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Aims and Scope

Journal of Electrodiagnosis and Neuromuscular Diseases (JEND) is a peer-reviewed journal concerning both normal and abnormal functioning of the muscle, the neuromuscular junction, and the peripheral motor, sensory and autonomic nerves. The journal publishes clinical studies, reviews, and case reports in the fields of electrophysiology, electrodiagnosis, imaging studies including ultrasonography, and management, about neuromuscular diseases. The journal is aimed to provide an open forum for original research in basic science and clinical research that will improve our fundamental understanding and lead to effective treatments of neuromuscular diseases.
Journal of Electrodiagnosis and Neuromuscular Diseases (JEND) is the official journal of the Korean Association of EMG Electrodiagnostic Medicine.

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신경통성 근위축증

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Neuralgic Amyotrophy

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In clinical practice, neuralgic amyotrophy (NA) is being frequently underdiagnosed, and its actual incidence is reported to be 1/1000 cases per year. Here, we present an overview of the essentials on NA. The cause of NA is known to be multifactorial (immunologic, mechanical, or genetic factors). Typically, patients with NA show sudden onset of pain in the shoulder region, followed by patchy flaccid paralysis of the muscles in the upper extremity. NA is suspected based on the clinical history and physical examination. However, prior to confirming the diagnosis of NA, other disorders showing similar clinical presentations with NA, such as cervical radiculopathy or rotator cuff pathology, should be ruled out. Electromyography, nerve conduction study, High-resolution ultrasound and magnetic resonance neurography are useful for confirming the diagnosis and judging the necessity of surgical operation. The prognosis depends on the degree of weakness of muscles innervated by involved nerves and the degree of axonal damage in nerve conduction study. Our review would be helpful for clinicians to diagnose NA and treat the symptoms of NA appropriately.

Keywords: Neuralgic amyotrophy; Muscle weakness; Pain; Diagnosis

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서론

신경통성 근위축증(neuralgic amyotrophy)은 주로 견갑부에서 갑작스럽게 발생하는 통증 이후 발생하는 일부 상완 근육의 이완성 마비(patchy flaccid paralysis)를 특징으로 하는 신경병증이다. 처음으로 1948년 Parsonage et al. 이 위와 같은 특징적 임상양상을 보이는 환자 136명을 모아 보고하였고, 이후 이를 처음으로 보고한 연구자들의 이름을 따서 "파르소니지-터너 (Parsonage- Turner) 증후군"이라고도 불려왔다[1]. 또한, 신경통성 근위축증은 급작스러운 염증에 의해 발생되는 것으로 여겨져, "상완신경총염 (brachial plexitis)"이라고도 불리기도 한다. 신경통성 근위축증이 처음 보고된 후 초창기에는, 신경통성 근위축증은 상완신경총에만 영향을 미치는 질환으로 알려져 있었지만, 현재는 전방골간신경 (anterior interosseous nerve), 요골신경 (radial)과 같은 상지의 신경 외에도 요천추신경총 (lumbosacral plexus)와 같은 하지의 말초신경 등의 여러 말초신경에도 영향을 미치는 것으로 알려졌다[2-4].

많은 문헌들에서 신경통성 근위축증은 희귀질환으로 기술되

어 있다. 그러나, 최근 연구에서 신경통성 근위축증은 임상의료 현장에서 심각하게 과소 진단되고 있으며, 실제 발생률은 1/1000이 될 것이라고 보고되었다[5]. 급성기 신경통성 근위축증 환자의 임상양상은 다른 근골격계 질환들과 증상이 일부 중복되는 경우가 많기에 의원이나 응급실에서 신경통성 근위축증 환자들이 윤활낭염 (bursitis)과 같은 어깨관절 병증 (shoulder joint pathology) 혹은 근육 염좌 (muscle strain)로 오진 되는 경우가 흔히 있다. 본 종설에서는 임상에서 신경통성 근위축증의 정확한 진단과 적절한 치료를 위해, 신경통성 근위축증의 특징적 임상양상과 진단 및 치료에 대해 기술하고자 한다.

역학

과거에는 신경통성 근위축증은 희귀질환으로 여겨졌으며, 발병률은 연간 100,000명 당 1~3명 정도로 알려져 왔으나, 최근의 연구에서는 이러한 발병률은 신경통성 근위축증에 대해 질환과 이의 임상양상에 대한 임상 의사들의 무지로 인해 비롯되었으며, 실제적으로는 연간 1,000명 당 1명이라는 보고가 있었다[5]. 신경통성 근위축증의 남녀의 성비는 2:1로 남자에서 더 호발하는 것으로 알려져 있다[5]. 신경통성 근위축증은 모든 연령에서 발생할 수 있으나, 평균 발병 연령은 40세 전후에 가장 많이 발생하는 것으로 알려져 있으며, 유전성 신경통성 근위축증의 경우의 경우 25세 전후에서 흔히 발병한다고 알려져 있다[6]. 또한, 유전성 신경통성 근위축증의 경우 75%에서 재발한다고 알려져 있다[6].

병인

신경통성 근위축증의 정확한 병태생리학적 기전이 확립되지는 않았지만, 이의 발생에는 다양한 요인들이 관련되어 있다고 알려져 왔다. 일반적으로 주로 면역학적 요인, 기계적 요인, 유전적 요인이 신경통성 근위축증의 발생과 관련이 있는 것으로 알려져 있다.

1) 면역학적 요인

신경통성 근위축증 환자의 절반 이상에서 감염, 예방접종, 수술, 임신, 신체적 또는 정신적 스트레스와 같은 면역계를 자극하는 선행인자가 확인되었다[6, 7]. 감염의 경우 세균 감염, 바이러스 감염 모두 신경통성 근위축증의 선행인자로 고려될 수 있는데, 최근 연구에서 인플루엔자 (influenza), 헤르페스 바이러스 (herpes virus), 수두대상포진 바이러스 (varicella-zoster virus) 외에도 E형 간염 바이러스 감염 (hepatitis E virus) 또한 하나의 요인이라는 보고가 있었으며, 신경통성 근위축증 환자의 약 10%에서 E형 간염 바이러스 감염 이 동반되어 있는 것으로 보고되었다[3,8-9].

일부 연구에서 급성 또는 아급성 신경통성 근위축증 환자를 대상으로 말초신경 생검을 실시하였다[10-13]. 환자의 상완신경총 생검에서, 림프 신경외막 혈관주위의 T 세포(T-cell) 침윤과 축삭 손상 및 신경 외막의 비대소견이 관찰되었다. 그 외에도 CD8+ T림프구 (CD8+ T-lymphocyte), CD68+ 대식세포 (CD8+ Macrophage), CD20+ B림프구 (CD20+ B-lymphocyte)들이 병발된 말초신경 주위의 혈관으로 침윤되어 있었다[10-13]. 그리고, 급성기 신경통성 근위축증의 T2-가중 자기공명영상 (T2-weighted magnetic resonance imaging)에서 이환된 말초신경이 고신호를 보이는 것은 신경통정 근위축증의 발병에 면역학적 요인이 관여한다는 가설을 뒷받침하는 것으로 보인다[14].

2) 기계적 요인

유의하게 많은 수의 신경통성 근위축증 환자들에서 일반 인구와 비교하여 스포츠 활동과 같이 과격한 활동 등 말초신경에 기계적 스트레스 (mechanical stress)를 유발할 수 있는 선행 사건들이 있었던 것으로 보고되었으며, 신경통성 근위축증 환자의 약 10%의 경우에서 격한 상지 운동의 과거력이 확인되었다[6]. 가장 흔히 신경통성 근위축증에 의해 침범되는 말초신경이 상완신경총 중 특히 상부줄기 (brachial plexus, upper trunk)이라는 사실은 이 질환의 발생이 기계적 요인에 기인할 수 있다는 가설을 뒷받침할 수 있다고 생각된다[15]. 견관절은 광범위한 운동범위를 가지는 관절로서 인체에서 가장 가동성이 높은 관절이다[16]. van Alfen et al.. [27]은 이러한 견관절의 광범위한 운동 범위와 가동성으로 인해 상완신경총 주변의 혈액-신경 장벽에서 마모가 발생하고, 이로 인해 약화된 혈액-신경 장벽으로 인해 면역 인자나 세포가 상완신경총에 접촉하여 신경통성 근위축증이 호발 될 수 있다 주장하였다[7].

1976년 Englert et al. [17]이 모래 시계 모양의 협착(hourglass-like constrictions)에 의한 신경 마비를 최초로 보고한 이후 몇몇 연구들에서 고해상도 초음파 (high-resolution ultrasound), 자기공명영상 신경조영술(magnetic resonance neuropathy)을 영상검사로 사용해 이 질환을 진단하였다[17-19]. 이 모래 시계 모양의 협착이 신경통성 근위축증으로 진단된 환자에게서 확인되었는데, 이러한 사실이 신경통증 근위축증 발생에서의 기계적 요인을 설명한다는 보고가 있었다. 이 모래 시계 모양의 협착은 신경통성 근위축증의 아형으로 분류되기도 한다.

3) 유전적 요인

일부 환자들에서는 신경통성 근위축증이 재발되기도 하는데, 이러한 환자에게서는 유전적 요인이 연관되어 있는 것으로 생각된다. 이 경우를 유전성 신경통성 근위축증(hereditary neuralgic amyotrophy)라 부른다[6,7]. 유전성 신경통성 근위축증은 상염

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색체 우성 희귀질환으로, 특발성 신경통성 근위축증 보다 대략 10배 정도 드물게 발생한다[20]. 유전성 신경통성 근위축증 환자의 약 절반이 17q25.3 염색체의 점 돌연변이(point mutation) 또는 유전자 SEPT9의 복제와 관련이 있다고 알려져 있다[20-22]. 또한, 신경통성 근위축증 환자의 10%에서 가족력이 확인되었는데, 이는 유전적 요인이 신경통성 근위축증의 발생과 관련이 있을 수 있음을 나타낸다[7].

임상양상

신경통성 근위축증의 전형적인 임상양상은 견갑부의 갑작스러운 극심한 통증 후 발생하는 일부 상완 근육의 이완성 마비(patchy flaccid paralysis) 및 근 위축이다. 흔히 한 쪽 사지에서만 병발하는 경우가 많으나, 환자의 10~30%에서 양측으로 병발한다[6].

상완신경총에 신경통성 근위축증이 발생한 환자의 대부분이 초기 증상으로 견갑부의 수 시간 내에 갑작스럽게 나타나는 급성 통증을 호소하는데, 이 통증은 목, 상완, 그리고 전완으로 방사상으로 퍼질 수 있다. 1~2%의 소수의 환자에서만 목, 견갑 또는 상완 등의 제한된 부위에만 통증이 있다[6,7]. 또한, 60%의 환자에서 통증이 야간에 시작되며 이에 많은 환자들이 중증도의 통증으로 인해 아침 일찍 잠에서 깨며, 이후 수 시간 내로 통증이 최대 중증도로 증가한다[6,7]. 통증의 양상은 보통 "예리한(sharp)", "찌르는 듯한(stabbing)", "욱신거리는 (throbbing)" 또는 "쑤시는(aching)" 양상으로 나타나며, 그 강도는 일반적으로 numeric rating scale (NRS) 7점 이상으로 심한 수준이다 [6,7]. 통증은 사지의 움직임에 의해 악화될 수 있다. 통증 지속시간은 광범위해서, 수 시간에서 수 주 동안 지속될 수 있으며 약 5%의 환자의 통증은 24시간 내에 호전이 되나, 10%는 2개월 넘게 지속된다, 평균적인 통증의 지속시간은 4주라고 알려져 있다[6,7].

일부 상완 근육의 근력저하 또한 신경통성 근위축증의 두드러진 증상이며, 특징적으로 통증의 중증도가 낮아질 때 발생한다. 신경통증 근위축증의 초기 통증 발생 후, 70%의 환자가 2주내 근력저하를 호소하며, 특히 30%가 24시간 내의 근력저하를 호소한다. 약 30%의 환자에서는 통증이 시작된 후 2주 이후에 근력저하가 발생할 수 있다. 근력저하는 일반적으로 환자의 약 70%에서 상완신경총 상부 줄기(upper trunk)의 지배를 받는 근육에서 병발하며, 그 중 약 70%에서 긴 흉벽신경(long thoracic nerve)의 지배를 받는 근육에서도 병발한다[7]. 다음으로 흔한 병발 부위는 상완신경총의 중간 및 하부 줄기(middle and lower trunk)이다. 상완신경총 외의 신경의 침범 또한 가능한데, 요천추신경총(lumbosacral plexus), 전방 및 후방 골간신경(anterior and posterior interosseous nerves), 횡격신경(phrenic nerve), 척추 신경근 등 에서도 병발 가능하다[6].

신경통성 근위축증은 한 쪽 사지에서만 병발하는 경우가 많지만, 환자의 10~30%에서는 양측(보통 비대칭적으로)으로 병발한다[6]. 일반적인 특발성 신경통성 근위축증 환자의 경우보다 유전성 신경통성 근위축증의 경우에서 상완신경총 외 신경침범이 흔하다. van Alfen et al. [27]에 따르면 특발성 신경통성 근위축증 환자의 17.3%에서 상완신경총 외 신경의 병발이 확인되었으며, 요천추신경총 (8.2%), 횡격신경 (6.6%), 되돌이후두신경 (2%), 기타 (2.6%) 순으로 그 빈도가 확인되었다. 유전성 신경통성 근위축증의 경우에서는 55.8%에서 상완신경총 외 신경에서 질환의 발생이 확인되며, 요천추신경총(32.6%), 횡격신경(14%), 되돌이후두신경(18.6%), 기타(7%) 순으로 그 빈도가 확인되었다[6].

신경통성 근위축증의 임상양상 중 감각증상은 환자의 70~80%에서 발현되는 것으로 알려져 있지만, 보통 근력저하 정도에 비해서는 경미하다[6]. 감각과민(hyperesthesia) 또는 감각이상(paresthesia)이 가장 일반적인 감각 증상이며, 감각저하(hypoesthesia) 역시 증상으로 나타날 수 있으며 삼각근 및 외측 상완 부위가 감각증상이 나타나는 가장 흔한 부위이다[6]. 매우 드물지만, 비복신경(sural nerve)과 천요골 감각신경(superficial radial nerves)를 침범한 근력저하가 동반되지 않은 순수한 감각증상만을 가진 경우도 발생한다[7,23].

진단

신경통성 근위축증은 임상 병력과 신체 검진을 토대로 의심된다[24]. 하지만, 임상양상이 다른 질환과 비슷한 경우가 많아, 확진을 위해서는 이 질환들이 배제되어야 한다. 다른 감별진단을 감별하고 신경통성 근위축증을 확진하기 위해, 실험실 검사, 전기진단검사 및 영상학적 검사가 이용될 수 있다.

실험실 검사 중, 신경통성 근위축증의 진단을 위한 면역반응을 증명할 수 있는 유효한 혈액 검사는 없으며, 질병이 전신감염 또는 결합조직질환과 관련된 경우가 아닌 한 적혈구침강속도 (erythrocyte sedimentation rate) 및 전혈구수 (complete blood count) 또한 일반적으로 정상이다[21,24]. 그 외에 동반된 바이러스 감염 및 신경통성 근위축증에 선행되는 기타 감염의 증거를 확인할 수 있는 경우가 있는데, HEV 감염에 동반한 경우 양측으로 신경증상이 발생하며, 실험실 검사에서 간 효소 수치의 상승 등을 확인 가능하다[8,21,25]. 뇌척수액 검사로는 백혈구 증가가 동반되지 않은 경미한 단백질 상승을 확인할 수 있다[21,24].

전기진단검사는 신경통성 근위축증의 진단 및 말초신경 병발 부위의 위치를 확인할 수 있으며 신경 손상의 정도를 평가할 수 있다[24,26]. 그러나, 급성기에는 전기생리학적 검사에서 비정상 소견이 확인되지 않을 수 있다. 비정상 소견은 신경전도 검사 및 근전도 검사에서 질환의 발현 후 각각 약 1주, 3주 후에 나타난다[26,27]. 운동 신경전도 검사에서는, 이환된 신경에서 복합근

육 활동전위 진폭의 감소를 확인할 수 있다. 감각 신경전도 검사에서는 30%~45%에서 이상 소견이 발견될 수 있다[27]. 근전도 검사에서는, 이환된 말초신경의 지배를 받는 근육에서 비정상 자발전위(abnormal spontaneous activity)인 양성 예파(positive sharp waves)와 세동 전위(fibrillation potential)가 나타난다[24,26]. 경추부 신경근병증 (cervical radiculopathy)은 임상양상이 근위축증 신경병증과 유사하여 반드시 감별이 필요한데, 근전도 검사가 도움이 될 수 있다. 경추 주위근(paraspinalis muscles)에 대한 침 근전도 검사에서 경추부 신경근병증의 경우 약 47%의 환자에서 비정상 자발전위가 확인되나, 신경통성 근위축증의 경우 1.5%의 환자에서만 비정상 자발전위가 확인됨이 보고되었다[6,28].

이 외에도 영상학적 검사가 신경통성 근위축증의 진단 및 감별진단을 위해 시행될 수 있다. 고해상도 초음파 영상검사에서는 이환된 신경의 국소적인 분절 팽창(segmental swelling), 협착(constriction), 염전(torsion) 등이 확인 될 수 있다[29]. 하지만 고해상도 초음파 영상검사의 민감도는 74%로 알려져 있으며, 이는 상완신경총이 아닌, 액와부 이하 부위의 상완 신경에서만 비정상 소견을 확인 가능한 것으로 알려져 있어 제한이 있다[29].

기존 고식적 자기공명영상(conventional MRI)에서는 신경통성 근위축증이 이환된 신경이 지배하는 견관절 주위 일부 근육 내의 탈신경성 변화(intramuscular denervation change)를확인할 수 있다[30]. 가돌리늄 조영 자기공명 영상에서는 이환된 신경의 염증 부위가 고 신호 강도(high signal intensity)로 확인되어 도움이 될 수 있다[31]. 하지만 신경통성 근위축증의 병변 확인에 있어 가돌리늄 조영 자기공명영상의 민감도가 낮아 이 질환에서의 신경의 병적인 변화를 확인하기에는 불충분하였다. 하지만 최근 새로이 개발된 자기공명 영상 신경조영술은 지방과 혈관의 영상신호를 억제함으로써, 말초신경의 고해상도 영상을 제공한다[32]. 이전 증례 보고들에서, 신경통성 근위축증 환자의 자기공명 영상 신경조영술 영상에서, 상완신경총을 포함하여 이환된 신경들의 비대 및 고 신호 변화를 확인할 수 있었다[18,19]. 하지만 이러한 상완신경총과 말초신경의 고 신호 변화는 경수 신경근병증 에서도 관찰될 수 있는 소견으로, 감별 진단에 있어 제한이 있을 수 있다[33]. 또한 신경통성 근위축증의 아형으로 분류되는 모래 시계 모양의 협착의 경우 자기공명 영상의 축상면(axial view)에서 특징적인 소견인 "bull's eye sign"을 확인함으로써 진단 가능하다[34].

초음파 및 자기공명 영상을 이용한 영상학적 검사는 신경통성 근위축증의 병변을 확인하는 역할 외에도, 확진 전 비슷한 임상양상을 보이는 추간판 탈출증, 척추협착증, 회전근개질환등의 근골격계 질환을 감별하기 위해 필수적이다[7].

감별진단

신경통성 근위축증은 이 질환과 유사한 증상을 보이는 다른 질환들로 오진되는 경우가 많다. 신경통성 근위축증의 확진을 위해 임상의들은 경추 신경근병증 및 회전근개 파열과 같은 기타 질환을 감별해야 한다[7]. 신경통성 근위축증의 확진을 위해 임상의가 배제해야 하는 질병과 그 각개의 임상 특성을 Table 1에 요약하였다.

치료

대부분의 신경통성 근위축증 환자들은 심한 통증을 호소하기에 다양한 진통제를 이용하여 환자의 통증에 적극적으로 개입해야 하며, 이에 비스테로이드성 진통제(non-steroidal anti-inflammatory drug)와 아편양 진통제(opiate)가 고려될 수 있다[10]. 그러나, 신경통성 근위축증의 회복을 위한 치료 방법은 매우 제한적인 상황이며, 이에 대한 이전의 연구도 경험적인 결과를 보고한 경우가 대부분인 실정이다. 이전의 일부 연구들에서, 2주간의 corticosteroid의 조기 투여(60mg/day 1주간 투여 후 감량)를 시행한 경우 대조군에 비해 통증 기간이 단축되었으며 (12.5일 vs 20.5일), 일부 환자의 근력회복에 긍정적인 영향을 미칠 수 있다는 보고가 있었다 (18% vs 6.3%) [21,35]. 하지만 van Alfen

Table 1. Disorders That Should Be Ruled Out Before Diagnosing Neuralgic Amyotrophy And Their Clinical Characteristics

Disorders	Clinical characteristics
Cervical radiculopathy due to herniated cervical disc	Acute onset, spurling sign (+), sensory and motor deficits in the same dermatome
Cervical radiculopathy due to cervical foraminal stenosis	Insidious onset, slow progression, spurling sign (+), sensory and motor deficits in the same dermatome
Rotator cuff tear	Pain during shoulder movement, easily differentiated by ultrasound
Mononeuritis multiplex (or vasculitic neuropathy)	Sudden onset, severe pain, distal parts of the limbs usually involved, elevated C-reactive protein, skin lesion (e.g. purpura, petechiae, and ulcer)
Multifocal motor neuropathy	Slow progression, no sensory symptom, no pain, distal parts of the limbs usually involved
Hereditary neuropathy with pressure palsies	Recurrent episodes of palsy, family history, focal neuropathy at susceptible pressure points
Complex regional pain syndrome	Diffuse pain, predominant vasomotor feature, history of stroke, trauma, or peripheral nerve injury

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et al. [27]의 메타분석 연구의 결론에서 corticosteroid는 신경통성 근위축증의 치료에 유의하게 긍정적인 효과가 없다고 확인되었으며, 장기 회복에 대한 효과 또한 분명하게 입증되지 않은 것으로 나타났다[20].

면역글로불린(immunoglobulin, IVIG)의 투여 후 임상양상이 호전된 증례는 많이 있어 왔으나, 역시 무작위 배정 임상 연구는 없었고 효과에 대한 증거가 부족한 실정이다[20,36,37].

3개월 동안 회복에 대한 임상 징후가 나타나지 않을 경우 자기공명 영상 신경조영술이나 고해상 초음파를 통한 신경의 영상학적 평가가 권장되며, 신경 협착(constriction)이 확인되면, 외과적 수술을 고려할 수 있다. 신경 협착으로 인한 신경 두께 감소 비율이 75% 미만인 경우 속내 신경박리술(intrafascicular neurolysis) 이 권장되며. 신경 협착이 75% 이상인 경우에는 신경봉합술(neurorrhaphy) 혹은 이식(grafting)을 고려해야 한다[25].

이전 한 연구에서는, 수술적 치료 없이, 재활치료를 시행한 환자에서 14개월 후 근위축의 변화에 대한 평가 시행하였으나, 근육의 크기에 변화를 보이지 못하였다[38]. 하지만, 신경통성 근위축증은 근력저하와 근 위축을 야기함으로써 병발 사지의 불안정성 및 관절 구축을 야기할 수 있어 일반적 근력 강화 목적이 아닌, 관절구축의 예방과 생체역학적 안정성(biomechanical stability)을 최대화하기 위한 목표의 재활치료는 도움이 될 수도 있다[24].

예후

신경통성 근위축증의 예후는 곁가지 재신경 지배(collateral reinnervation)가 일어나는 정도에 따라 달라진다. 전체적으로 발병 2~3년 후 이전 상태의 80~90%를 회복하지만, 70% 의 환자 경우에서는 근력저하가 잔존한다[6,7]. 전기진단검사와 근력저하에 대한 평가가 신경통성 근위축증의 예후 예측에 도움이 될 수 있다. 만약 전기진단검사상 복합근육활동전위(compound muscle action potential) 진폭이 70% 이상하여 감소하거나 첫 평가 당시의 근력저하가 Medical Research Council 상 3점 미만으로 저하되었다면 곁가지 재신경 지배가 불완전하여 예후가 좋지 않을 가능성이 있다[7].

결론

임상 의료 현장에서 임상의사들은 통증을 동반한 근력저하를 호소하는 많은 환자들을 만나게 되는데, 이들 중 신경통증 근위축증 환자가 있을 수 있다. 그러나 이 질병에 대한 무지로 인해 실제 임상 현장에서 오진되는 경우가 많다. 우리는 본 종설에서 신경통증 근위축증에 대한 필수 정보에 대해 기술하였고, 이 질환의 정확한 진단과 치료에 도움이 될 것이라 기대한다.

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신경전도검사를 이용하여 경추 6-7 신경공협착에 의한 경추 포착 신경병증과 감각신경 활동전위의 연관성 연구

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Relationship Between Cervical Nerve Root Entrapment Due to Cervical 6-7 Foraminal Stenosis and Sensory Nerve Action Potentials in Nerve Conduction Study

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Objective: To compare the sensory nerve action potentials (SNAPs) between patients with cervical 6-7 foraminal stenosis (FS) and those without.

Methods: We enrolled patients who underwent a bilateral nerve conduction study (NCS) between January 2016 and December 2017. The FS group (n=35) included patients whose lesion was located at C6-7 FS. The non-FS group (n=75) had no lesions at C6-7 foramen. The amplitudes and latencies of SNAPs on the affected side were compared between the two groups. The proportion of those with abnormal SNAP responses was also evaluated.

Results: Compared with the non-FS group, the FS group had a significantly lower amplitude (30.5 vs. 40.2 ÄV; age-adjusted p<0.001) and a longer latency (3.31 vs. 3.17 ms; age-adjusted p=0.001) of the median nerve. Abnormal amplitude was observed in 20.0% of the FS group and 4.0% of the non-FS group (age-adjusted p=0.007), and abnormal latency was observed in 45.7% of the FS group and 26.7% of the non-FS group (age-adjusted p=0.010). The median SNAP ratio between the affected side and the unaffected side was not significantly different between the two groups.

Conclusion: Patients with C6-7 FS had a lower amplitude and a longer latency of the median nerve than did those without. Abnormal SNAP amplitude and latency were significantly more common in patients with FS. SNAP amplitude may be used as a predictor of cervical FS.

Keywords: Electrodiagnosis; Cervical foraminal stenosis; Sensory nerve action potential; Median nerve

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서론

척추에서 신경근이 나가는 길은 해부학적으로 척주관내, 신경공내, 신경공외 구역으로 나뉜다. 신경공을 통해서 운동신경과 감각신경이 말초와 중추신경계로 이어지며 특히, 신경공내에는 후근신경절이 위치하고 있다. 따라서 신경공내, 신경공외에서 신경공협착이나 추간판탈출증 등 신경을 압박할 수 있는 질환으로 후근신경절이 눌리게 되고, 이로 인해 통증, 이상감각과 신경학적 감각증상이 유발되고, 운동신경의 압박으로 근위약이 발생할 수 있다[1-3]. 신경근 손상을 진단하기 위해서 신체진찰뿐만 아니라, 단순방사선 사진, 컴퓨터 단층 촬영, 자기공명영상, 전기진단검사 등을 시행한다. 자기공명영상에서 구조적인 이상 소견이 관찰되지 않으나 신경근병증이 있는 경우도 있다. 이는 영상검사의 높은 위음성 비율과 관련이 있다[4]. 이와 같은 경우에는 전기진단검사가 유용한 진단 도구이다[2].

한편, 신경근병증에서 감각신경전도검사는 대체로 정상소견을 보이나, 복합근활동전위와 F파는 이상소견을 보일 수 있다[5,6]. 신경근병증은 병변 위치가 후근신경절보다 근위부위에 있는 경우, 감각신경전도검사는 정상소견을 보인다. 또한, 통증과 관련된 C섬유는 전 도속도가 느려서 통상적인 감각신경전도검사에서는 이상 소견이 관찰되지 않는다[5]. 이로 인해 감각증상이 있는 환자에서 신경공 협착이 있음에도 불구하고 근전도 검사에서 정상인 경우가 흔하다.

Muneharu 등[7]은 요추5번 신경근병증이 있는 환자를 신경공내 또는 신경공외 협착이 있는 환자와 척주관협착이 있는 환자로 나누어서 표재비골신경의 감각신경 활동전위를 비교하는 연구를 하였다. 감각신경 활동전위는 건측과 환측의 비율로 비교하였다. 신경공내 또는 신경공외 협착이 있는 환자에서 표재비골신경의 감각신경 활동전위 진폭의 비율이 낮게 나타났다.

Mondelli 등[8]의 연구에서는 요추5번 신경병증에서 표재비골신경 활동전위의 진폭이 감소되었으며, 천추1번 신경병증에서 비복신경 활동전위의 진폭이 감소되었으며, 이들은 모두 신경공 협착이 있는 환자였다.

요추에서 신경공 협착과 감각신경 활동전위의 연관성을 본 연구는 있으나, 경추의 신경공 협착과 감각신경 활동전위의 연관성에 대한 연구는 아직 없다. 이에 따라 본 연구는 경추의 신경공 협착증이 있는 환자와 없는 환자에서 감각신경의 활동전위의 잠시, 진폭이 어떤 상관성이 있는지 확인하고자 하였다.

연구 대상 및 방법

1) 연구대상

2016년 1월 1일부터 2017년 12월 31일까지 서울아산병원 재활의학과에서 상지에 대해 전기진단 검사를 시행한 환자를 대상으로 후향적 조사를 실시하였다.

포함기준으로 목과 상지에 통증, 저림감, 둔한감각, 위약이 있는 환자, 전기진단 검사를 양측으로 시행한 환자, 자기공명영상 검사 결과가 있는 환자를 대상으로 하였다. 제외기준으로는 정중신경병증, 척골신경병증, 팔신경얼기병증, 말초신경병증이 있거나, 이전에 경추 수술을 받은 과거력이 있거나, 경추 6-7의 신경공 부위에 낭종, 암 등 종괴가 있거나, 자기공명영상 검사상 경추 6-7에 양측이 동일하게 신경공협착이 있는 환자로 설정하였다.

2) 연구방법

환자의 의무기록을 열람하여 근전도검사 기록, 자기공명영상 검사를 확인하였다. 전기 진단 검사는 NicoletTM (Natus neurology, Middleton, U.S.A.) 근전도 기기를 이용하였으며, 소인 속도 10 ms, 민감도 20 μV, 여과 범위는 20 Hz에서 2 kHz 로 하였다. 표면 전극은 Ambull®사의 118mm × 86 mm 타원형 전극을 이용하였다. 근전도실의 실내온도는 26˚C에서 28˚C 사이를 유지하였다.

근전도검사는 정중신경의 감각신경 활동전위를 확인하였다. 활동전극과 참고전극을 정중신경은 3번째 손가락에 부착하였으며, 활동전극은 첫마디뼈에, 참고전극은 끝마디뼈에 부착하여 측정하였다. 그리고 활동전극으로부터 근위부 14cm의 손목에서 최대초과자극을 역방향으로 자극하였다. 해당 연구는 정점잠시(peak latency)와 기저선 음극정점진폭 (baseline to peak amplitude)를 사용하였다. 정상 값은 정중신경의 잠시는 3.3 ms [9] 이며, 정중신경의 진폭은 20 μV [6] 로 하였다.

자기공명영상검사에서 경추6-7의 신경공협착 유무에 따라 2개 그룹으로 나누었다. 신경공협착이 일측에만 있거나, 양측에 있어도 영상 검사의 판독상 협착의 중증도가 한쪽이 더 심하게 있는 경우에는 신경공 협착이 있는 그룹으로, 신경공 협착이 없는 경우에는 신경공 협착이 없는 그룹으로 나누었다. 신경공 협착이 있는 그룹에서는 신경공 협착이 있는 쪽을, 신경공 협착이 없는 그룹에서는 증상이 있는 쪽을 영향을 받은 쪽으로 설정하고 분석하였다.

두 그룹간 비교를 위해서 정중신경의 진폭과 잠시의 절대값 및 건측과 환측의 비율을 사용하였으며, 이상값을 가진 환자수의 백분율을 확인하였다.

3) 통계 분석

통계 분석은 윈도우 용 SPSS version 18.0을 이용하였고, 통계학적 유의성은 $p<0.05$ 수준으로 하였다. 환자 특성은 독립 표본 t 검정을 사용하였으며, 성별과 영향을 받은 부위의 좌우 비율에 대한 비교는 Pearson 카이제곱 검정을 사용하였다. 각 그룹간의 감각신경 활동전위 값과 비율의 비교는 독립 표본 t 검정을 사용

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하였고, 신경공 협착이 있는 그룹과 없는 그룹의 감각신경활동전위 비정상 수는 Pearson 카이제곱 검정 또는 Fisher의 정확한 검정을 사용하였다. 그룹간 비교에서 연령 보정을 위해 로지스틱 회귀분석을 사용하였다.

결과

2016년 1월부터 2017년 12월까지 2년간 본원에서 근전도 검사를 시행한 환자는 1564명이었으며, 이 중에서 자기공명영상 검사가 있는 환자는 539명 이었다. 정중신경병증, 척골신경병증, 팔신경얼기병증, 말초신경병증있거나, 이전에 경추 수술을 받은 과거력이 있거나, 경추 6-7의 신경공 부위에 종괴가 있거나, 자기공명영상 검사상 경추 6-7에 양측 신경공협착이 있는 환자를 제외하고 최종적으로 총 110명을 대상으로 분석하였다. 110명은 모두 목이나 상지에 신경학적인 증상을 동반하였다.

대상환자군 중 경추 6-7에 신경공 협착이 있는 그룹은 35명, 신경공 협착이 없는 그룹은 75명이었다. 신경공 협착이 있는 그룹에서 여자는 17명, 남자 18명 이었으며, 신경공 협착이 없는 그룹에서 여자는 38명, 남자 37명이었다. 양 그룹의 평균 나이는 각각 58.7 ± 10.7 세, 49.4 ± 13.1 세 이고, 체중은 각각 70.4 ± 13.4 kg, 63.1 ± 13.6 kg 으로 양 그룹간 유의미한 차이가 있었으나, 키, 신체질량지수, 영향을 받은 부위의 좌우 비율은 양 그룹간 유의미한 차이가 없었다 (Table 1).

경추 6-7에 신경공 협착이 있는 그룹과 없는 그룹에서 정중신경의 진폭은 각각 30.5 ± 12.4 μV, 40.2 ± 15.0 μV 로 유의미한 차이가 확인되었다 (p-value 0.003). 정중신경의 잠시의 경우 각각 3.31 ± 0.31 ms, 3.17 ± 0.38 ms 로 확인되었다 (p-value 0.090). 연령보정을 하였을 때는 정중신경의 진폭과 잠시 모두 통계적으로 유의미한 차이를 보였다 (연령보정 p-value는 각각 <0.001, 0.001) (Table 2).

감각신경 활동전위의 비정상 환자수를 비교하였을 때, 정중신경의 진폭과 잠시는 신경공 협착이 있는 그룹에서 20.0 %, 45.7 % 로 확인되었으며, 신경공 협착이 없는 그룹에서는 4.0 %, 26.7 % 로 나타났다 (p-value는 각각 0.011, 0.047). 연령 보정을 시행하였을 때는 정중신경의 진폭과 잠시 모두에서 통계적으로 유의미한 차이가 확인되었다 (연령보정 p-value는 각각 0.007, 0.010) (Table 3).

건측과 환측의 비율을 경추 6-7에 신경공 협착이 있는 그룹과 없는 그룹에서 비교하였을 때, 정중신경의 진폭은 1.04 ± 0.43. 1.08 ± 0.42 로 나타났으며, 잠시는 0.99 ± 0.10, 1.00 ± 0.05 로 유의미한 차이는 관찰되지 않았다 (연령보정 p-value는 각각 0.108, 0.439) (Table 4).

고찰

정중신경의 감각신경 활동전위는 경추 신경공 협착의 예측인

Table 1. Baseline Characteristics

	C6-7 foraminal		p-value
	Stenosis (+) (n = 35)	Stenosis (–) (n = 75)	
Sex (Female : Male)	18:17	37:38	0.941†
Age (years)	58.7 ± 10.7	49.4 ± 13.1	<.001
Weight (kg)	70.4 ± 13.4	63.1 ± 13.6	0.023
Height (cm)	163.1 ± 9.5	163.2 ± 10.1	0.984
BMI (kg/m^2)	26.3 ± 3.3	24.2 ± 7.3	0.169
Side of the foraminal stenosis or symptoms (Right : Left)	15:20	29:46	0.676†

Values are mean ± SD.
†Pearson chi-square test; otherwise independent t-test was used.

Table 2. Comparison of the Median Nerve Sensory Nerve Action Potential Parameters between Patients with C6-7 Foraminal Stenosis and Those without

		C6-7 foraminal		p-value*	Age-adjusted p-value**
		Stenosis (+)†	Stenosis (–)††		
Median nerve	Amplitude (μV)	30.5 ± 12.4	40.2 ± 15.0	0.003	<.001
	Latency (ms)	3.31 ± 0.31	3.17 ± 0.38	0.090	0.001

Values are mean ± SD.
†Value of the affected side was used.
††Value of the symptomatic side was used.
*Independent t-test.
**Univariate logistic regression analysis.

Table 3. Comparison of the Proportion of Patients with Abnormal Median Nerve Sensory Nerve Action Potential Parameters between Patients with C6-7 Foraminal Stenosis and Those without

		C6-7 foraminal		p-value	Age-adjusted p-value***
		stenosis (+)	stenosis (–)		
Median nerve	Patients with abnormal amplitude (%)	7 (20.0)	3 (4.0)	0.011*	0.007
	Patients with abnormal latency (%)	16 (45.7)	20 (26.7)	0.047**	0.010

Cut-off values for abnormal findings in the median nerve: latency, 3.3ms; amplitude, 20μV.
*Fisher's exact test.
**Pearson Chi-square test.
***Binary logistic regression analysis.

Table 4. Comparison of the Median Nerve Sensory Nerve Action Potential Ratio between Patients with C6-7 Foraminal Stenosis and Those without

		C6-7 foraminal		p-value*	Age-adjusted p-value**
		stenosis (+)†	stenosis (–)††		
Median nerve	Ratio of amplitude (μV)	1.04 ± 0.43	1.08 ± 0.42	0.628	0.108
	Ratio of latency (ms)	0.99 ± 0.10	1.00 ± 0.05	0.399	0.439

Values are mean ± SD.
†The ratio between the affected side and the unaffected was used.
††The ratio between the symptomatic side and the asymptomatic side was used.
*Independent t-test.
**Univariate logistic regression analysis.

자로 사용 가능하다. 두 신경의 감각신경 활동전위 진폭과 정중신경의 감각신경 활동전위 잠시가 신경공 협착증이 있는 환자에서 유의미하게 감소되었다. 또한, 활동전위의 진폭과 잠시의 이상 소견이 있는 경우가 신경공 협착이 있는 환자군에서 유의미하게 더 많이 확인되었다.

Mondelli 등[8]의 연구를 보면, 요추 5번, 천추 1번에 신경근병증이 있는 환자의 7%에서 표재비골신경 또는 비복신경의 활동전위 진폭이 이상소견이 확인되었다. Mondelli 등은 감각신경의 활동전위의 진폭 감소가 신경근병증에서는 드물다고 하였지만 표재비골신경, 비복신경에서 이상소견이 있는 환자는 모두 신경공 협착이 있었다. 그리고 Ho 등[10]의 연구에서 요추 5번의 신경근병증이 있는 경우와 없는 경우를 비교하였을 때 표재비골신경의 이상이 요추 5번 신경근병증에서 더 많이 관찰되었다. 이러한 점은 본 연구에서 신경공 협착이 있을 경우, 감각신경 활동전위에 이상을 유발한다는 점과 일치한다.

Pawar 등[2]은 경추 신경근병증 진단을 보조하는 도구로써 근전도 검사가 유용하며, 특히 감각신경 활동전위가 운동신경 활동전위에 비해 특이성이 더 높다고 하였다. 하지만 해당 연구는 경추 신경근병증 진단 방법, 신경근병증의 위치에 대해서는 언급되어 있지 않으며, 신경근병증 위치의 구분 없이 비교분석을 진행하였다는 제한점이 있다. 본 연구는 정중신경과 경추 6-7의 신경공 협착증의 상관관계를 보았으며, 이는 Pawar의 연구처럼 감각신경이 신경근병증을 예측하는데 사용될 수 있음을 보여준다.

Muneharu 등[7]은 요추 5번 척주관협착이 있는 환자와 신경공 내 또는 신경공외 협착이 있는 환자를 대상으로 표재비골신경 감각신경을 확인하였으며, 신경공내 또는 신경공외 협착이 있는 환자에서 활동전위 진폭이 감소된 경향을 보였다. Muneharu의 연구는 요추 5번을 대상으로 한 연구이기는 하지만, 신경공 협착과 감각신경 활동전위의 상관관계를 확인한 연구이다. 이러한 점은 본 연구에서 경추 신경공 협착과 감각신경의 활동전위가 상관관계가 있다는 결론과 일치한다.

본 연구의 제한점으로 후향적 연구라는 점, 단일기관 연구이며 대상환자 수가 적다는 점, 자기공명영상검사와 근전도 검사 시기의 차이가 존재한다는 점이 있을 수 있다. 또한 신경공협착의 중증도에 따른 감각신경의 연관성을 비교하지 않았다. 따라서 향후 연구는, 전향적 연구로 다기관에서 대상환자 수를 늘려서 시행하고, 협착정도에 따른 연관성을 분석하는 것이 좋겠다. 또한 가능하다면, 신경공협착을 예측할 수 있는 기준값 확인을 고려하는 것이 필요하다.

결론

근전도 검사에서 정중신경의 감각신경 활동전위는 경추 신경공 협착을 예측하는데 유용한 것으로 판단된다. 따라서, 정중신경의 감각신경 활동전위가 비정상 이거나 진폭이 이상 소견이 있는 경우 신경공 협착을 의심하고 추가적인 검사를 고려해 봐야 한다.

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CASE REPORT

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길랑 바레 증후군으로 오인된 중증의 감각운동 다발성 신경병증 환자에서 급성 간헐 포르피린증 선별의 중요성: 증례 보고

손우철, 김자영, 김대열

울산대학교 서울아산병원 재활의학과

Importance of Screening Acute Intermittent Porphyria in Severe Sensorimotor Polyneuropathy Misdiagnosed as Guillain-Barré Syndrome: A Case Report

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Acute intermittent porphyria (AIP) is a rare genetic disorder. Its clinical manifestations include abdominal pain, neurological symptoms, and sensorimotor polyneuropathy. Herein we report the case of a 23-year-old woman who first presented with abdominal pain, followed by severe sensorimotor polyneuropathy, which was first reckoned as Guillain-Barré Syndrome (GBS) or critical illness polyneuropathy (CIP). Her clinical findings and laboratory data corresponded to AIP, which can cause severe peripheral axonal polyneuropathy, that is, porphyric neuropathy. On the basis of a review of the present case, we confirmed that porphyric neuropathy should be considered as a differential diagnosis in patients with an acutely progressing severe symmetric sensorimotor polyneuropathy with particular similarities to GBS or CIP because considerable recovery can be expected in patients with porphyric neuropathy who receive an early diagnosis and treatment.

Keywords: Porphyria, Neuropathy, Guillain–Barré Syndrome, Critical illness polyneuropathy

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Introduction

Acute intermittent porphyria (AIP) is a rare genetic disorder that induces severe disabilities. The development of symptoms is associated with various exacerbating factors, from medications to poor oral intake or stress, in patients with genetic factors. The clinical manifestations of AIP include abdominal pain, neurological symptoms, and sensorimotor polyneuropathy. Considerable recovery can be expected in cases that receive early diagnosis and treatment [1,2].

Porphyric neuropathy is typically described as a motor-dominant axonal neuropathy with preceding abdominal pain (from a few days to weeks). Its symptoms, signs, and results of cerebrospinal fluid (CSF) analysis are similar to those of Guillain-Barré

Syndrome (GBS) or Critical illness polyneuropathy (CIP). Clinical features such as asymmetric, proximal dominant weakness, accompanied with psychosis or abdominal pain, as well as axonal-type polyradiculopathy or polyneuropathy verified by nerve conduction studies, can be used for differentiating porphyric neuropathy from GBS or CIP [3]. However, AIP is rare disease that is difficult to diagnose at the first inspection of clinical data; therefore, patients with AIP rarely receive a diagnosis at an early disease stage. Here, we described the clinical course in a patient with AIP that was first reckoned as other polyneuropathies.

Case report

A 23-year-old unmarried woman without any medical history presented to the hospital with cramping abdominal pain on February 11, 2018. An abdominal computed tomography (CT) scan showed two sites of intussusception. She underwent laparoscopic manual reduction and postoperative care with limited oral intake. She was discharged from the hospital 10 days following the intervention.

Six hours after being discharged, she experienced a sudden deterioration in her consciousness level and suffered four serial convulsions lasting for 3-5 min each. The convulsions were of a generalized tonic-clonic type. Brain CT and CSF tapping were normal, but the patient's condition continued to deteriorate with the requirement for admission to the intensive care unit (ICU), intubation, mechanical ventilation, and tracheostomy. Magnetic resonance imaging (MRI) was performed and posterior reversible encephalopathy syndrome was observed without a definitive etiology (Fig. 1). Tests using an anti-nuclear antibody; anti-neutrophil cytoplasmic antibodies; and anti-GM1, GD1b, and GQ1b antibodies were performed, but no positive signals were detected.

The onset of motor weakness in both the upper and lower limbs was noted on the 23rd postoperative day (POD). Physical examination revealed that her motor power was of Medical Research Council (MRC) grade 2/5, with progressive deterioration to grade 1/5 1 day later; this was accompanied with the absence of all deep tendon reflexes in all limbs. The sensory functions of all limbs, as assessed using both pinprick and light-touch tests, were also impaired compared with the cheek. A nerve conduction study (NCS) revealed the presence of severe sensorimotor axonal polyneuropathy, with no response in a routine NCS of the motor and sensory nerves in the median, ulnar nerves, motor NCS of deep peroneal, posterior tibial nerves, and sensory NCS of superficial peroneal and sural nerves. The patient was diagnosed with peripheral polyneuropathy such as GBS or CIP, and was treated with intravenous immunoglobulin administration for 5 days, starting on POD 31, with no definitive effect [4].

Abdominal pain, neurological symptoms, and unexplained severe polyneuropathy raised the suspicion of AIP. For confirming this diagnosis, urine porphobilinogen, delta-aminolevulinic acid, coproporphyrin, porphyrin, and uroporphyrin levels, as well as blood porphyrin concentration, were assessed. A urine color change was also observed after 24 h of exposure to sunlight (Fig. 2). Ten days following the suspicion of AIP, a diagnosis of this condition was confirmed by urine porphobilinogen levels (102.01 mg/day) on POD 40 (Table 1). Two 4 day courses of intravenous hemin were administered after the diagnosis, for proper management of the disease. None of her extremities exhibited a change in power after hemin injection.

A follow-up NCS was performed 2 months after the onset of quadriplegia, without any change in muscle strength or sensory functions. There were no responses in any of the sensory and

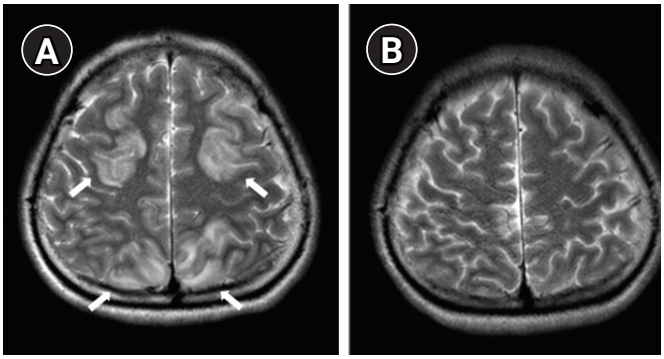

Fig. 1. Brain magnetic resonance imaging performed on POD 10 (A) and POD 21 (B). Axial T2-weighted images (A) revealed signal intensity changes in both the frontal and parietal cortices and in the subcortical white matter(white arrows). POD 21 images (B) revealed complete remission. POD, postoperative day.

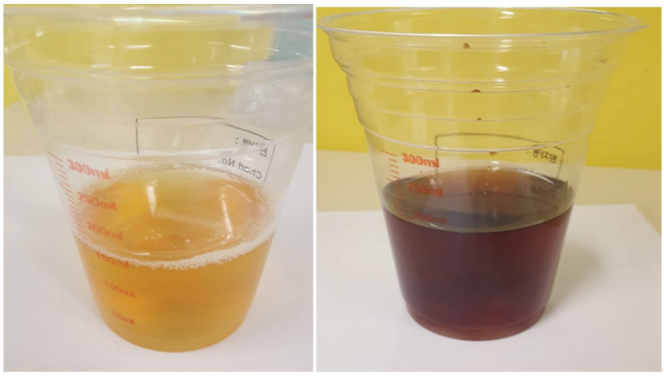

Fig. 2. Urine color change after 24 h of exposure to sunlight, from a straw to a red-brownish color.

motor nerves tested. We observed abnormal spontaneous activities in all muscles tested, and no motor unit action potential. Three months later, the motor power of the proximal upper limbs and lower extremities was improved, to an MRC grade of 3/5. Although there was no improvement in the NCS findings, some regeneration signs, that is, polyphasic motor unit action potential(MUAP) and 2-3 different MUAPs were recruited, were observed in the left biceps brachii, which were absent in the tests performed 3 months previously (Fig. 3). Other muscles tested, which were right first dorsal interossei, rectus femoris, tibialis anterior showed no regeneration signs. These muscles showed no motor unit action potentials in both studies.

Discussion

Acute intermittent porphyria is a type of acute hepatic porphyria that usually presents with gastrointestinal, neurological, and psychiatric symptoms. The common clinical manifestations of this condition include severe abdominal pain accompanied with nausea, vomiting, and ileus; and neurological symptoms, such as seizure, PRES, chronic neuropathic pain, psychosis, and motor-dominant polyneuropathy [5,6]. It is well known that antiepileptic medications, which induce hepatic cytochrome P450 enzymes, act as exacerbating factors of AIP, as do etomidate, ketamine, NSAIDs, and rifampin. Glucose acts as a suppressor of delta-aminolaevulinic acid synthase expression, which participates in the heme biosynthetic pathway. Circumstances such as starvation, which may lead to caloric deficiency, also exacerbate AIP, and can trigger an acute attack [1]. In the present case, phenytoin, valproic acid, topiramate, and etomidate were administered during ICU management, and starvation after laparoscopic operation might also have contributed to the exacerbation of her condition. These drugs and circumstances elicited recurrent acute attacks with neurological symptoms. The clinical manifestations of abdominal pain, seizure, PRES, chronic neuropathic pain, tachycardia, and LFT elevation accompanied the sensorimotor polyneuropathy in our patient. These neurological and gastrointestinal symptoms are extraordinary in general neuropathies.

Porphyric neuropathy mostly presents as a primary motor neuropathy, usually sparing sensory function. Onset distribution is important to rule out other neuropathies. The disease starts in

Table 1. Laboratory Data of Case Patient at POD 31

Laboratory tests	Values	Normal range
Porphobilinogen, 24hour Urine	102.01mg/day	0 - 2.5mg/day
Delta-Aminolevulinic acid, 24 hour Urine	31.0mg/day	1.5– 7.5mg/day
Total porphyrin, blood	Negative	
Total porphyrin, urine	Positive	
Spot urine Coproporphyrin	Positive	
Urine Porphyrin	Positive	
Urine Uroporphyrin	Positive	

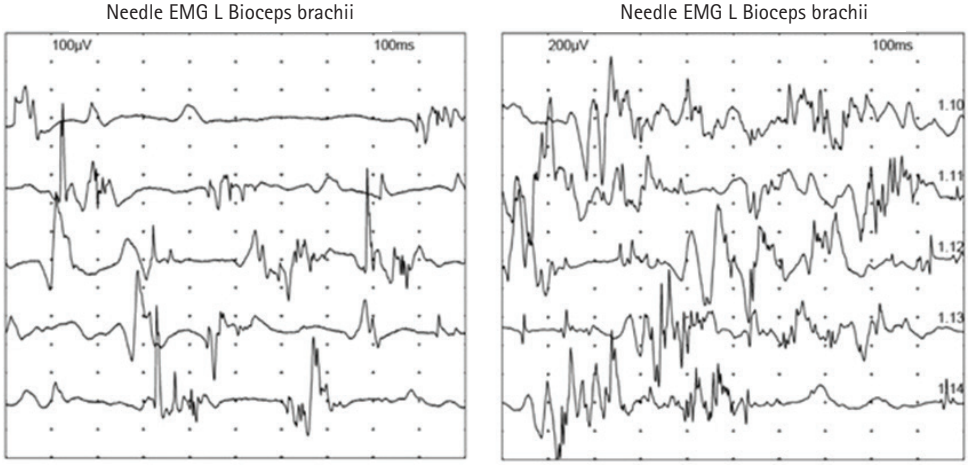

Fig. 3. Polyphasic MUAP of the left biceps brachii observed in follow-up electromyography performed 6 months after the onset of symptoms. MUAP, motor unit action potential.

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the upper extremities in 50% of patients, whereas proximal muscles, rather than distal ones, are involved in 80% of cases [6-8]. But in this case, symmetric sensorimotor involvement was found in early stage. It suggests that we should also consider porphyric neuropathy as one of the diagnosis even in patient with symmetric sensorimotor polyneuropathy. To the best of our knowledge, no clear severity criteria have yet been established. In the present case, we assumed the presence of a severe form of neuropathy in both the upper and lower extremities; moreover, facial and bulbar symptoms were involved with sensory loss. Those symptoms occurred progressively, one by one; thus, the clinical features might be different if early treatment is administered.

Nerve conduction studies and electromyography show no pathognomonic findings for porphyric neuropathy; however, these tests remain important as differential diagnostic tools. In general, porphyric neuropathy shows axonal-type neuropathy in nerve conduction studies, while sensory conduction is often spared. This can be an important clue to differentiate this disease from GBS, which generally presents with a demyelinating type, rather than an axonal one, even though it can also be of axonal type. Some limitations are still existing, such that acute motor axonal neuropathy (AMAN) type is prevalent among Far East Asian, and that severe cases without any action potentials are difficult to specify [9]. Unlike most of other cases, a follow up study was conducted after 3 months, and we could find out some regeneration signs as symptoms improved [3,6-8]. With more studies, it could provide some chances to follow up NCS, EMG as a prognostic tool that might be correlated with clinical features.

Regarding perspective of prognosis, neuropathy improved in a slower manner compared with other symptoms. Moreover, it is related to the magnitude of axonal degeneration. The prevention of recurrent attacks is also an important issue. Weakness and sensory impairment improve over several months, and sometimes incomplete recovery of motor function is observed. Pischik et al. mentioned the severity of abdominal pain, muscle weakness degree, duration of mechanical ventilation, bulbar palsy, consciousness impairment, and hyponatremia as prognostic factors in neuropathy [10]. In our case, mot of factors were severely involved, which helped predict a poor prognosis for this patient. The patient could gait a few steps alone and was able to roller-walker gait with supervision 1 year later.

Acute intermittent porphyria may cause severe peripheral axonal polyneuropathy. Porphyric neuropathy should be considered as a differential diagnosis in patients with an acutely progressing severe polyneuropathy for better prognosis.

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폐암환자에서 신생물딸림 증후군의 초기 증상으로 나타난 보행장애

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Gait Disturbance as an Initial Manifestation of the Paraneoplastic Neurologic Syndrome in a Patient with Lung Cancer

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We report a patient with clinical presentation of Lambert-Eaton myasthenic syndrome (LEMS) and paraneoplastic cerebellar degeneration (PCD) prior to the diagnosis of small cell lung cancer (SCLC). A sixty-year-old man suffered from subacute dizziness, ataxia, dysarthria, ptosis, and proximal weakness of limbs. A repetitive nerve stimulation (RNS) test showed a decremental responses at low-frequency (LF) stimulation and an incremental responses at high-frequency (HF) stimulation, suggesting the pattern of LEMS. There was no evidence of a specific lesion on brain magnetic resonance imaging (MRI), so we concluded his cerebellar ataxia, nystagmus and dysarthria were caused by PCD. We suspected that the PCD was coexistent with LEMS as a paraneoplastic neurological syndrome (PNS) associated with SCLC. Initial screening for cancer was negative; however, SCLC was detected through computed tomography of the thorax at four-months follow-up. Early recognition of PNS and repeated screening tests can lead to a timely diagnosis and earlier initiation of treatment of SCLC.

Keywords: Paraneoplastic syndromes, Nervous system, Lambert-Eaton myasthenic syndrome, Small cell lung cancer

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Introduction

Paraneoplastic neurological syndrome (PNS) can affect the central nervous system (e.g., paraneoplastic cerebellar degeneration [PCD]), and the neuromuscular junction (e.g., Lambert-Eaton myasthenia syndrome [LEMS] and myasthenia gravis [MG]) or the peripheral nervous system (e.g., autoimmune neuropathy) [1]. A small percentage of patients with small cell lung cancer (SCLC) have PNS, of which the most frequent type is LEMS [2]. Here, we report a case where the symptoms of PNS arose before the diagnosis of SCLC, and we emphasize that, there is a need to pay attention to these patients with PNS. Re-

peated cancer screening tests, such as computed tomography (CT) or F-fluorodeoxyglucose- positron emission tomography (FDG-PET) scans, are recommended for a patient with limb weakness due to a neuromuscular junction disorder (especially the LEMS type) [2]. Early diagnosis of cancer allows for earlier initiation of anti-cancer therapy, and thereby improves survival. We experienced a patient who presented initially with LEMS, and cerebellar symptoms and was finally diagnosed with PNS associated with SCLC.

Case report

A sixty-year-old male patient, previously a civil engineer, was admitted to the neurology department because of ataxia, dysarthria, ptosis, dizziness progressive weakness in the proximal limbs, and difficulty walking for two-weeks. On physical examination, he exhibited spontaneous nystagmus, hypo-reflexia, and disturbances in standing and tandem walk test. Orthostatic hypotension was diagnosed by head-up tilt test. Brain magnetic resonance imaging (MRI) revealed no specific abnormal findings. Serology results for antibodies related to non-cancer-associated syndrome (AChR-Ab, anti-GQ1b IgG and IgM, and anti-MuSK Ab) and paraneoplastic antibodies (anti- Hu, anti-Ri, anti-Yo, anti-amphiphysin, anti-CV2, anti-recoverin, anti-SOX1, and anti-PNMA2) were negative. The thyroid function test results were normal. Initial repetitive nerve stimulation (RNS) testing at the neurology department revealed that a compound muscle action potential (CMAP) amplitude in the abductor digiti minimi (ADM) was within the normal reading, and there was a 24% decrement in the response of the ADM on low-frequency (LF) (3Hz) RNS. The CMAP amplitude of the ADM did not increase after brief exercise. These results suggested the pattern of MG (Table 1). A chest CT was performed to look for signs of thymoma, and showed a mass in the anterior mediastinum, suggesting thymoma without evidence of cancer. The patient underwent thymectomy, but a subsequent pathological examination revealed only a thymic cyst. He was treated with a combination of oral prednisolone (60 mg/day for 7days, tapering by 10 mg/day to achieve a 10 mg/day maintenance dose over a total 15-week course) and pyridostigmine (180mg/day maintenance dose) under the impression he had MG.

However, despite the addition of intravenous immunoglobulin therapy for 7 days, the effect of the medical treatment was unclear. His bilateral proximal weakness, truncal ataxia, nystagmus, and gait disturbances continued for several months. The patient was unable to ambulate even with a walker. Therefore, we re-evaluated his neurophysiology study at four months after the initial RNS test. A motor nerve conduction study (NCS) indicated slightly delayed latencies and significant low amplitudes, but his conduction velocities were preserved. His sensory NCS results were normal. On electromyography, proximal and distal limb muscles showed motor units changes with short durations and variable amplitudes. The second RNS test showed a 24% decremental response in the ADM on LF (3Hz) stimulation at rest and the CMAP amplitude of left ADM in post-exercise (immediately) was increased by 260% compared to the baseline. The CMAP of left ADM exhibited an 80.2 % incremental response under high-frequency (HF) stimulation (20 Hz). The 20 Hz HF-RNS test could not be sufficiently performed, but the first 10 potentials demonstrated a tendency to facilitation. These findings were consistent with the pattern of LEMS (Table 1, Fig. 1). Although the patient refused a test for Voltage-gated calcium channels (VGCC) antibody, we considered that he had LEMS based on his clinical symptoms and electrophysiologic test findings.

We conducted chest CT and FDG-PET scans to screen for lung cancer, but no significant lesions were detected (Fig. 2A). Considering the cerebellar signs and ptosis, and the proximal motor weakness with fatigue, we strongly suspected PNS manifesting with PCD, coexisting with LEMS associated with lung cancer. At a four-month follow-up after the initial screening, we performed chest CT and FDG-PET scans again, which showed significant lymphadenopathy in the right paratracheal area (Fig. 2B). The pathology of endobronchial ultrasound bronchoscopy fine needle aspiration finally confirmed a mediastinal lymph node metastasis of SCLC (Fig. 3A, B). Bone scanning showed no evidence of distant metastasis. The patient has since received chemotherapy and radiation therapy. At one year follow-up, abdomen CT, brain

Table 1. Initial RNS Test and Follow-up Test after 4 Months

	Initial test	2nd test (4 months follow-up)
CMAP Amplitude of ADM (mv)		
Rest	12.7	3.1
Immediately post-exercise	13.1	11.2
Increment (%)	3%	260%
3 Hz LF RNS		
Decrement (%)		
resting	-24%	-24%
Immediately post-exercise	-18%	-18%
3 min after exercise	-29%	-23%
20Hz HF RNS		
Increment (%)		80.20%

CMAP, compound motor actioni potential; RNS, repetitive nerve stimulation test; LF, Low-frequency; ADM, abductor digiti minimi; HF, high-frequency.

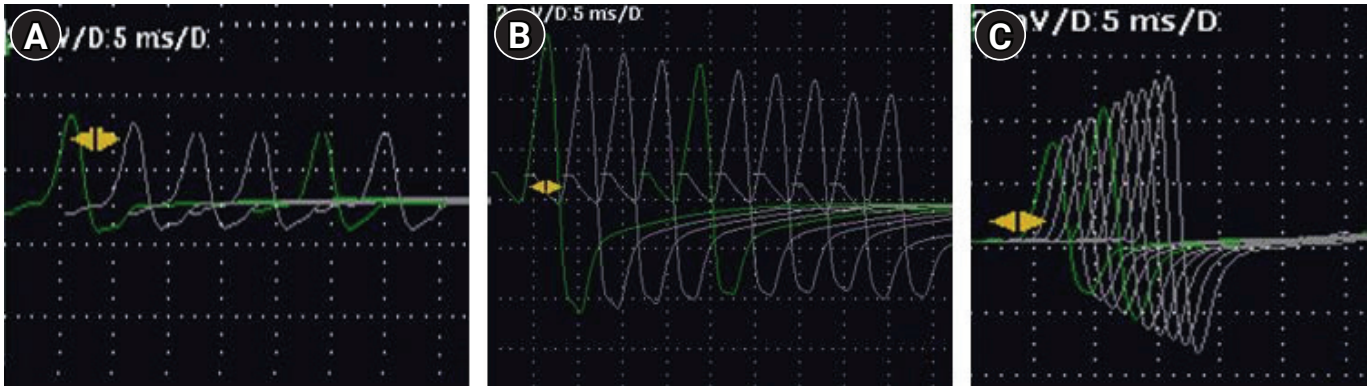

Fig. 1. The second follow up repetitive nerve stimulation (RNS) test in the patient with Lambert-Eaton myasthenic syndrome (LEMS) (A) Baseline: the decreased initial compound muscle amplitude potential (CMAP) amplitude of the left abductor digiti minimi (ADM) and the decremental responses by 24% during 3Hz low-frequency (LF) stimulation (B) immediately post-exercise: Increased the CMAP amplitude of left ADM by 360 % compared to the baseline and the decremental responses by 17.5% during 3Hz LF stimulation C) 20hz high-frequency (HF) RNS test : incremental responses by more than 80% between 1st and 10th responses.

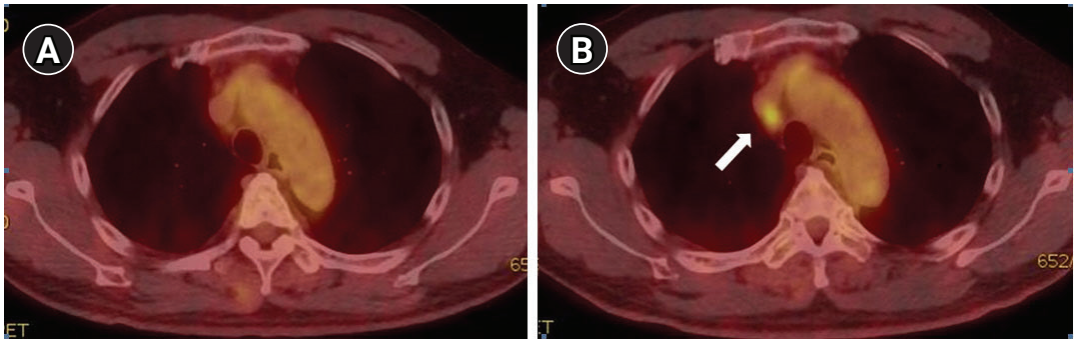

Fig. 2. (A) Initial FDG-PET showed no abnormal lesion. (B) FDG-PET at four-month follow-up evaluation showed newly appeared lymphadenopathy (white arrow) in the right upper paratracheal area.

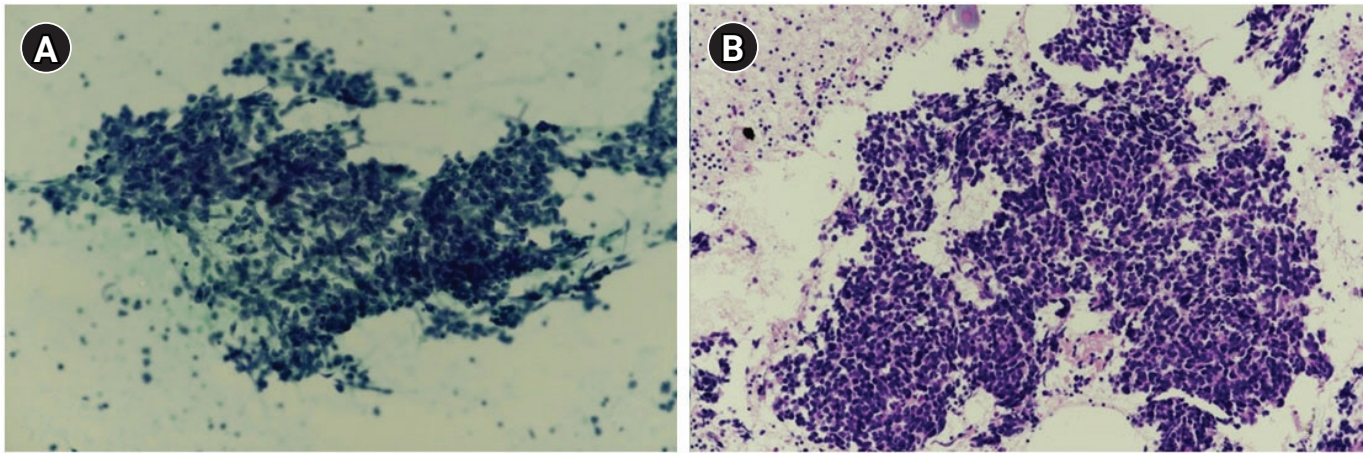

Fig. 3. Cytologic findings of lymph node by endobronchial ultrasound bronchoscopy-fine needle aspiration. (A) Smear (PAP Stain) (B) Cell block (H&E stain). Microscopic findings showed the clusters of round to oval cells with finely dispersed nuclear chromatin, nuclear molding with scanty cytoplasm and apoptotic bodies.

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MRI, and bone scan results showed no distant metastasis. Presently, he can ambulate with a cane after gait and balance training for several months.

Discussion

PNS can manifest in various forms including PCD and as neuromuscular junction disorders (e.g., LEMS and MG) [1]. LEMS and PCD can be present together as PNS in rare cases associated with SCLC.3 In one study, at least 16% of patients with SCLC and PCD had LEMS [4]. Some reports indicated that 41% of patients with PCD and SCLC had P/Q type VGCC antibodies, which are presumed to have a role in the pathogenesis of the cerebellar dysfunction, and 43% of these patients had LEMS [5]. In patients with SCLC, PCD may occur with or without Hu anti-neuronal antibodies (HuAb) [4].

The results of RNS tests are important in diagnosing LEMS. The classical pattern of LEMS shows a low compound muscle action potential (CMAP) amplitude at rest, a 10-15% decremental response in LF-RNS, and a >100% incremental response in HF(10-30Hz)-RNS [5]. In the initial RNS test of our patient, the CMAP amplitude of the left ADM at rest was within normal and did not increase at post-exercise. This pattern of RNS suggested MG, so HF-RNS was not performed at that time. Although the patient was treated under the impression of MG, the possibility of other diseases was suspected, because he was negative for AChR-Ab and anti-MuSK Ab and he showed little response to pyridostigmine. On the second RNS test at 4 months follow-up after the initial RNS, this case showed a low CMAP amplitude (3.1 mV) which increased by 260 % (11.2 mV) after brief exercise. We considered that the CMAP amplitude of ADM at rest gradually decreased as LEMS progressed. If symptoms of LEMS are mild or in an early stage, it is not easy to differentiate it from MG because the CMAP amplitude can be normal, and it does not exhibit the characteristic post-exercise facilitation [6]. Therefore, serial follow-up studies with high rate RNS are important when treating patients with LEMS.

The clinical symptoms of PCD can manifest with ataxia, dizziness, nystagmus, dysarthria, and dysphagia [7]. The criteria of PCD are: no evidence of significant cerebellar atrophy more than the expected on MRI, subacute neurologic symptoms within three months, and a Rankin score of at least 3 (moderate disability; requiring some help) caused by the cerebellar syndrome. Our patient was unable to walk independently due to ataxia, his cerebellar symptoms started within two weeks, and his brain MRI was negative for other causes. Therefore, the cerebellar symptoms in our case were considered to be caused by the PCD.

A repeated screening of radiologic imaging such as chest CT and FDG-PET is important for early recognition of SCLC in LEMS and PCD. The CT scanning of the thorax has revealed a sensitivity of 83% at primary screening and 92% overall in patients with LEMS [8]. FDG-PET can help in the diagnosis of lung cancer in patients with a negative CT thorax result at primary screening [2]. If suspicion of a malignancy remains high, the first repetition of screening should be performed after 3 or 4 months [2]. According to a large cohort study, two years of screening is sufficient for patients with LEMS [8].

A limitation of this report was that we did not perform a VGCC antibody test due to the patient's refusal. Patients with LEMS who are positive for the P/Q-type VGCC antibody showed a prolonged survival [5]. The results of the VGCC antibody test would have been helpful for diagnosis and prognostication in this case.

In conclusion, we experienced a case with progressive weakness of the limbs, ataxia, ptosis, and dysarthria as the initial symptoms of lung cancer without respiratory symptoms. These symptoms were found to be diagnosed with LEMS and PCD. Although initial screening for lung cancer was negative, small cell lung cancer was finally diagnosed through repeated screening. Therefore, early recognition of PNS and repeated screenings would improve the speed of diagnosis and allow for earlier initiation of treatment of the SCLC.

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신경통 근위축증 환자에서 국소 및 전신 스테로이드 병용 치료: 증례 보고

이현성, 김창범, 김은석, 김명옥, 좌경림, 박찬혁, 김창환

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The Successful Treatment of Combined Local and Systemic Steroids in a Patient with Brachial Plexus Neuritis: A Case report

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Brachial plexus neuritis (BPN), also known as Parsonage-Turner syndrome, is an uncommon neurological disorder that can manifest with acute extreme upper arm pain followed by patchy muscle paralysis. There is no evidence supporting any recommended treatments for BPN from randomized trials. Non-randomized studies have supported the effectiveness of early administration of high-dose oral corticosteroids during the painful phase. We present a case of a 41-year-old-female with a history of acute left lateral shoulder pain and shoulder girdle weakness who was diagnosed with acute BPN. To reduce the risk of complications arising from high-dose oral steroid administration, we used an ultrasound-guided brachial plexus blockade combined with low-dose oral steroids. The combined treatment approach successfully restored shoulder function and effectively alleviated pain while avoiding complications associated with systemic steroid administration.

Keywords: Brachial plexus neuritis, Interventional ultrasonography, Steroid

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Introduction

Brachial plexus neuritis (BPN) is a rare peripheral nervous disorder characterized by acute attacks of neuralgia and patchy paresis on upper extremities followed by weakness and atrophy. The pathophysiologic mechanism underlying BPN is presumed to involve localized inflammatory or immune processes affecting the brachial plexus [2]. High-dose oral corticosteroid therapy is the conventional treatment strategy for ameliorating the acute painful phase of BPN [1,3]. Among patients who receive high-dose oral steroid therapy, 20% to 30% experience systemic side effects, including gastrointestinal complaints and mood swings [1,4]. As for other treatments, pain medications (including opioids and gabapentin) or physical therapy (including transcutaneous elec-

trical nerve stimulation) can be applied in the acute and delayed phases [3].

To reduce localized brachial plexus inflammation and to minimize the systemic side effects of steroids, we performed three ultrasound-guided brachial plexus blockades combined with low-dose oral steroid administration. This treatment strategy achieved full restoration of shoulder-girdle function and elimination of residual shoulder pain within 3 months after onset.

Case report

Two weeks after recovering from a common cold, a 41-year-old, right-handed woman experienced sharp and acute pain in her left shoulder and lateral upper arm, followed by a marked loss of left shoulder mobility. The symptoms were so severe that the patient had difficulty completing daily activities. She visited our hospital 2 weeks after the onset of pain. She rated her pain as 7 out of 10 on a numeric rating scale (NRS) ranging from “no pain" (0) to the "most severe pain imaginable” (10). She described a tingling sensation and a dull ache affecting the left shoulder and lateral upper arm. She denied a previous history of neck pain or radiating pain before this event. There was no history of hereditary or metabolic disease. Pain medications, such as opioids, provided no significant pain relief.

Upon physical examination, the patient had prominent left shoulder girdle muscle weakness without atrophy. Shoulder flexion and abduction exhibited Medical Research Council (MRC) power gradings of 2/5, and elbow flexion was graded as 4/5. Her muscle power was normal in the left wrist and hand, as well as the entire right upper extremityLight touch and pin-prick test results were unremarkable. Passive shoulder movement achieved the full range of motion in all directions. Spurling’s test results, which assesses radicular pain, was unremarkable. The Neer and Hawkins-Kennedy tests for identifying rotator cuff pathology were negative.

Her white blood cell count, C-reactive protein and rheumatoid factor results were within normal limits. HbA1c was 5.6. The motor and sensory nerve conduction studyresults were also within normal limits (Table 1). However, electromyography revealed signs of denervation of the left infraspinatus, deltoid, biceps bracii, and left cervical paraspinalis muscles (Table 2). Cervical magnetic resonance imaging (MRI) showed a herniated disc at the C4-5 level with mild protrusion in the left foraminal zone and bulging discs at the C5-6 and C6-7 levels without neural foraminal stenosis or severe cervical root compression.

As a differential diagnosis, cervical radiculopathy was deemed unlikely since the patient presented with sudden pain and severe weakness of the shoulder girdle in a short period without any history of trauma or neck pain. Additionally, the physical examination findings did not support this diagnosis, nor did the mild herniated disc at C5-6—it was not a lesion that could cause severe acute paralysis. Rotator cuff disease was ruled out because of intact range of motion, along with the negative Neer and Hawkins-Kennedy tests and the intact rotator cuff tendon revealed by ultrasound examination. We conducted a brachial plexus MRI with contrast enhancement after suspecting a diagnosis of acute idiopathic BPN. The MRI findings were as follows: 1) mild swelling at the post-ganglionic level of the C5 nerve, 2) swelling and high signal intensity in the left trunks, divisions and the lateral cord and 3) mild T2 high signal intensity at the left supraspinatus and infraspinatus muscles, suggesting denervation changes related to the suprascapular nerve (Fig. 1A, B, and C). Based on the clinical manifestations and radiologic findings, an initial working diagnosis of idopathic BPN was made.

Considering the acute painful phase of the BPN, we initiated oral steroid therapy (30 mg prednisolone for the first 5 days, ta-

Table 1. Motor and Sensory Nerve Conduction Study Results

Motor	Right side				Left side			
	Lat. (ms) Prox./Dis.	Amp. (mV) Prox./Dis.	CV (m/s)	F-M Lat (ms)	Lat. (ms) Prox./Dis.	Amp. (mV)	CV (m/s)	F-M Lat (ms)
Median	2.8/6.4	9.3/8.7	57.9	19.7	3.1/6.6	8.1/8.0	59.8	19.8
Ulnar	2.6/5.3	10.8/10.5	66.7	20.0	2.8/5.8	10.8/10.5	59.6	19.1
Radial	1.8/4.7	5.4/5.4	60.1		1.8/4.8	5.4/5.0	60.3	
Axillary	3.5	3.5			3.2	6.6		
MC	3.6	5.4			3.3	6.1		
Sensory	Lat (peak) (ms)	Amp. (μV)	CV (m/s)		Lat. (ms)	Amp. (μV)	CV (m/s)	
Median	2.9	49.4	48.1		2.8	53.0	50.2	
Ulnar	3.1	53.9	45.6		2.9	64.3	48.6	
Radial	2.5	56.0	55.1		2.7	44.9	52.6	
LABCN	2.0	23.6	50.5		2.0	23.6	58.8	

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Table 2. Needle Electromyography

Muscle	At rest		MUPs			Recruit
	Fibs.	PSW	Poly	Amp	Dur	
Lt. ISP	+++	+++	++			Reduced
BB	++	++	+			Slightly R
Deltoid	+	+	+			Slightly R
FCR	S	S	Normal			Full
ECR	S	S	Normal			Full
EDC	S	S	Normal			Full
FDI	S	S	Normal			Full
Cervical PVM (C5,6)	++	++				

Lat, latency; Amp, amplitude, units; mV in nerve conduction study and uV in sensory conduction study, Prox, proximal stimulation; Dis, distal stimulation; CV, conduction velocity; MUPs, motor unit potentials; Fibs. fibrillation potentials; PSW, positive sharp waves; Poly, polyphasia; Dur, duration; R, recruitment; S, silent; MC, musculocutaneous nerve; LABCN, lateral antebrachial cutaneous nerve; ISP, infraspinatus; BB, biceps brachii; FCR, flexor carpi radialis; ECR, extensor carpi radialis; EDC, extensor digitorum communis; FDI, first dorsal interosseous; PVM, paravertebral muscle.

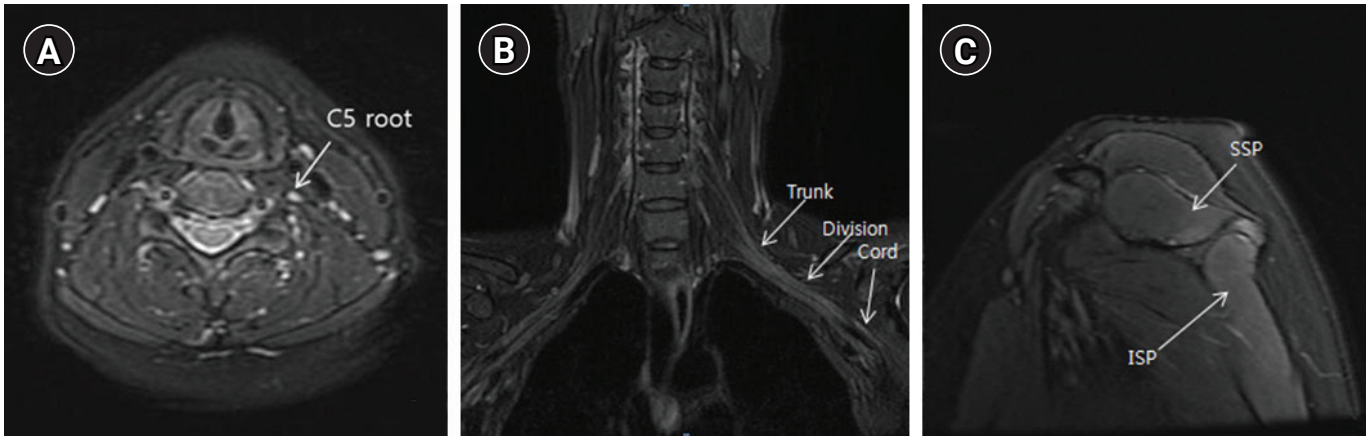

Fig. 1. Brachial plexus MRI with contrast enhancement for a patient with BPN. A) T2 water-excited sagittal image: mild nodular swelling observed in the post-ganglionic level of the C5 nerve root; B) T2 water-excited coronal image:s swelling and high signal intensity observed in the left trunks, divisions, and lateral cord; C) T2 water-excited sagittal image: mild-high signal intensity in the left supraspinatus (SSP) and infraspinatus (ISP), suggesting a denervation change related to the suprascapular nerve.

pered to 20 mg for 3 days, 10 mg for 1 week, and ending with 5 mg as a maintenance dose). The timing of oral prednisone cessation was to be guided by the patient's symptoms. The patient was scheduled for ultrasound-guided brachial plexus blockades to facilitate an oral steroid dosage reduction and provide direct control of local inflammation.

To evaluate the outcomes of the treatment, we assessed the MRC power grades for the three muscle pairs (shoulder flexors, abductors, and elbow flexors), and pain was reassessed using the NRS (Fig. 2). An experienced physician administered an initial injection (dexamethasone 5 mg + lidocaine 25mg) to the C5 brachial root 2 weeks after the onset of symptoms. Two weeks after the first injection, the power in the shoulder flexors and abductors improved from MRC grade 2 to grade 3, and the NRS dropped from 7 to 3. Since the patient was experiencing pain relief and muscle strength improvement, along with greater overall satisfaction, the second injection was administered to the same site with the same drug concentration. Two weeks after the second injection, the pain score went down further, from 3 to 2, and the MRC grades for shoulder abduction and flexion improved from 3 to 4. Since the upper arm pain and shoulder weakness persisted somewhat, a third injection (triamcinolone 20mg + lidocaine 25mg) was finally administered at the trunk level (Fig. 3), and we stopped the oral prednisolone. During the period taking oral steroids, she had no side effects such as nausea, gastrointestinal discomfort, weight gain. Three months after the onset of symptoms, the patient's muscle power was fully restored, and her pain had subsided. Monitoring for side effects was conducted after each injection; however, no side effects of the intervention such as dyspnea, hoarseness, paralysis and local infection, or

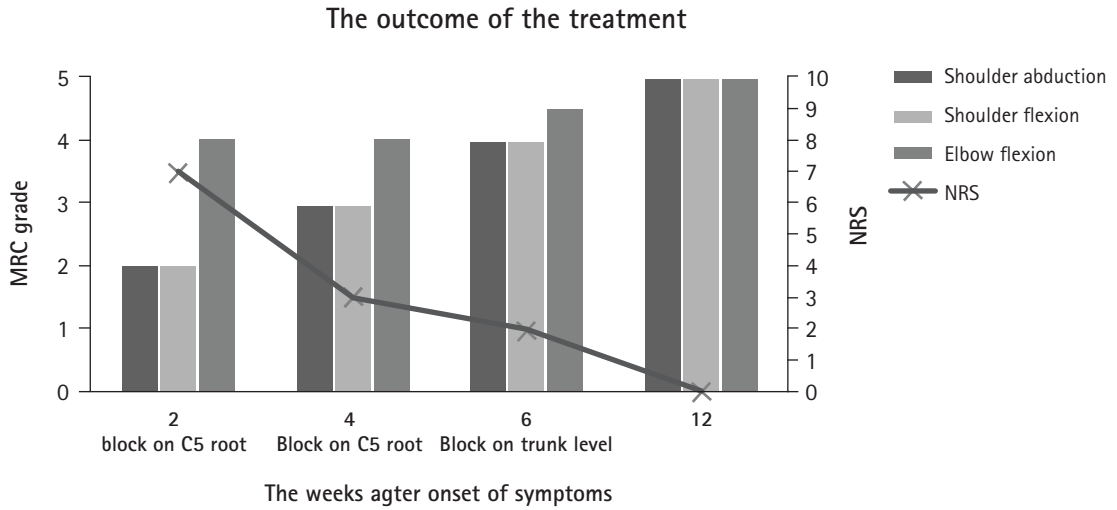

Fig. 2. The outcome of treatement : The graph shows the changes in the shoulder flexors, abductors, and elbow flexors of medical research council (MRC) grades and the numeric rating scale (NRS) for pain following each steroid injection.

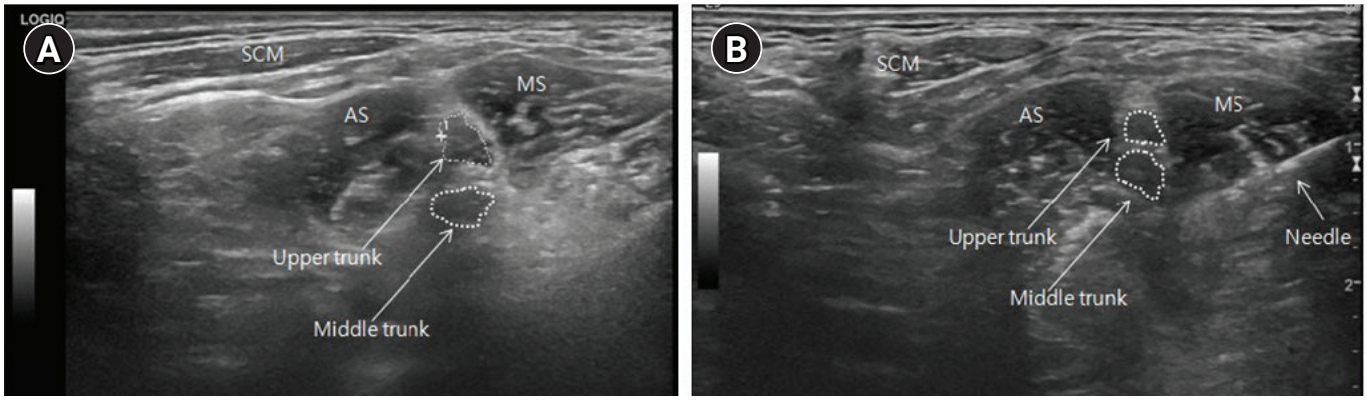

Fig. 3. Ultrasound-guided injection to left brachial plexus at the trunk level with 20mg triamcinolone acetonide and lidocaine 25mg. A) Before the injection; B) During injection. SCM: sternocleidomastoid, AS: anterior scalene, MS: middle scalene.

bleeding were observed. She reported no functional limitations in the performance of daily activities and returned to work.

Discussion

Although the precise pathophysiology of BPN remains unclear, the condition is assumed to have unique or multiple focal inflammatory, immunologic triggers [2,3]. From the pathological perspective, the effectiveness of anti-inflammatory and immunomodulatory drugs, including high-dose oral steroids or intravenous immunoglobulin therapy, has been reported [1,5]. These drugs are administered to reduce the fibrous, stenotic changes of some fascicular hourglass-like constrictions and eventually prevent axonal damage [3].

Van Eijk et al. [1] suggested a regimen "60 mg/day (prednisolone) in the first week, tapered by 10 mg every day during the next 5 days, ending with 5 mg on day 13" for acute-onset BPN (< 1 month) patients.After one year, this regimen provided higher recovery rates than management strategies that omitted corticosteroid therapy (56% vs. 11%) [1]. However, about 20% to 30% of patients are reported to experience systemic side effects, including mood swings and gastrointestinal complaints, due to oral corticosteroid therapy [1,4]. In addition, The short-term use of oral prednisone decreases glucose tolerance in a dose-dependent manner, and more than 6 days of high-dose prednisone (>60mg) raises the possibility of hypothalamic-pituitary-adrenal axis suppression even among healthy adults [6].

If attacks of BPN are presumed to be pathologically immune-mediated, we considered localized immune control through steroid injections. Although no clinical studies on this technique have been reported, one report suggests that local anesthetic and steroids can be used as excellent adjuncts to other treatments [7].

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The intervention is helpful for pain relief while oral steroids are being titrated to an effective level [7]. In one case report, an 83-year-old man with BPN was treated with an interscalene brachial plexus block consisting of bupivacaine and dexamethasone [8]. This approach is not without risks, since the procedure itself can result in some complications, including post-interventional pain, ipsilateral hemidiaphragmatic paresis, Horner's syndrome, and hoarseness [9]. However, the ultrasound-guided approach used by an experienced practitioner would improve the quality of the injection because the approach allows the dynamic visualization of the relevant anatomical structures and needle [9].

In this case, the rationale behind the initial approach to the C5 brachial root was as follows: 1) there was motor paralysis in the C5 myotome and pain in the lateral shoulder area corresponding to the C5 dermatome, 2) the supraspinatus and infraspinatus showed denervation changes on a brachial MRI, and these muscles are innervated by the nerves above the brachial trunk level, and 3) brachial plexus MRI with contrast enhancement also showed a high signal intensity in the post-ganglionic C5 root with mild swelling. The third injection to the brachial plexus in the trunk was finally applied to control the local inflammation shown in the brachial plexus MRI and to prevent progressing axonal damage. These outcomes were desirable in treating our patient.

There are some limitations to consider in association with this report. First, from a case report, we cannot decipher whether the treatment strategy described herein is superior to conventional oral steroid therapy in terms of effecting pain reduction and functional recovery. Further investigations of the efficacy of ultrasound-guided brachial plexus blockades and into the development of a standardized treatment regimen should be undertaken.

In conclusion, althougth, the precise pathophysiology of BPN is unknown, it is thought to be associated with a localized inflammatory immune attack. Additionally, conventional high-dose oral steroid therapy is associated with systemic side effect risk. In our case, the combination of ultrasound-guided steroid injections with low-dose oral steroid therapy successfully relieved pain while avoiding complications associated with systemic steroid administration.Therefore, this approach may be an alternative treatment method to the patients with idiopathic BPN who had a risk of high-dose oral steroid side effect.

Acknowledgements

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CASE REPORT

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내시경적 요추 감압술 후 큰 가성동맥류와 함께 나타난 요근 혈종에 의해 발생한 요추부 신경얼기병증: 증례 보고

이승열, 석현, 김현정, 안준영, 김상현

순천향대학교 부천병원 재활의학과

Psoas Hematoma with a Large Pseudoaneurysm Causing Lumbar Plexopathy after Endoscopic Lumbar Decompression: A Case Report

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We report a case of psoas hematoma with a large pseudoaneurysm causing lumbar plexopathy following endoscopic lumbar decompression. A 34-year-old man had received posterolateral endoscopic lumbar decompression for the treatment of herniated intervertebral disc at L4/5 level. After endoscopic lumbar decompression, his low back pain, groin pain and motor weakness of his left lower extremity were more aggravated. Psoas hematoma with a pseudoaneurysm causing lumbar plexopathy was detected by computed tomography scan and confirmed by electrodiagnostic testing. In order to prevent permanent and catastrophic disability, a proper evaluation for detecting possible complication and immediate management should be needed during the early post-operative phase.

Keywords: Hematoma, Lumbar plexopathy, Pseudoaneurysm, Endoscopic lumbar decompression

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Introduction

Iliacus hematoma is a rare cause of femoral neuropathy [1]. Hematoma in the psoas muscle is also rare cause of lumbar plexopathy [2]. In patient with anticoagulation therapy or hemophilia, spontaneous hematoma on retroperitoneal space can be occurred occasionally [3,4]. However, in patient without bleeding tendency, psoas hematoma causing lumbar plexopathy developed after surgery has been known as a much rarer phenomenon [5,6]. Though there is no consensus for the treatment of lumbar plexopathy from psoas hematoma, delayed diagnosis or treatment can lead to severe and permanent disability.

We report a unique case of psoas hematoma with a large pseudoaneurysm causing complete lumbar plexopathy after postero-

lateral endoscopic lumbar decompression, which was confirmed by computed tomography (CT) and electrodiagnostic testing.

Case report

A 34-year-old man first visited our outpatient clinic with low back pain, severe radiating pain and weakness of left lower extremity and groin pain (Numeric rating scale, NRS 9/10). He had no past history of chronic medical disease, such as hypertension and diabetes. He said his intermittent low back pain had begun about 3 years ago, however, 2 months earlier, radiating pain to left lower extremity was newly developed. Therefore, 3 weeks ago, he had received posterolateral endoscopic lumbar decompression on left lumbar 4/5 level at the local spine clinic under diagnosis of herniated intervertebral disc at lumbar 4/5 level with left radiculopathy. Unexpectedly, his low back pain and radiating pain to left lower extremity was not improved but more aggravated and weakness of left lower extremity was newly developed after endoscopic surgery. About 1 week after endoscopic decompression, proximal weakness of left lower extremity was newly detected.

On physical examination, according to the medical research council (MRC) grading system, the motor strength of left hip flexor, hip extensor, knee flexor, knee extensor, ankle dorsiflexor and ankle plantar flexor was 1, 3, 3, 0, 3 and 4, respectively. There were decreased sensation on L2 ~ L5 dermatome of left lower extremity and a diminished knee jerk reflex. Above all, he complained left lower extremity and groin pain (NRS 9/10). Abdomen and pelvic CT revealed a huge hematoma in left psoas muscle with a large pseudoaneurysm at posterior aspect of the hematoma (Fig. 1). According to CT findings, emergent embolization at proximal and distal portion of pseudoanerysm of left 4th lumbar artery was done by the intervention specialist (Fig. 2).

His lower extremity pain was slightly improved after intervention. On next day after embolization, lumbar spine contrast-enhanced magnetic resonance imaging was done and revealed huge hematoma in the left psoas muscle encasing left L4 nerve root (Fig. 3). Six days after intervention, his pain was more improved, but weakness of his left lower extremity was not improved. From then, comprehensive rehabilitation for restoration of weakness and gait function was begun. Electrodiagnostic testing revealed left lumbar plexopathy with axonal degeneration, mainly femoral and obturator nerves involved, complete in nature (Tables 1, 2). Three months after endoscopic lumbar decompression, his motor weakness was persisted and atrophy of left quadriceps and adductor muscle was detected. Motor weakness of left knee extensor (grade 0/5) and hip flexor (grade 1/5) was not improved. Hypesthesia of his left anteromedial thigh was still existed.

Discussion

Iliopsoas hematoma often occurs in hemophilia patients, anti-

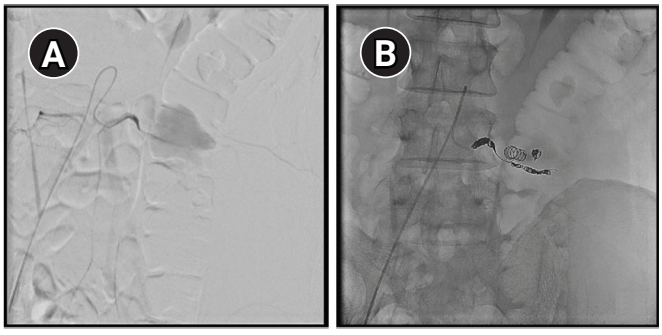

Fig. 2. (A) Pseudoaneurysm at 4th left lumbar artery. (B) 7 microcoil embolization was done at proximal and distal portion of pseudoaneurysm of left 4th lumbar artery.

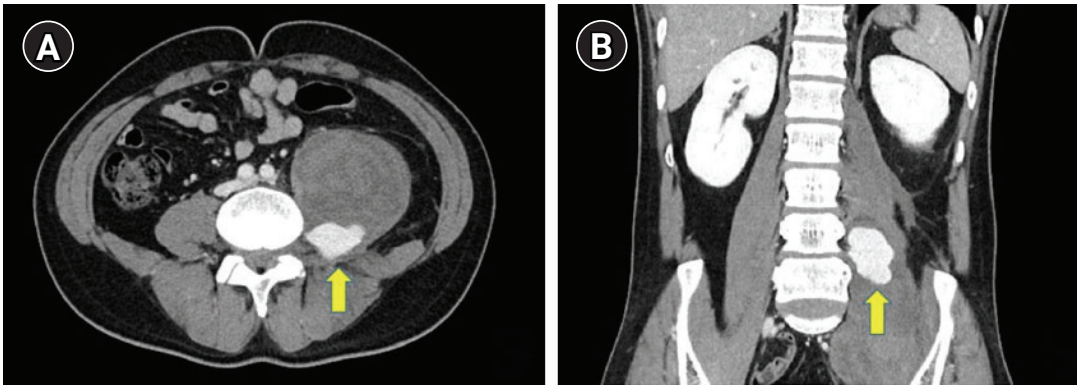

Fig. 1. Abdomen and pelvic CT scan with contrast enhanced. Axial (A) and Coronal view (B). A huge hematoma in left psoas muscle with a large pseudoaneurysm at posterior aspect of the hematoma (arrows).

coagulant recipients, patient with arteriovenous malformations who undergo a low-energy trauma [2]. The mechanism for compression injury of lumbar plexus caused by such retroperitoneal hematoma has already been established [7]. In the case of hematoma at iliacus, the femoral nerve is often compressed. On the other hand, in psoas hematoma, the femoral and obturator nerves can be mainly co-compressed, resulting in diffuse lumbar plexus injury, although it is a rare complication [2]. Previous published three cases of complete lumbar plexus injury secondary to compression by a hematoma were mostly spontaneous hematoma associated with bleeding tendency, such as in patients with hemophilia or other diseases (leukemia, disseminated intravascular coagulation) [2]. And it has also been reported lumbar plexopathy after lumbar plexus block for analgesia in hip surgery in patient treated with enoxaparin [8].

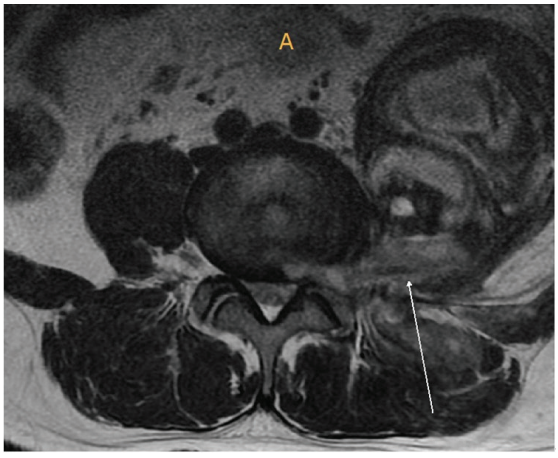

Fig. 3. Lumbar spine contrast-enhanced MRI T2 weighted image on next day after embolization revealed a huge hematoma in the left psoas muscle encasing left L4 nerve root (arrows).

Not the spontaneous hematoma mentioned above, in case of an iatrogenic psoas hematoma without anticoagulant therapy, there were few cases reported. Two cases of iliacus hematoma causing femoral neuropathy associated with hip arthroplasty were previously reported [5,6].Robinson et al. presented a case report of iliopsoas hematoma with femoral neuropathy after spinal decompression [9]. In this case, 8 weeks after a posterior, mid-line, spinal decompression, mild groin pain and a femoral neuropathy developed. After exploration, the compartment was fully decompressed with resolution of the nerve root symptoms within 48 hours. However, after endoscopic lumbar decompression, this is the first case of psoas hematoma presenting a large pseudoaneurysm causing lumbar plexopathy. In this case reported, psoas muscle hematoma with lumbar plexopathy following posterolateral endoscopic lumbar decompression might have been associated with the 4th lumbar artery. Lumbar arteries pass beneath the psoas and develop into posterior and spinal branches [9], in this case, a posterior branch may have been injured, leading to psoas hematoma with a large pseudoaneurysm. Therefore, if there was a groin pain and/or weakness of lower extremity was newly developed or aggravated after endoscopic lumbar decompression, the possibility of psoas hematoma caused by vessel

Table 1. Nerve Conduction Study

Nerve and sites	Lat, ms	Amp, mV	CV, m/s
Motor conduction study			
Lt peroneal nerve (EDB)	4.00	3.2	43.5
Rt peroneal nerve (EDB)	4.25	2.9	47.2
Lt tibial nerve (AH)	3.73	20.7	45.0
Rt tibial nerve (AH)	3.57	28.9	45.7
Lt femoral nerve (RF)	-	-	NT
Rt femoral nerve (RF)	4.71	11.9	NT
Sensory conduction study			
Lt superficial peroneal nerve	2.29	12.5	43.7
Rt superficial peroneal nerve	1.84	18.8	54.3
Lt sural nerve	1.82	22.7	54.9
Rt sural nerve	1.85	25.9	54.1
Lt LFCN	-	-	-
Rt LFCN	2.49	11.0	72.3
Lt saphenous nerve	-	-	-
Rt saphenous nerve	2.54	4.7	55.1

Rt, right; Lt, left; EDB, extensor digitorum brevis; AH, abductor halluces; RF, rectus femoris; Lat, latency; Amp, amplitude; CV, conduction velocity; LFCN, lateral femoral cutaneous nerve; NT, not tested.

Table 2. Needle Electromyography

Muscle	Fib	PSW	Recruitment
Lt Adductor longus	2+	2+	0/5
Lt Adductor magnus	2+	2+	1/5
Lt Tibialis anterior	2+	2+	3/5
Lt Peroneus longus	2+	2+	4/5
Lt EHL	1+	1+	3/5
Lt Gastrocnemius	–	–	5/5
Lt Vastus medialis	2+	2+	0/5
Lt Iliopsoas	2+	2+	0/5
Lt Rectus femoris	3+	3+	0/5
Lt Sartorius	3+	3+	0/5
Lt Gluteus medius	-	-	4/5
Lt Gluteus maximus	-	-	4/5
Lt L3 paraspinalis	-	-	
Lt L4 paraspinalis	2+	2+	
Lt L5 paraspinalis	3+	3+	
Lt S1 paraspinalis	–	–	

Lt, left; Fib, fibrillation potential; PSW, positive sharp wave; EHL, Extensor hallucis longus; NT, not tested.

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injury should be kept in mind. In a case similar to this patient, it would be very significant to investigate the presence of hematoma through early CT scan. As well as, immediate decision of treatment modality should be needed for preventing the other neurologic deficit.

This case has some features that distinguished from other previous ones. First of all, due to the existing L5 radiculopathy with herniated nucleus pulposus at left L4/5 level, it was accompanied by pain and mild weakness in lower extremity even before surgery. Therefore, early diagnosis of hematoma was quite challenging. If muscle strength is more weakened after endoscopic lumbar decompression, it is important to closely observe changes in muscle strength through a delicate physical examination including manual muscle test. It is also important to discriminate whether myotome of weakened muscle was involved with existing radiculopathy or not. In addition, if previous radicular pain characteristics were changed after surgery, for instance, a newly onset groin pain was developed or severity of pain was aggravated, clinicians have to strongly suspect a possibility of newly developed nerve compromised lesion. Consequently, early differential diagnosis can be very essential and challenging.

Electrodiagnostic testing is essential tool when muscle strength is weakened after spine surgery. In this case, a newly developed complete lumbar plexopathy mainly involved femoral and obturator nerves was confirmed accompanied with existing radiculopathy mainly involved L5 nerve roots by electrodiagnostic testing. If the existing lumbar herniated nucleus pulposus was involved with not L5, but L3 or L4 radiculopathy, the results of existing radiculopathy and newly developed plexopathy on the needle electromyography test might appear similar pattern, so the nerve conduction study needs to be closely examined and careful interpretation of electrodiagnostic testing would be necessary.

In conclusion, psoas hematoma should be considered in the differential diagnosis of severe groin pain and lower extremity weakness after endoscopic lumbar decompression. Abdomen and pelvis CT scan could be a useful diagnostic tool in the early period after surgery. If surgical evacuation or embolization of hematoma was not done promptly or delayed, damage to lumbar plexus can lead to prolonged or permanent disability of lower extremity.

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발음성 삼두근 증후군의 근전도 및 초음파 소견: 증례 보고

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Findings of Electrodiagnosis and Ultrasonography in Snapping Triceps Syndrome: A Case Report

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Snapping triceps syndrome is a rare disorder in which the medial head of the triceps muscle moves anteriorly over the medial epicondyle during elbow flexion. Characteristically, visible and palpable snapping can be observed during elbow flexion. Several hypotheses have been proposed about its cause, but the etiology is still unclear. The diagnosis of snapping triceps syndrome is based on clinical symptoms and imaging studies. Early diagnosis is important to prevent symptom worsening and unnecessary surgical treatment. Snapping triceps syndrome is often accompanied by ulnar neuropathy. However, few electrodiagnostic findings have been reported for snapping triceps syndrome. Therefore, we report the ultrasonographic and electrodiagnostic findings in a patient with snapping triceps syndrome.

Keywords: Snapping triceps syndrome, Ulnar neuropathies, Electrodiagnosis

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Introduction

Snapping triceps syndrome was first reported by Rolfsen in 1970 [1]. It is a relatively rare condition in which a part of the medial head of the triceps dislocates beyond the medial epicondyle during elbow flexion, and it is often accompanied by ulnar nerve dislocation [2,3]. Isolated ulnar nerve dislocation is relatively common and is found in approximately 16% of healthy subjects [4]. However, snapping triceps syndrome is an uncommon disease and no study has reported its prevalence.

Patients with snapping triceps syndrome may complain of symptoms of medial elbow pain, paresthesia in the ulnar nerve distribution, and visible, audible, or palpable snapping during elbow flexion. Such symptoms may be unilateral or bilateral and become worse after excessive activity using elbow joints. However, the syndrome can be asymptomatic. If the diagnosis is clinically unclear, magnetic resonance imaging (MRI) can be performed. However, because MRI is a static imaging modality, it should be performed on the elbow in both fully flexed and extended positions so that the ulnar nerve and medial head of the triceps muscle are dislocated.5 Therefore, dynamic ultrasonography, which can visualize movements during elbow joint motion, is considered the imaging modality of choice [5].

Previous studies on snapping triceps syndrome have reported

ultrasonographic findings and surgical treatment [3,5], but electrodiagnostic findings have rarely been reported. Therefore, we report the findings of a nerve conduction study and electromyography in a patient with snapping triceps syndrome confirmed by ultrasonography.

Case report

A 55-year-old man who had been training with weights for about 10 years visited the local orthopedic clinic with a chief complaint of left shoulder pain and a limitation in elbow extension when the left shoulder was flexed greater than 90°. After conservative management, the pain was alleviated but the limitation of the elbow extension persisted, and he was referred to a tertiary-care hospital. He had a medical history of 5 years of hypertension, 5 years of dyslipidemia, and early gastric cancer treated with an endoscopic mucosal resection 2 years ago. And he had a left elbow fracture when he was young, and the left elbow showed an increased cubitus valgus compared with the right side. When his left medial elbow was physically contacted, he felt a tingling sensation in his left fourth finger. No abnormal findings were detected on upper limb radiographs and brachial plexus MRI, but left foraminal stenosis was found between the sixth and seventh cervical disks (Fig. 1). On physical examination, palpable and visible snapping was observed when he flexed his left elbow over 90° with shoulder flexion over 90°. There was no tenderness around the medial epicondyle. Therefore, we performed dynamic ultrasonography to determine which structure was snapping during elbow flexion motion. Ultrasonography showed that the medial head of the triceps dislocated anteriorly over the medial epicondyle with the ulnar nerve when the elbow flexion was more than 90-100° but there was no enlargement of the left ulnar nerve compared with the right ulnar nerve (Fig. 2). The findings could be diagnosed as snapping triceps syndrome.

Motor and sensory nerve conduction studies (NCS) of the bilateral median, ulnar and radial nerves showed normal findings (Table 1). In particular, motor and sensory NCS of ulnar nerves were conducted to identify ulnar neuropathy because the patient felt a tingling sensation in the left fourth finger and the dislocated medial head of the triceps can compress the ulnar nerve during elbow flexion. However, no ulnar neuropathy findings were obtained, such as conduction block or focal slowing. Needle electromyography (EMG) was performed for the left paracervical and upper extremity muscles (Table 2). Regarding needle EMG, all the examined muscles, including the ulnar nerve innervated muscles, showed no abnormal findings, except that the triceps muscle showed increased insertional activity and denervation potentials such as positive sharp waves and fibrillation potentials.

Because electrodiagnosis showed no evidence of ulnar neuropathy, he underwent selective transforaminal epidural block (STEB) as a diagnostic and therapeutic option for radiculopathy caused by left foraminal stenosis between the sixth and seventh cervical disks. There was no immediate pain relief after STEB, so it was unclear whether radiculopathy was the cause. A nonsteroidal anti-inflammatory agent and pregabalin were prescribed for him to relieve pain. After 1 month of treatment, the tingling sensation in the left fourth finger and shoulder pain were improved, but the elbow extension limitation and snapping symptoms persisted.

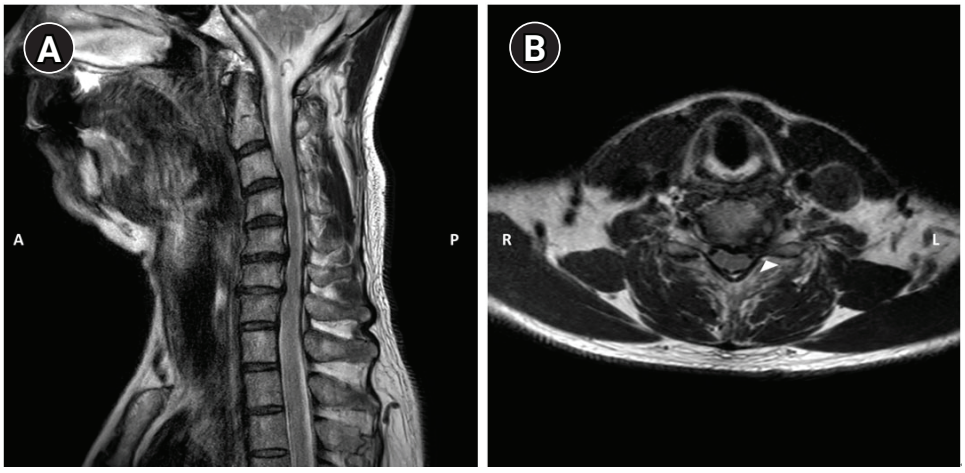

Fig. 1. An mDIXON T2-weighted image (A) sagittal view. (B) Axial view. Left foraminal stenosis between the sixth and seventh cervical disks (arrowhead). A: anterior, P: posterior, R: right, L: left.

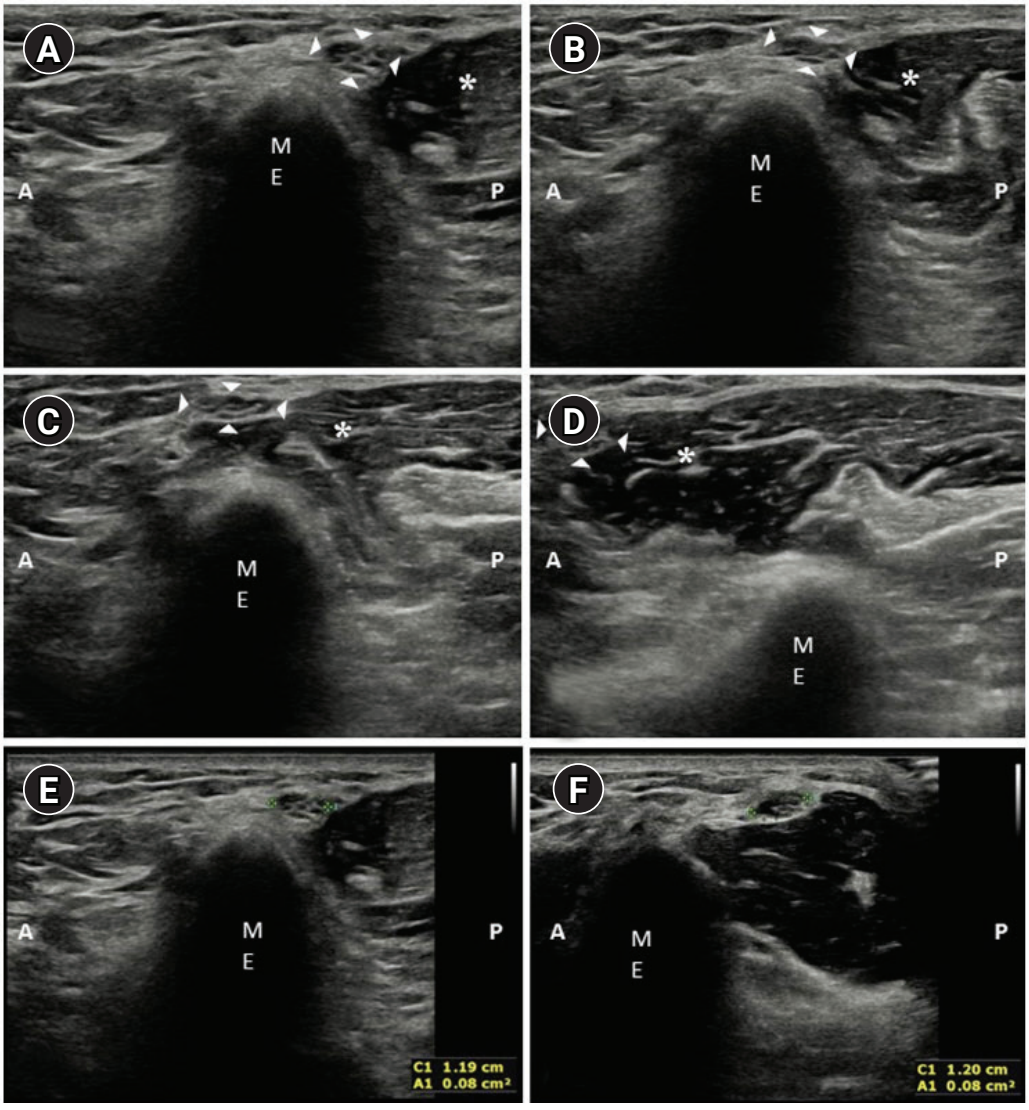

Fig. 2. Left elbow in (A) 0° flexion, (B) 45° flexion, (C) 90° flexion, and (D) 120° flexion. The medial head of the triceps (asterisk) dislocated anteriorly to the medial epicondyle with the ulnar nerve (arrowhead) when the elbow flexion was over 90-100°. Cross-sectional area (CSA) of the ulnar nerve on the (E) left elbow (0.08 cm^2) and (F) right elbow (0.08 cm2). An ultrasound transducer was applied to a virtual line connecting the medial epicondyle and olecranon in the elbow extension position. ME: medial epicondyle, A: anterior, P: posterior.

Discussion

Snapping triceps syndrome is a rarely diagnosed disease. According to the first case of snapping triceps syndrome reported by Rolfsen [1], only anterior ulnar nerve transposition was performed to relieve the ulnar nerve symptoms. However, the snapping symptoms continued, resulting in further resection of the medial triceps muscle. Therefore, it is important to diagnose snapping triceps syndrome correctly to prevent additional operations.

Several studies have attempted to fully explain this syndrome, but it is thought to be due to structural problems of the muscle belly, tendon or fascia of the medial triceps [3]. The causes can be divided into congenital and acquired. Congenital causes may include incorrect insertion of the triceps into the olecranon, accessory triceps tendon and fourth head of the triceps muscle. Acquired causes may include hypertrophy of the medial head of the triceps due to excessive muscle use or cubitus varus deformity that occurs after trauma [3,6]. In this case, long-term weight training may be the cause of snapping triceps syndrome; however, further confirmation is required.

Prior studies on snapping triceps syndrome have mostly been conducted on its diagnostic imaging and surgical treatment, but only few have been conducted on electrodiagnostic testing. Boon

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Table 1. Motor and Sensory Nerve Conduction Studies

Side	Motor nerve	Stimulation	Recording	Latency (msec)	Amplitude (mV)	Distance (cm)	NCV
Right	Ulnar	Wrist	ADM	2.5	18.1		
		BE	ADM	6.1	17.7	23.5	67.4
		AE	ADM	7.5	16.9	10	68.6
		Axilla	ADM	9.0	16.4	11.5	69.8
Left	Ulnar	Wrist	ADM	2.2	17.9		
		BE	ADM	5.8	17.3	23.5	64.0
		AE	ADM	7.2	16.8	10	68.6
		Axilla	ADM	8.6	16.2	10	73.1
Right	Median	Wrist	APB	2.7	16.8		
		Elbow	APB	6.6	16.3	24.5	62.8
Left	Median	Wrist	APB	3.1	13.4		
		Elbow	APB	7.3	12.2	24.6	57.9
Right	Radial	Forearm	EIP	1.4	10.6		
		Elbow	EIP	3.5	9.5	14	68.3
Left	Radial	Forearm	EIP	1.4	10.6		
		Elbow	EIP	3.3	9.8	14	73.7
Side	**Sensory nerve**	**Stimulation**	**Recording**	**Onset latency (msec)**	**Amplitude (uV)**		
Right	Ulnar	Wrist	V digit	2.2	43.6		
Left	Ulnar	Wrist	V digit	2.6	40.7		
Right	Median	Wrist	II digit	2.8	33.8		
Left	Median	Wrist	II digit	2.6	33		
Right	Radial	Forearm	1st web space	1.5	38.9		
Left	Radial	Forearm	1st web space	1.2	48		

NCV: nerve conduction velocity, BE: below elbow, AE: above elbow, ADM: abductor digiti minimi, APB: abductor pollicis brevis, EIP: extensor indicis proprius

Table 2. Needle Electromyography

Side	Muscle	Insertional activity	Spontaneous activity	Motor unit action potentials			IP
				Polyphasia	Amplitude	Duration	
Left	Paracervicalis	N	-				F
	Deltiod	N	-				F
	Biceps	N	-				F
	Tricpes	I	F&P(+++)				F
	EDC	N	-				F
	FCU	N	-				F
	FDI	N	-				F
	APB	N	-				F
	ADM	N	-				F

IP, interference pattern; N, normal; I, increase; F, full; F&P, fibrillation potentials & positive sharp waves; EDC, extensor digitorum communis; FCU, flexor carpi ulnariss; FDI, first doral interosseous; APB, abductor pollicis brevis; ADM, abductor digiti minimi.

et al. [2] compared the firing patterns of the medial, lateral, and long head of the triceps muscles between patient and control groups to verify the hypothesis that snapping triceps syndrome is caused by abnormal firing patterns in each muscle segment. However, studies have shown no significant difference between the patient and control groups. Kang et al. [7] performed ultrasonographic and electrophysiologic evaluations of ulnar nerve instability and the triceps medial head position during elbow flexion in healthy subjects. They reported that the instability of the ulnar nerve increased with more elbow flexion, which was associated with triceps snapping. However, their study was conducted only in healthy subjects; the subjects did not have real

snapping symptoms or paresthesia in the ulnar nerve innervation area.

Snapping triceps syndrome accompanied by ulnar neuropathy was reported previously [3,8]. However, no abnormal findings of ulnar NCS were found in this case. The ulnar nerve was dislocated beyond the medial epicondyle tip in this case, but the snapped medial head of triceps did not compress it and the sonographic morphology of the ulnar nerve did not change. This might be the cause of normal NCS of the ulnar nerve in this case. Nevertheless, friction between the ulnar nerve and medial epicondyle caused by repeated elbow flexion and extension might have resulted in ulnar nerve irritation and associated paresthesia symptoms.

In the needle EMG of the patient, denervation potential was observed only in the left triceps muscle, not in the ulnar innervated muscles. To the best of our knowledge, there is no report concerning needle EMG findings in snapping triceps muscle, and this is the first report of denervation potentials in snapping triceps muscle. This denervation may be caused by radiculopathy due to left foraminal stenosis between the sixth and seventh cervical vertebrae. However, this explanation is unlikely because no other C7-innervated muscles and paracervical muscles showed denervation potentials. Another explanation may be direct muscle injury caused by repeated snapping between the medial epicondyle and medial triceps head. Denervation potentials from direct muscle injuries have been reported in several previous studies [9], including repetitive trauma in the foot intrinsic muscle of normal elderly people [10].

Treatment of snapping triceps syndrome is conservative, and consists of avoiding the activity of repeating elbow flexion as much as possible, using non-steroidal anti-inflammatory agents, and attempting corticosteroid injection around the triceps or ulnar nerve [3]. If the symptoms do not improve despite 3-6 months of non-operative management, surgical treatment could be considered [3]. Some surgical treatments have been reported and include excision of the medial head of the triceps muscle, transposition of the triceps, and medial epicondylectomy [3,8], but no consensus exists.

In this patient, abnormal findings were not observed in the NCS of the ulnar nerve and symptoms were alleviated by one month of oral medications: a nonsteroidal anti-inflammatory agent and pregabalin. And we observed denervation potentials in the triceps muscle that may be caused by friction between the medial head and medial epicondyle, suggesting that the friction caused by snapping may lead to significant muscle trauma. However, further studies are needed to confirm whether isolated denervation potential due to muscle injury occurs in other patients with snapping triceps muscle.

To the best of our knowledge, this is the first report of a patient with snapping triceps syndrome confirmed by ultrasonography and accompanied by denervation potentials in the dislocated triceps muscle.

Acknowledgements

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CASE REPORT

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천측두동맥-중대뇌동맥 우회로 형성술 시행 중 유발전위의 변화 양상

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Changes in Evoked Potentials during Superficial Temporal Artery-Middle Cerebral Artery Bypass Surgery: A Case Series Report

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Superficial temporal artery-middle cerebral artery (STA-MCA) bypass is a surgical treatment for ischemic stroke caused by large artery occlusion. Intraoperative neuropsychological monitoring (IONM) is useful for ensuring cerebral perfusion. Herein, we report 5 cases of patients with ischemic stroke who underwent STA-MCA bypass with IONM. We investigated whether the improved cerebral perfusion was reflected in real time on the evoked potentials (EP) of IONM. To measure changes in cerebral perfusion, all patients underwent pre- and post-operative perfusion-weighted imaging (PWI). Additionally, functional assessments were performed before and after the surgery. We found that the motor evoked potential (MEP) amplitudes showed a consistent increase at the end of surgery. The somatosensory evoked potential (SSEP) latencies decreased consistently, but this change was not large. SSEP amplitudes showed no consistent change. Most of the PWI findings and functional assessments of our patients also improved after the surgery. After bypass surgery, MEP showed more reliable and consistent changes than SSEP.

Keywords: Cerebral Revascularization, Cerebral blood flow, Intraoperative neurophysiological monitoring

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Introduction

Revascularization treatment methods for ischemic stroke include intravenous thrombolysis, endovascular recanalization, and mechanical thrombectomy [1]. In patients with cerebral infarction caused by occlusion of large arteries such as the middle cerebral artery (MCA) or the internal carotid artery (ICA), superficial temporal artery to middle cerebral artery (STA-MCA) bypass surgery may be considered as a treatment method. This is considered particularly when endovascular recanalization therapy is ineffective and the compensatory blood circulation corresponding to the reduced cerebral blood perfusion does not occur adequately [2].

Monitoring cerebral blood flow is very important during STA-MCA bypass surgery. Sonographic flowmetry and indocyanine green angiography are two methods currently being used to assess blood flow in real time. These tests employ flow-oriented monitoring, and are used in the operating room because they can provide immediate and visual feedback [3]. Pre- and post-operative perfusion weighted magnetic resonance imaging (PWI) is another method that can be used to check the cerebral perfusion and the improved blood flow quantitatively. PWI images are also helpful in checking the pattern of blood flow changes according to the different brain regions [4].

Intraoperative neurophysiological monitoring (IONM) has been widely applied as an important diagnostic tool during cerebrovascular surgeries. With IONM, neurologic integrity and functional status as well as overall perfusion status can be monitored during the entire operation. Patient safety can be ensured by real-time detection of abnormal signs due to insufficient cerebral perfusion or a decrease in local blood. Numerous studies have demonstrated that the application IONM during cerebrovascular surgeries could prevent neurological damage [5]. However, only a few reports of the application of IONM during STA-MCA bypass surgery currently exist. Moreover, no report has been presented as to whether the improvement in blood flow seen during direct bypass surgery could be reflected as a change in the evoked potential (EP) on IONM.

Case report

This is a case series report of 5 patients who underwent the STA-MCA bypass surgery with IONM. This study protocol was reviewed and approved by the Institutional Review Board (approval No. PSSH0475-201911-HR-004-01).

For each patient, we observed the changes in EP on IONM that occurred between baseline and the end of the operation. The amplitude of the motor evoked potential (MEP) and the latency and amplitude of the somatosensory evoked potential (SSEP) were measured on the lesion side. MEP recorded from the abductor pollicis brevis muscle (APB-MEP) and tibialis anterior muscle (TA-MEP) were measured. For SSEP, median SSEP recorded from the median nerve at the wrist and tibial SSEP recorded from the posterior tibial nerve at the ankle level were measured. Total intravenous anesthesia regimen (propofol, 3-5 μg/mL and remifentanil, 3-5 ng/mL) was applied during the surgery. A single bolus of neuromuscular blocking (NMB) agent (rocuronium, 0.4-0.5 mg/kg) was used before intubation. The demographic factors of patients are summarized in Table 1.

We observed the PWI findings before and 1 week after surgery. The region of interest was positioned at the centrum semiovale (CS) and the subcortical level of primary motor and sensory cortex area (SPMS). For functional assessment, we measured the modified Rankin scale (mRS) and the modified Barthel index (MBI) at the same time as the PWI. The results of EP, PWI, and functional assessments are summarized in Table 2.

1) Case 1

An 80-year-old female patient who had left MCA occlusion underwent STA-MCA bypass surgery 2 months after stroke onset. She had hypertension, which was a vascular risk factor. Median SSEP showed a decrease in N20 latency and an increase in amplitude, but the degree of change was 2.37% and 7.16%, respectively. For tibial SSEP, P37 latency decreased by 2.3%; however, at the

Table 1. Characteristics of Patients

Case	Age (years)	Sex	Occlusion	Lesional side	Onset to OP	Vascular risk factors
1	80	female	MCA	Left	2 months	HTN
2	57	female	MCA	Right	2 weeks	HTN, DM
3	45	male	MCA	Left	2 weeks	DM
4	42	male	ICA	Left	6 months	HTN, SM
5	52	female	MCA	Right	6 months	HTN, DM, CAD

OP, operation; MCA, middle cerebral artery; ICA, internal carotid artery; HTN, hypertension; DM, diabetes mellitus; SM, smoking; CAD, coronary artery disease.

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Table 2. Findings of IONM, PWI, and Functional Assessments

		Somatosensory evoked potential*				Motor evoked potential*		PWI†		Clinical data†	
		Median nerve		Tibial nerve		APB	TA	SPMS-MTT (s)	CS-MTT (s)	mRS	MBI
		N20 (ms)	Amplitude (uV)	P37 (ms)	Amplitude (uV)	Amplitude (uV)	Amplitude (uV)				
Case 1	Pre	20.65	11.73	43.5	0.87	2670	1290	13.9	12.4	2	88
	Post	20.16	12.57	42.5	0.6	2700	1710	11.9	10.3	2	88
	Δ	-0.0237	0.0716	-0.023	-0.3103	0.0112	0.3256				
Case 2	Pre	20.88	0.76	37	1.53	617.86	764.69	11	11.7	1	96
	Post	20.53	0.74	36.75	1.1	1110	798.66	10.1	9.9	0	100
	Δ	-0.0168	-0.0263	-0.0068	-0.281	0.7965	0.0444				
Case 3	Pre	NR	NR	NR	NR	NR	NR	10.1	10.7	5	5
	Post	NR	NR	66.5	0.19	NR	NR	10.5	9.1	4	37
	Δ			Appeared	Appeared						
Case 4	Pre	18.9	0.67	37.75	1.31	559.49	1530	13.5	12	3	70
	Post	18.55	0.75	35.26	1.26	2590	1690	8.1	8.6	3	70
	Δ	-0.0185	0.1194	-0.066	-0.0382	3.6292	0.1046				
Case 5	Pre	17.97	1.25	36.25	1.27	NR	674.45	6.9	6.9	3	70
	Post	17.73	3.04	33.5	1.34	318.94	1660	8.2	7.1	2	88
	Δ	-0.0134	1.432	-0.0759	0.0551	Appeared	1.4613				

IONM, intraoperative neurophysiological monitoring; PWI, perfusion weighted imaging; APB, abductor pollicis brevis; TA, tibialis anterior; SPMS, subcortical level of primary motor and sensory cortex area; CS, centrum semiovale; MTT, mean transit time; mRS, modified Rankin scale; MBI, modified Barthel index; Δ, delta; NR, no response.
*Values were obtained from the baseline wave and wave at the end of surgery.
†They were measured before and 1 week after surgery.

end of the surgery, the amplitude decreased by 31.03%. The amplitude of APB-MEP increased by 1.12%, which was a minute degree of change. The amplitude of TA-MEP increased by 32.56%. PWI revealed that the mean transit time (MTT) at the SPMS and CS had shortened. Functional assessments showed no changes.

2) Case 2

A 57-year-old female patient who had right MCA occlusion underwent STA-MCA bypass surgery 2 weeks after stroke onset. She had hypertension and diabetes. Both the median and tibial SSEP showed decreases in latency, but the degree of change was 1.68% and 0.68%, respectively. SSEP amplitudes decreased at the end of the surgery. An increase in amplitude was observed in all the measured MEPs. The amplitude of APB-MEP increased by 79.65% indicating a large amount of change. Meanwhile, the amplitude of TA-MEP increased by only 4.44%, which was a slight increase. In the PWI findings, both SPMS and CS showed the most reduced MTT at 1 week after surgery. Even before the surgery, the patient had an MBI score of 96 and had not experienced a significant decline in daily activities. Thus, the patient was able to resume normal function following the surgery.

3) Case 3

A 45-year-old male patient who had left MCA occlusion underwent STA-MCA bypass surgery 2 weeks from the stroke onset. He had diabetes. Both SSEP and MEP were not evoked at the baseline. At the end of the surgery, a small waveform of tibial SSEP was evoked; however, the delay in P37 latency was still severe and the amplitude was small. Median SSEP and all MEPs were not evoked at the end of surgery. PWI conducted at 1 week after surgery revealed improved MTT at the CS (Fig. 1). However, at the SPMS, PWI showed no clear improvement in perfu-

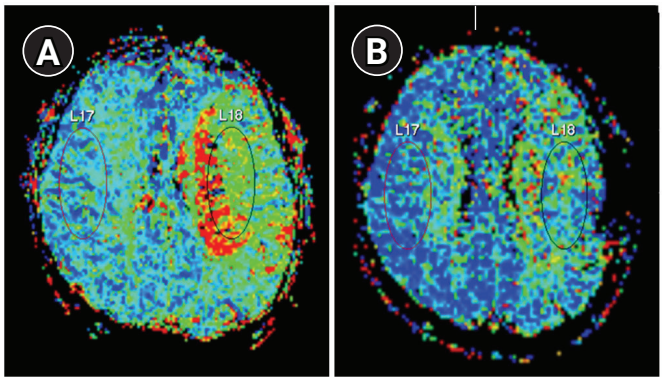

Fig. 1. Perfusion weighted magnetic resonance imaging (PWI) of case 3. Preoperative PWI showed prolonged mean transit time (MTT) (A). As can be seen, PWI at 1 week after surgery showed that MTT decreased following STA-MCA bypass (B). Region of interest was positioned at the centrum semiovale level.

sion. The patient scored a 5 and a 4 on the mRS before 1 week after surgery, respectively. His MBI before surgery was 5. Thereafter, the patient showed an improvement in the functional status by scoring an MBI of 37 at 1 week after surgery.

4) Case 4

A 42-year-old male patient who had left ICA occlusion underwent STA-MCA bypass surgery 6 months from the stroke onset. He had hypertension and was a smoker. Median SSEP showed a decrease in N20 latency by 1.85% and an increase in amplitude by 11.94%. For tibial SSEP, P37 latency decreased by 6.60%. In contrast, the amplitude decreased, but the degree of change was small. The amplitude of APB-MEP increased by 362.92%, and TA-MEP increased by 10.46%. On postoperative PWI, MTT at the CS and the SPMS showed an obvious reduction. Functional assessments showed that his mRS score and MBI before surgery were 3 and 70, respectively, and there were no changes in these scores 1 week after surgery.

5) Case 5

A 52-year-old female patient who had right MCA occlusion underwent STA-MCA bypass surgery 6 months from the stroke onset. She had hypertension, diabetes, and coronary artery disease. Both median SSEP and tibial SSEP showed decreases in their latencies and increases in amplitude. Median SSEP showed a considerable change with an increase in amplitude by 143.20%. APB-MEP was not evoked at the baseline. At the end of the surgery, however, we observed a waveform representing a significant change (Fig. 2). TA-MEP also showed a significant change with an increase of 146.13% in amplitude at the end of surgery. The patient scored 3 on the mRS and 70 on the MBI before surgery. At 1 week after surgery, the patient's functional status improved (mRS score, 2 and MBI, 88). Although findings from both EP and functional assessments indicated improvements, PWI showed an increase in MTTs.

Discussion

With IONM, functional integrity of motor and sensory pathways can be detected in real time during surgery. IONM is distinguished from other real-time blood flow monitoring methods or PWI as it has the advantage of reflecting the patient's neurological and functional states [3]. In this study, we investigated whether the improved cerebral perfusion after vascular anastomosis during STA-MCA bypass surgery was reflected in real time on the EP on IONM.

MEP and SSEP are key IONM modalities that are mainly used for cerebrovascular surgeries. MEP is sensitive as it reflects the functional state of motor pathways and rapidly reacts to neurological deterioration; however, it has the disadvantage of having a high variability [6]. Contrastingly, SSEP reflects the functional state of sensory pathways and is also closely related to other factors such as blood pressure and body temperature as well as overall cerebral blood flow. SSEP generally has a higher specificity and relatively lower sensitivity than MEP. SSEP response to intraoperative events has also been shown to be slower than that of MEP [7]. Due to these characteristics, in case of subcortical ischemia, MEP has a rapid response, but SSEP can display false negatives [8]. On the other hand, according to the anatomical features of the brain motor pathway, lower extremity SSEP may react more sensitively to ischemia than MEP in the case of anterior cerebral artery or an anterior choroidal artery procedures [9]. In addition, since MCA supplies both primary motor and sensory cortices, changes in SSEP can indirectly reflect motor deficit [5]. In most cases, MEP and SSEP are monitored simultaneously. Such multimodal EP monitoring gives the advantage of providing complementary information to physiatrists and surgeons.

IONM's role as a preventive measure for ensuring patient safe-

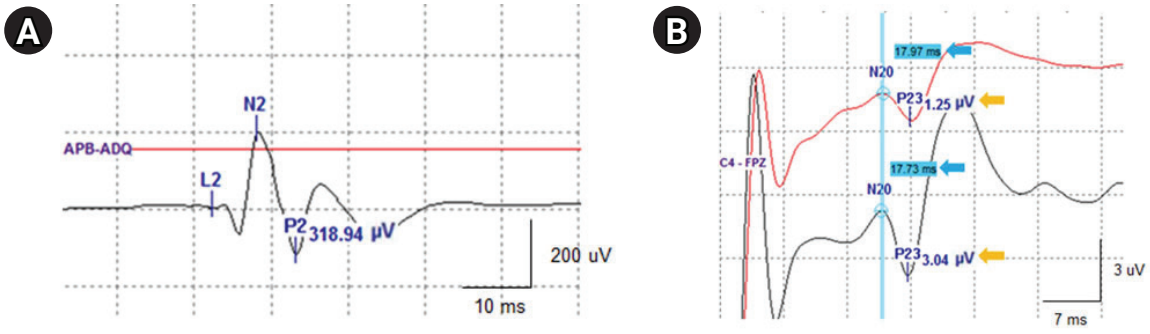

Fig. 2. Results of evoked potentials in case 5. Baseline MEP (red line) recorded from the abductor pollicis brevis muscle was not evoked but appeared at the end of surgery (black line) (A). Median SSEP showed a 143% increase in amplitude compared with the baseline (orange arrows), but there was no significant change in N20 latency (blue arrows) (B).

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ty is being continuously emphasized. Based on previous reports, our warning criteria for IONM during cerebrovascular surgery are as follows: 1) MEP amplitude is reduced by 50% or more, 2) SSEP amplitude is reduced by 50% or more, or 3) SSEP latency is increased by 10% or more [7,9]. Such warning signals indicate not only neural insults such as blood flow reduction or nerve damage during surgery but also represent significant changes concerning systemic conditions such as hypoxia, hypothermia, hypotension, and anesthesia [5]. On the other hand, little has been known about the change in EP when blood flow improved in patients with ischemia. One previous study reported that the degree of recanalization after mechanical thrombectomy was related to changes of EP and functional outcomes in patients with hyperacute stage stroke [10]. However, there has been no study demonstrating the relationship between improvement of cerebral blood flow and corresponding EP changes after the direct bypass surgery in the subacute stage of stroke patients with large artery occlusion.

In this study, the MEP amplitude showed an average of 50% or more improvement in amplitude. In case 5, APB-MEP was not evoked at baseline. However, at the end of the surgery, APB-MEP was evoked in addition to improvements in other EP findings. In case 3, no MEP and SSEP were evoked at baseline. This patient was transferred from other hospital with left MCA infarction. Upon initial assessment, according to Medical Research Council (MRC), muscle strength in his right upper and lower extremities were 1/5 (Trace). Preoperative EP study was performed 24 hours prior to the surgery, and neither MEP nor SSEP was evoked at the right upper and lower extremities. However, MEP and SSEP were normally evoked in the left upper and lower limbs. Therefore, unobtainable EP on the right side at the baseline study were more likely to be the result of cerebral infarction rather than the possibility of polyneuropathy, such as diabetes. Most MEP and SSEP were not evoked after the surgery, however, at the end of the surgery, although it was a minute change, a waveform was evoked on tibial SSEP.

In our opinion, this SSEP change was associated with the improvement of MTT in CS on PWI performed one week after the surgery. However, there was no sufficient recovery of cerebral blood flow to induce dramatic improvement of EP during surgery.

A comprehensive analysis of the aforementioned EP findings revealed that MEP amplitude shows an immediate response to the improvement in cerebral perfusion after STA-MCA anastomosis. We view that it is because MEP itself has high sensitivity and MEP showed a more dramatic change because the motor tract is mainly supplied by the MCA. All patients' baseline EP data were gained just before the opening of dura mater. EP data obtained after the skin closure were used as the end of operation data. In this study, baseline EP data were obtained just before the opening of dura mater. This means that the baseline MEP data used in this study had a time interval to minimize the effect of the NMB agent, and thus, the effect of the NMB agent can be excluded in the interpretation of MEP findings. The time taken to obtain baseline EP data from the first train-of-four response was 85.4 minutes (75-101).

On the other hand, SSEP did not show consistent improvements as expected. Consequently, our reasoning is that the intra-operative hemodynamic changes after the vascular anastomosis are not enough to reduce the SSEP latency or increase SSEP amplitude in terms of temporal aspect.

In our study, most MTTs were reduced at 1 week after surgery. In case 5, postoperative PWI showed that MTT was delayed after surgery. PWI findings were in contrast to EP findings and functional assessments, both of which showed improvements. However, judging by the fact that both mRS and MBI results improved, we infer that MTT delay is not necessarily related to a functional change. In a previous study comparing PWI results after STA-MCA bypass surgery, MTT showed gradual improvements in the 1-week and 3-month postoperative PWI studies, and these results were consistent with those of our study [4].

In STA-MCA bypass surgery, multimodal EP monitoring can be used to predict the improvement in brain perfusion intraoperatively. In particular, in this case report, MEPs showed more reliable and consistent changes than did SSEPs during bypass surgery.

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Instructions for Authors

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Journal of Electrodiagnosis and Neuromuscular Diseases (JEND), an official journal of the Korean Association of EMG Electrodiagnostic Medicine, is published twice a year. It regards all aspects of EMG, electrodiagnostic medicine, and neuromuscular diseases, including clinical practice, experimental and applied research, and education, and its formal abbreviated journal name is J Electrodiagn Neuromuscul Dis.

The manuscript guidelines for JEND are based on the "Uniform Requirements for the Manuscripts Submitted to Biomedical Journals" published by the International Committee of Medical Journal Editors (http://www.icmje.org), and instructions which are not mentioned in the present guidelines are referred to the guidelines stated in the Uniformed Requirements.

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4) Online resource

Morse SS. Factors in the emergence of infectious diseases. Emerg Infect Dis [serial online] 1995 Jan-Mar [cited 1996 Jun 5]; 1(1): [24 screens]. Available from: URL:http://www.cdc.gov/ncidod/EID/eid.htm

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