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A case of neuromyositis in a patient with dermatomyositis

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Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by proximal muscle weakness and typical skin lesions such as heliotrope rash, Gottron rash. Peripheral nerve involvement in DM is extremely rare and has been reported as neuromyositis since 1983. But the pathogenesis of neuromyositis is not clear. In Korea, there is no case reports of neuromyositis associated with DM. Herein, we report a 42-year-old man with DM who presented with neuromyositis. A 42-year-old man visited our clinic with limbs weakness and skin lesions on the face and hands. He was diagnosed with DM with interstitial lung disease based on the criteria of Bohan and Peter and started taking steroid and azathioprine. Unusually, electromyography (EMG) study on upper and lower extremities showed normal at the time of DM diagnosis, but nerve conduction study (NCS) suggested peripheral neuropathy on right peroneal nerve and left median nerve. Additional EMG study was not performed on the affected areas because there was no clinical symptom and sign of peripheral neuropathy. Four months after treatment, he complained a right ankle weakness that developed gradually for five days. We performed electrodiagnostic test again to determine the exact state. Both NCS and EMG results showed a right common peroneal neuropathy at knee level with moderate partial axonotmesis and denervation potentials. At that time, the muscle strength of proximal limbs was improved and laboratory findings such as creatine kinase were all normal. Therefore, there was no evidence that clinically DM worsened. Also, other causes such as trauma and infection which may cause a right ankle weakness could not be found. Finally, we diagnosed neuromyositis, a peripheral nerve involvement in DM. We decided that more slowly tapering of steroid and increasing dose of azathioprine. Since then, the right ankle weakness was improved little by little and almost completely recovered after 6 months, which was confirmed in follow-up NCS. This case is the first case of neuromyositis in DM reported in the literature in Korea and indicates that peripheral neuropathy is one of the important extramuscular manifestation of DM.

Table 1. Results of nerve conduction study

	1st study			2nd study			3rd study		
Motor nerve	**Lat (ms)**	**Amp (mV)**	**CV (m/s)**	**Lat (ms)**	**Amp (mV)**	**CV (m/s)**	**Lat (ms)**	**Amp (mV)**	**CV (m/s)**
Left Peroneal	3.0	5.3	47.7	3.92	3.2	52.4	4.04	7.2	53.4
Right Peroneal	3.42	2.8	47.2	4.13	1.14	48.8	4.90	3.3	50.0
Sensory nerve	**Lat (ms)**	**Amp (uV)**	**CV (m/s)**	**Lat (ms)**	**Amp (mV)**	**CV (m/s)**	**Lat (ms)**	**Amp (uV)**	**CV (m/s)**
Left Peroneal Superficial	2.4	5.7	58.3	2.98	5.7	47.0	2.69	13.2	52.0
Right Peroneal Superficial	0.0	0.0	0.0	3.09	1.46	45.3	2.51	7.3	55.8

1st study means a study performed at the time of dermatomyositis diagnosis.
2nd study means a study performed at the time of neuromyositis diagnosis.
3rd study means a study performed after recovery of neuromyositis.
Lat, latency; Amp, amplitude; CV, conduction velocity

Table 2. Results of eletromyogrphy study

	2nd study						
	Spontaneous Activity				Voluntary Activity		
Muscle	**Fib**	**PSW**	**Fasc**	**Amp**	**Dur**	**Polyph**	**IP**
Right EHL	1+	1+	None	Normal	Normal	+	PIP-CIP
Right PL	2+	2+	None	Normal	Normal	Normal	PIP-CIP
Right TA	2+	2+	None	Normal	Normal	+	PIP-CIP

2nd study means a study performed at the time of neuromyositis diagnosis.
EHL, extensor hallucis longus; PL, peroneus longus; TA, tibialis anterior
Fib, fibrillation potentials; PSW, positive sharp waves; Fasc, fasciculation; Amp, amplitude
Dur, duration; Polyph, polyphasic; IP, interference pattern; PIP, partial interference pattern; CIP, complete interference pattern

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폐출혈을 동반한 육아종증다발혈관염 1예

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육아종증다발혈관염은 대표적인 전신 혈관염으로, 육아종을 형성하거나 괴사를 유발하는 염증질환이다. 상기도, 폐 및 신장을 침범하여 다양한 임상양상으로 발현하며, Proteinase 3-antineutrophil cytoplasmic antibody (PR3-ANCA 또는 C-ANCA) 양성 소견이 진단에 도움이 된다. 기관 및 기관지 등 상기도 침범 시 협착으로 인한 기류 폐쇄를 일으키고, 폐실질 침범 시에는 다발성 결절, 폐출혈 또는 폐쇄성 폐렴을 일으킨다. 저자들은 폐출혈을 동반한 육아종증다발혈관염으로 진단 후 혈장교환술 및 Rituximab 치료로 호전된 증례를 경험하였기에 보고하는 바이다. **증례:** 환자는 만성 부비동염과 중이염 외에 특별한 과거력이 없던 70세 남자로 2주전부터 시작된 체중감소, 두통, 무기력증의 증상으로 입원하였다. 내원 시 백혈구 10900/mm^3(다형백혈구 83.7%), C-반응단백 200.3mg/L, 적혈구침강속도 104mm/hr BUN/Cr 31.8/1.17mg/dl이었으며, 소변 검사에서 적혈구 many/HPF의 혈뇨가 발견되었다. 흉부 X-ray에서 양측 상엽의 다발성 결절 발견되었고, 기관지 내시경적 조직검사에서 만성 육아종성 염증소견이 관찰되었다. 입원 7일째 양쪽 다리에 점상 출혈이 나타났고, BUN/Cr 42.5/3.73mg/dl으로 상승소견 보였으며, 입원 10일째 갑자기 발생한 객혈 및 호흡곤란 소견으로 재시행한 흉부 컴퓨터 단층촬영에서 양 폐야에 변연이 불규칙한 간유리모양 음영 소견 관찰되었고, hemoglobin의 저하가 발생하였으며, 기관지 내시경 검사에서 기관지 내 출혈 소견이 있어 미만성 폐출혈에 합당하였고, 위의 결과들과 함께 C-ANCA 양성 소견으로 육아종증다발혈관염을 진단하였다. 미만성 폐출혈의 치료를 위해 스테로이드 충격요법을 포함한 고농도 스테로이드 요법과 함께 혈장교환술, Rituximab 요법을 시행하였고, 신기능 감소로 혈액투석을 시행하였다. 미만성 폐출혈과 신기능 감소, 부비동염과 중이염 등 환자의 증상은 급속히 호전되었고, 현재 혈액투석을 중단하고 안정적으로 추적관찰 중이다.