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Multiple Symmetric Lipomatosis Presenting with Bilateral Brachial Plexopathy

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Dear Editor,

Multiple symmetric lipomatosis (MSL), also called Madelung's disease, is a rare disease characterized by the growth of nonencapsulated masses of adipose tissue.[1] The disease causes not only cosmetic issues but also various clinical problems depending on the location and extent of the lesion,[2] with neuromuscular manifestations such as polyneuropathy, myopathy, and ataxia having been reported.[3] Herein we report on a case of bilateral brachial plexopathy caused by extensive lipoma associated with MSL.

A 55-year-old man presented with progressive bilateral hand weakness. He first noticed a weak grip with his right hand 2 years before the presentation. The symptom progressed slowly, and his left hand also became weak a few months later. The patient often experienced paresthesia in both arms, but did not complain of proximal arm or leg weakness. He had been drinking about 200 mL of alcohol every day for 30 years. A physical examination revealed excessive soft tissues around his neck, shoulder, and forearm (Fig. 1A). His hand grip, finger abduction, wrist flexion/extension, and elbow extension were extremely weak (Medical Research Council scale grades from 1 to 3) on both sides with muscle atrophy in the first dorsal interossei, hypothenar, and pectoralis major muscles. Deep tendon reflexes were absent in both arms. A nerve conduction study revealed no potentials in the ulnar and radial motor nerves or the medial and lateral antebrachial cutaneous nerves of the left arm. Electromyography revealed positive sharp waves and fibrillation potentials in the first dorsal interosseous, abductor pollicis longus, and extensor carpi radialis of the left arm. Long-duration, high-amplitude motor unit action potentials (MUPs) were present in all muscles of the left arm with reduced recruitment of MUPs. Magnetic resonance images showed extensive

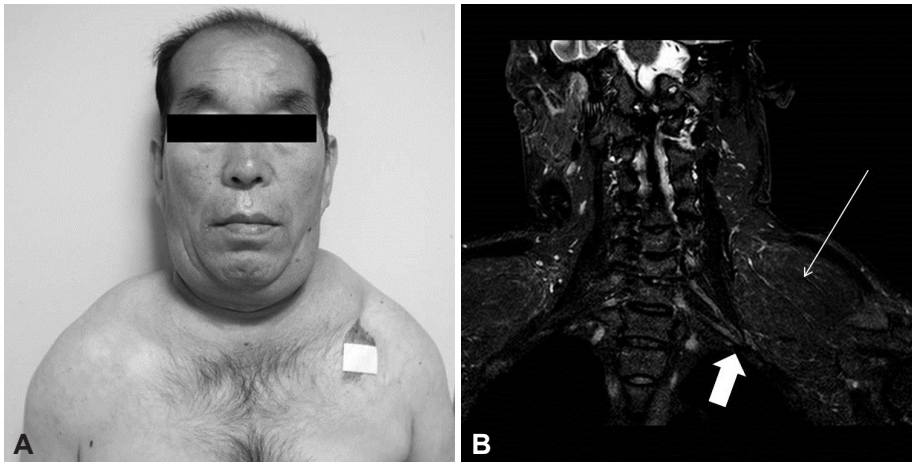

Fig. 1. A: Excessive fatty tissues around the neck and shoulder. B: Coronal fat-suppressed T2-weighted MRI reveals the brachial plexus (thick arrow) compressed by lipoma (thin arrow).

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fat proliferation and infiltration at the face, neck, shoulder, and axilla that resulted in compression of the brachial plexus (Fig. 1B). He was diagnosed with bilateral brachial plexopathy secondary to MSL, and was operated on to relieve the compression of the brachial plexus. After the surgery, the weaknesses in his hands were slightly improved.

The incidence of MSL is highest in Mediterranean countries, and it is uncommon in Asia (including Korea). A case-series study of Korean MSL patients found that most patients visited hospital because of a mass lesion and some of them were complicated with peripheral neuropathy.[4] The etiology of the disease is not fully understood, but most patients included in previous studies have had a history of chronic excessive alcohol consumption.[5] While the exact role of alcohol in the pathophysiology remains elusive, defects in adrenergic stimulated lipolysis or hypertrophy of the brown fat cells have been proposed as an alternative explanation for the pathomechanism.[6] The differential diagnosis should include other forms of generalized lipomatosis such as Bannayan-Zonana syndrome, Cowden syndrome, Proteus syndrome, and Dercum disease.[7] Familial multiple lipomatosis manifests as multiple encapsulated lipomas with autosomal dominant inheritance, but with relative sparing of the head and shoulders.[8] Surgical excision is the treatment of choice. Liposuction can be applied alone[9] or in combination with lipectomy.[10]

Conflicts of Interest

The authors have no financial conflicts of interest.

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