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Madelung Disease

To the Editor:

A 42-year-old Japanese man had a neck mass for 6 months that gradually grew in size. Similar masses appeared in the posterior part of bilateral auricles and upper arms. His medical history was only remarkable for alcoholic liver disease. He had been drinking 2 liters of beer per day for 20 years and smoking 20 cigarettes per day for 20 years. His medication and family history were unremarkable. Physical examination revealed symmetrical, soft, mobile masses on the neck and bilateral upper arms (Figure 1). Laboratory tests revealed elevated aspartate aminotransferase (150 U/L) and gamma-glutamyl transpeptidase (1078 U/L). Magnetic resonance imaging of the head and neck showed nonencapsulated soft-tissue masses dispersed over the superficial and deep fascial spaces (Figure 2); accordingly, Madelung disease was considered. Complete tumor resection was performed. Postoperative pathology was consistent with Madelung disease. The patient recovered from surgery uneventfully without obvious recurrence.

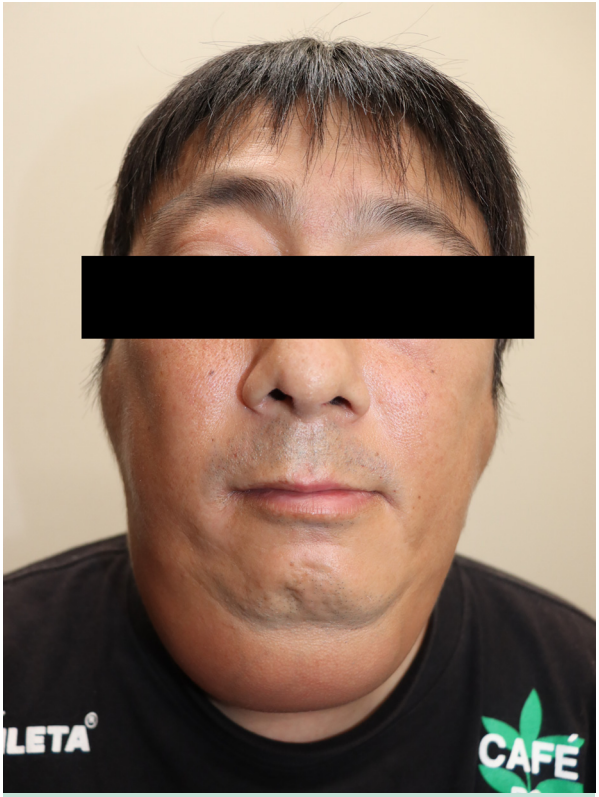

Figure 1 Physical examination revealed symmetrical, soft, mobile masses on the neck and bilateral upper arms.

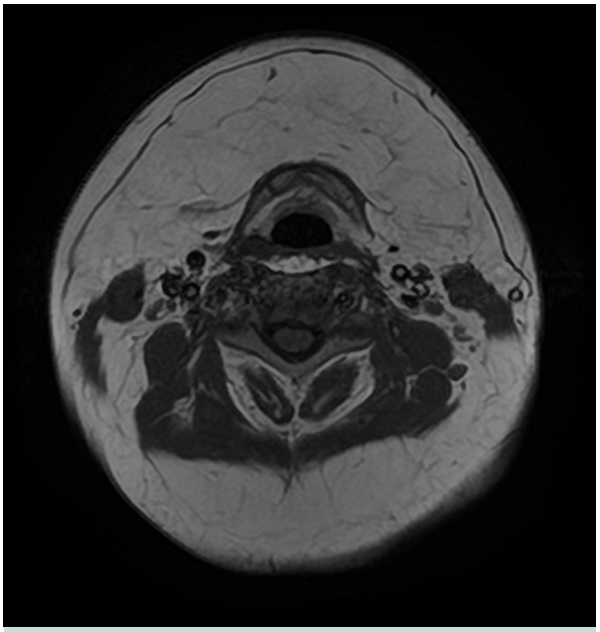

Figure 2 Magnetic resonance imaging of the head and neck showed nonencapsulated soft tissue masses dispersed over the superficial and deep fascial spaces.

Madelung disease is an exceedingly rare disorder of adipose metabolism in Japan, presenting as multiple, symmetrical, nonencapsulated fatty masses in the maxillofacial region, neck, shoulder, trunk, limbs, and other regions.[1,2] Madelung disease primarily affects men, especially those from the Mediterranean region.[1] Most cases of Madelung disease (90%) have a history of alcoholism.[1] Dysphagia and dyspnea may result from laryngeal or mediastinal involvement. Most instances are sporadic; however, a familial form characterized by maternally inherited mitochondrial gene mutation has been reported.[3] Differential diagnoses of Madelung disease include solitary lipoma,

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encapsulated lipoma, familial multiple lipomatosis, and liposarcoma.[1] Extensive lipectomy, the standard treatment for Madelung disease, can help achieve dramatic functional improvements and increase the patient's quality of life.[1,2] However, the recurrence of Madelung disease commonly occurs.[4] Abstinence from alcohol is not associated with spontaneous regression, but it reportedly reduces the recurrence rate.[5]

In conclusion, multiple fatty deposits involving the mandible in heavy drinkers should be differentiated from Madelung disease.

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