

Madelung's disease in a patient with chronic alcohol abuse

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Submitted: 4 December 2012

Accepted: 5 December 2012

Arch Med Sci 2014; 10, 6: 1268–1269
DOI: 10.5114/aoms.2013.36914
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A 47-year-old man with a history of chronic alcohol abuse was admitted to our hospital because of symmetrical face soft tissue enlargement for differential diagnosis. The lesions had appeared 4 years earlier and had been asymptomatic.

Physical examination revealed painless, elastic tumors of the subcutaneous tissue in the mandibular angle region, with no clear relation to lymph nodes or salivary glands (Figure 1 A). The laboratory tests showed elevated aspartate aminotransferase (ALT), 127 U/l; elevated alanine aminotransferase (ALT), 91 U/l, elevated γ -glutamyl transpeptidase (GGT), 643 U/l, and elevated lactate dehydrogenase (LDH) activity, 342 U/l. Test for anti-nuclear antibodies (ANA) was negative. HIV and HCV infections were excluded. Lipid profile results were within the normal range. An abdominal ultrasound revealed a fatty liver, but no other abnormality was noted. Ultrasonography of the parotid, submandibular and cervical region confirmed the correct size and structure of the salivary and thyroid glands and slight enlargement of the lymph nodes in both submandibular regions. In the place of visible symmetric thickening there was a tissue described as lipofibroma, 49 mm \times 24 mm on the right side and 44 mm \times 23 mm in size on the left side. Fine-needle biopsy revealed lipomatous tissue. A diagnosis of Madelung's disease was made.

At the follow-up visit after 1 year, the patient had a computed tomography (CT) scan performed, which showed a slight increase in the primary fatty deposit and the appearance of another in the neck area (Figure 2 A–C). Six months later the patient was disqualified from surgical treatment. The decision was motivated by the non-life-threatening condition and asymptomatic nature of the masses.

Madelung's disease (also known as benign symmetric lipomatosis or Launois-Bensaude syndrome) is a rare metabolic disorder of unknown etiology [1]. This disease usually affects men at the age of 30 to 60 years from the Mediterranean area. In reported cases, almost 90% of patients had a history of long-term alcohol abuse [2]. The disease is characterized by the presence of prominent, symmetrical deposits of non-encapsulated adipose tissue in the region of head, neck, nape and upper parts of the trunk [2, 3]. In contrast to other cases described in the literature, in our patient, the primary subcutaneous fat masses appeared in the parotid region, while the most common initial location is the neck. Although benign in nature, sometimes the fatty masses can reach very large sizes causing dyspnea, dysphagia, fatigue, and reduced neck movement ability [4]. The differential diagnosis includes familial multiple lipomatosis, simple obesi-

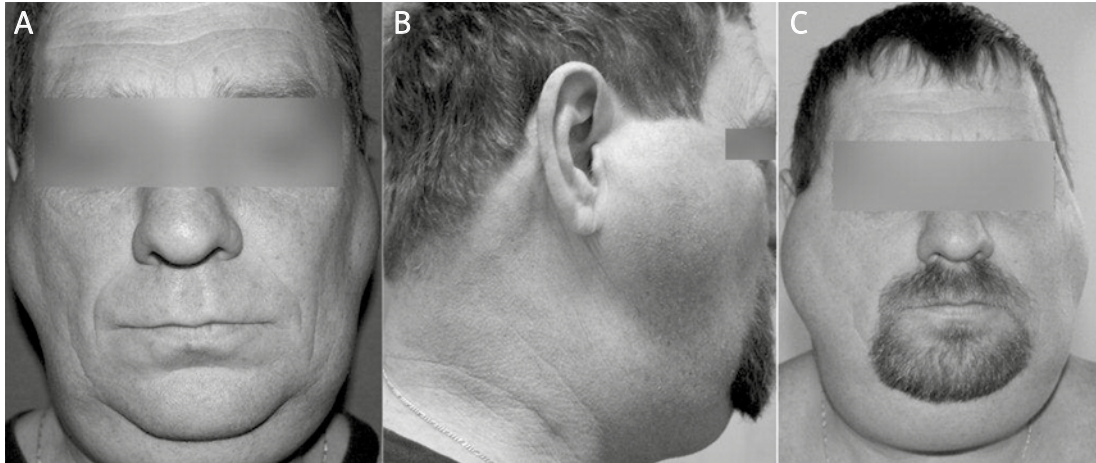


Figure 1. A – Frontal view of the patient at time of first hospitalization with visible symmetrical fatty masses. B, C – Frontal and lateral view of the patient at follow-up visit after 1 year; increase in the primary fatty deposit

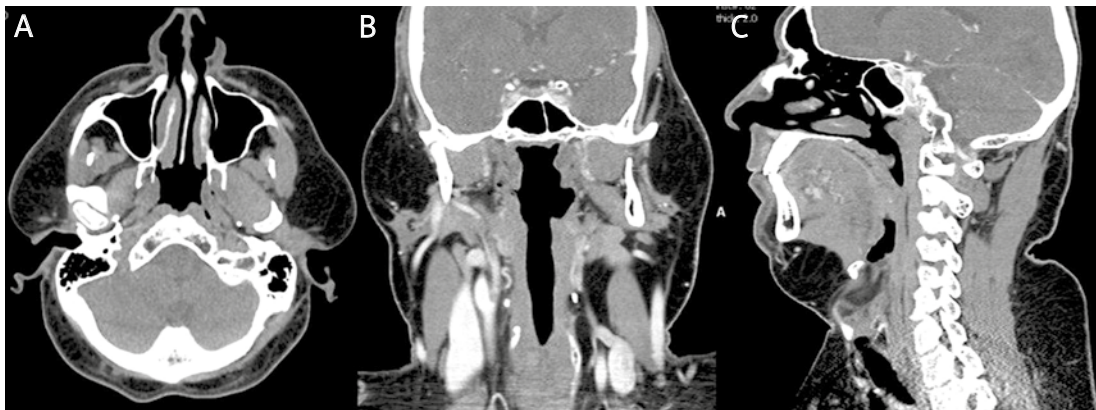


Figure 2 A-C. Head and neck CT showing the symmetrical enhancement of the lipomatous tissue

ty, Cushing's syndrome, angiolipoma, neurofibroma and liposarcoma [2, 4, 5]. The most effective treatment is surgery, especially in patients with esthetic deformity or compression of the aerodigestive tract [1, 3].

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