

# MULTIPLE SYMMETRIC LIPOMATOSIS (MADELUNG'S DISEASE): REPORT OF TWO CASES

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Multiple symmetric lipomatosis (Madelung's disease) is a rare disease characterized by non-encapsulated adipose deposits in the neck, the superior part of the trunk and, very rarely, in the limbs. It is common in middle-aged Caucasian Mediterranean males but very rare in Chinese people. We describe two cases of Madelung's disease: both patients presented complaining of a development of a painless neck mass that was increasing in size. Panendoscopy was normal. Both underwent incisional biopsies that revealed benign adipose tissue characterizing Madelung's disease. We discuss the clinical presentation, associated morbidity, and treatments for Madelung's disease.

**Key Words:** Madelung's disease, multiple symmetric lipomatosis  
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Multiple symmetric lipomatosis (Madelung's disease), a rare disease for which the cause remains undetermined, is characterized by diffuse multiple symmetric, non-encapsulated, fatty accumulations involving the head and neck region and upper trunk [1]. It occurs most frequently in alcoholic men from Mediterranean countries and is very rare in Chinese people [1,2]. We report our experience with two Madelung's disease patients and discuss the clinical presentations of Madelung's disease, the associated morbidity, and methods of treatment.

## CASE PRESENTATIONS

### Case 1

The first case was a 63-year-old male who presented with a 2-year history of multiple bilateral large disfiguring neck masses that were increasing in size (Figure 1). The masses

were soft, with no definite margin, and movable; there was no tenderness or inflammation. The patient, a heavy smoker and heavy drinker, had a 2-year history of gout. Laboratory examination revealed abnormal liver function and hyperuricemia. Neck computerized tomography (CT) scan revealed diffuse prominent large fatty tissue formations around the neck, supraclavicular regions and upper neck subcutaneous area (Figure 2A). Panendoscopy was normal. A biopsy was taken from one of the neck lumps. Pathology revealed the mature adipose tissue found in patients with Madelung's disease (Figure 3). There was no specific treatment for the neck lesions. Care was directed towards the patient's hyperuricemia and liver disease.

### Case 2

A 29-year-old male came into our hospital with progressive bilateral neck enlargement and a submandibular mass that had extended into the supraclavicular region over 6 months. Physical examination found that the neck mass was soft, but there was no tenderness or inflammatory changes. The patient had a history of heavy alcohol and cigarette use. Laboratory examination revealed abnormal liver function and hyperuricemia. The results of neck CT scan (Figure 2B) and pathology, and the management were similar to those in Case 1.

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**Figure 1.** Case 1: marked symmetric accumulation of adipose tissue deposits in the neck and shoulder.

## DISCUSSION

Madelung's disease is characterized by diffuse and symmetric deposits of non-encapsulated fat, generally affecting the cervical region and upper trunk. It was first described

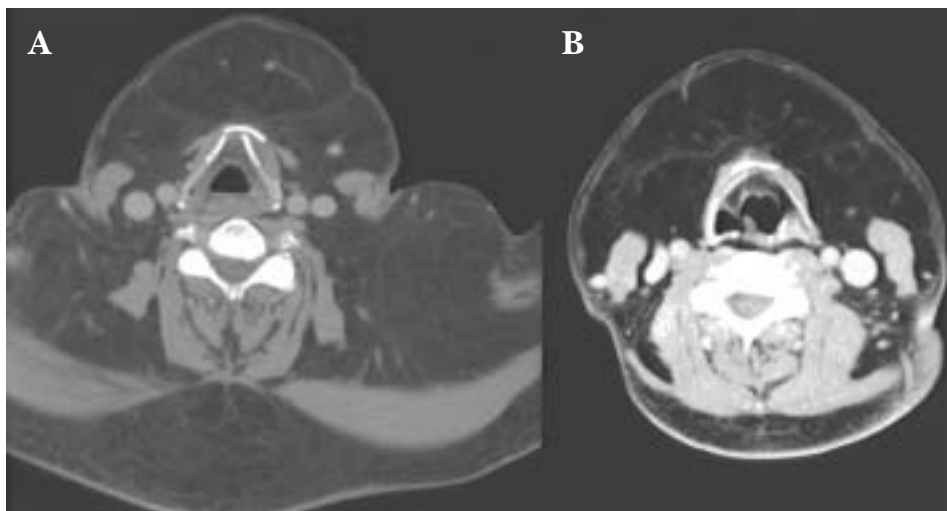
by Brodie in 1846. In 1888, Madelung reported 33 patients with cervical lipomatosis. Ten years later, Launois and Bensaude further defined this syndrome as multiple symmetric non-encapsulated fatty accumulations diffusely involving the head and neck region and upper trunk.

The etiology of this disease is unknown. According to the literature [1–3], 60% to 90% of patients with Madelung's disease have a history of alcoholism. Hyperuricemia, gout, liver disease, polyneuropathy, diabetes and glucose intolerance have occasionally been identified in patients with Madelung's disease [3]. Our two patients had a history of alcoholism, liver disease and hyperuricemia.

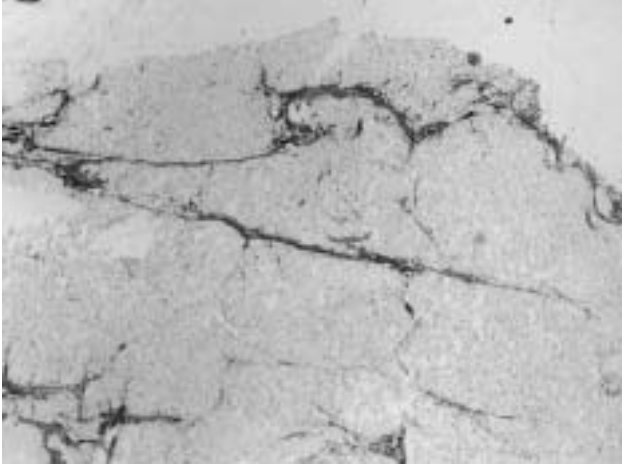
Madelung's disease usually has no symptoms apart from cosmetic disfigurement. Massive symmetric deposition of fat leads to cosmetic deformities in the parotid region ("hamster cheeks"), cervical region ("horse collar") and posterior neck ("buffalo hump") [4]. In some cases, the growth of adipose tissue can cause compression in the aerodigestive tract and lead to dyspnea, dysphagia, and dysphonia. In serious cases, emergency tracheostomy has been used to relieve airway obstruction.

Diagnosis of Madelung's disease is based on accurate history taking and clinical examination. Panendoscopy is necessary to exclude the possibility of malignancy originating in the head and neck region. Imaging techniques such as magnetic resonance imaging and CT provide useful information, but sonography does not [1]. Fine-needle aspiration may assist in the diagnosis.

According to one report, Madelung's disease was treated successfully with the  $\beta$ 2-agonist salbutamol, which acts on lipolysis via adrenergic stimulation [3]. Surgical excision is reserved for patients who have aesthetic deformities and/or significant compression of the respiratory tract [3].



**Figure 2.** (A) Case 1: neck computerized tomography (CT) scan reveals diffusely prominent, large fatty tissue formations around the neck, supraclavicular regions, and upper neck subcutaneous area. (B) Case 2: neck CT scan showing similar findings.



**Figure 3.** Case 1: photomicrograph of excised specimen shows normal, mature adipose tissue (hematoxylin & eosin,  $\times 1$ ).

Complete surgical removal of the tumor may jeopardize important anatomic structures because the lipomas can infiltrate or encase these structures [3,4]. Liposuction techniques may offer a better alternative to standard surgical therapy in some cases and seems to be the best way to reduce cosmetic scarring [5]. There was no airway compression or cosmetic concerns in our patients. The treatment of their disease focused on metabolic disturbances and liver dysfunction.

Madelung's disease has degenerated into malignancy in one reported case [6], but the prognosis for Madelung's

disease patients is generally good, although recurrence is common because of the difficulty in completely excising the tumors.

The occurrence of Madelung's disease in our patients may indicate that the disease may no longer be solely associated with Caucasian Mediterranean men; physicians in Asia should be aware of the possibility of Madelung's disease in alcoholic patients with multiple symmetric neck mass deposits.

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# 多發對稱性脂肪沉著病 (Madelung's Disease) — 二病例報告

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多發對稱性脂肪沉著病 (Madelung's disease) 是一種罕見的疾病，主要的特徵為脂肪組織廣泛的沉基於頸部及上胸部，一般好發於地中海地區中年男性且有酗酒習慣者，發生於中國人種更屬相當罕見。本文報告二病例，二病例皆主述無痛性頸部腫塊一段時間，鼻咽喉內視鏡檢查無異常發現，電腦斷層檢查發現腫塊廣泛分佈於前、後頸部及上胸部，經手術切片，病理報告皆為良性脂肪組織，與臨床診斷 Madelung's disease 相符合。由於此病在東方人種上實屬罕見，是故特究其臨床表現、症狀和徵象、影像學特徵、治療方法提出報告。

**關鍵詞：**多發對稱性脂肪沉著病，Madelung's disease

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