

# Madelung's disease: a case report and literature review\*

*Doença de Madelung: relato de caso e revisão da literatura*

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**Abstract** Madelung's disease is characterized by the presence of symmetric masses of adipose tissue on the neck, shoulders, arms and the upper trunk. Computed tomography demonstrates characteristic imaging findings, and it is considered to be the method of choice for diagnosis, pre-operative staging, and postoperative follow-up. The authors report the case of a man with typical tomographic findings of Madelung's disease.

**Keywords:** Madelung's disease; Benign multiple symmetric lipomatosis; Lipomas; Alcoholism.

**Resumo** A doença de Madelung é caracterizada por massas simétricas de tecido adiposo no pescoço, ombros, braços e parte superior do tronco. A tomografia computadorizada demonstra achados de imagem característicos e é considerada o método de escolha para diagnóstico, estadiamento pré-operatório e acompanhamento pós-cirúrgico. Relatamos o caso de um homem com aspectos tomográficos típicos da doença de Madelung.

**Unitermos:** Doença de Madelung; Lipomatose simétrica múltipla benigna; Lipomas; Alcoolismo.

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## INTRODUCTION

Multiple symmetric lipomatosis, also known as Madelung's disease or Launois-Bensaude syndrome, is a rare entity characterized by painless, subcutaneous diffuse deposition of adipose tissue on the neck, upper trunk, arms and legs<sup>(1)</sup>.

Historically, Madelung's disease has been most frequently observed in men (male-female ratio = 15:1) aged between 30 and 60 years. The disease prevalence is increased among the Mediterranean population (reported incidence in Italy: 1/25,000 men), and there is a relationship between this disease and excessive consumption of alcohol, particularly red wine<sup>(2)</sup>. Such a condition develops over a period of months to years. The patients complain of their appearance although, in advanced cases, the increase in fat deposits may cause dyspnea or dysphagia<sup>(3)</sup>. Association of Madelung's disease with diabetes, gout,

alcoholism and other systemic diseases has been observed<sup>(4)</sup>.

The authors report the case of a 50-year-old man with painless masses in the cervicofacial region and in the upper trunk, which increase over a period of three to four years (Figure 1).

## CASE REPORT

The patient's clinical history indicated systemic arterial hypertension effectively controlled with medication. The patient denied increase in caloric intake and reported alcohol consumption of 1.0 liter of distilled beverage per day during 28 years. At physical examination, foci of volume increase were observed on the face, neck and upper

trunk, with no sign of edema. Subsequently, the patient was submitted to head and neck computed tomography (CT) with basis on a clinical hypothesis of lipomas.

The CT findings showed a symmetric and diffuse subcutaneous distribution of homogeneous, nonencapsulated adipose material in the cervicofacial region and the upper trunk, with no sign of contrast enhancement. The deep muscle planes demonstrated preserved morphologies, contours and attenuation coefficients, slightly displaced by the adipose material, but with no signs of direct infiltration. Lymph node enlargement, mass with soft tissue attenuation, marked lymphatic malformations or vascular dilatation were not observed (Figure 2).



**Figure 1.** Lateral and frontal views of the patient demonstrating symmetrical enlargement of soft tissues resulting from fat deposition.

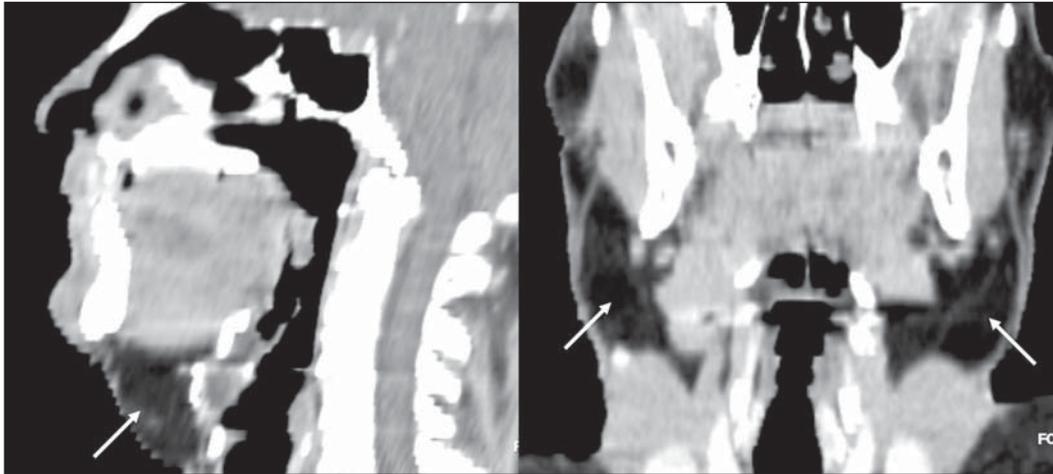
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**Figure 2.** Sagittal and coronal reconstructions demonstrating deep and diffuse deposition of adipose tissue (arrows).

## DISCUSSION

Madelung was the first author to extensively describe this entity in a series of patients in 1888; hence the name Madelung's disease. In 1889, Launois and Bensaude reported 65 cases, expanding the description of this disease that also has been called as Launois-Bensaude syndrome. Currently, reports on more than 270 cases are found in the literature<sup>(2,5)</sup>.

Multiple symmetric lipomatosis is classified into two types, as follows: type I, with lipomatous masses in the parotid region, neck, suprascapular and deltoid regions. In this group, deep involvement causing dyspnea and dysphagia may be observed in some patients. In type II, lipomatosis is diffuse, with clinical appearance of simple obesity, and without the symptoms of deep involvement present in type I<sup>(6)</sup>. Typically, the masses do not respect cleavage planes, penetrating all levels of the tissue, so their surgical exeresis is more difficult<sup>(4)</sup>.

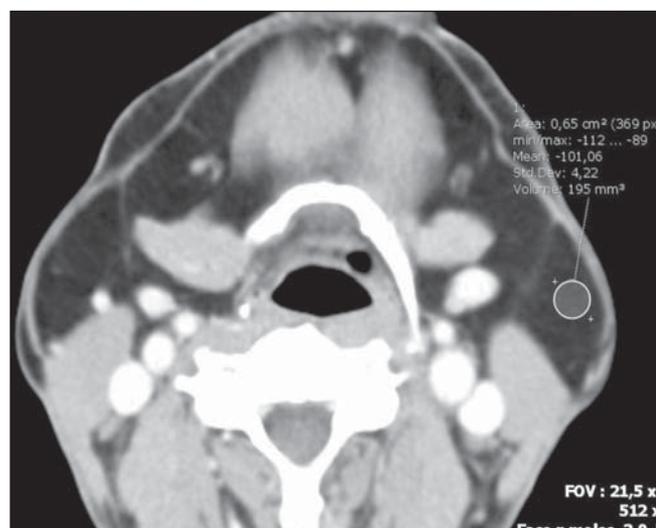
Several types of lipomatosis typically differentiated by anatomic location and varied clinical symptoms have been described. Such types include multiple symmetric lipomatosis, congenital infiltrative lipomatosis of the face, encephalocraniocutaneous lipomatosis, scapular waist lipomatosis, adiposis dolorosa, pelvic lipomatosis and mediastinal lipomatosis<sup>(5)</sup>. The clinical diagnosis is based on the recognition of the unique appearance of the patient, but in atypical cases, there may be confusion with lymphoproliferative disorders and other conditions<sup>(4)</sup>.

The etiology of Madelung's disease still remains unknown. Alcohol ingestion of more than 80 g per day during at least ten years – a factor observed in up to 90% of cases described in the literature -, was also observed in the present case. Polyneuropathy, described as a sensory, motor and autonomic dysfunction, and detected in about 85% of patients with Madelung's disease, generally develops several years after the onset of lipomas, and was not present in this case<sup>(7)</sup>.

Chest radiography may show mediastinal enlargement. At CT, lipomatous, non-encapsulated and homogeneous tissue is observed, possibly in association with deep deposition (submuscular) of adipose tissue (Figure 3). This latter presentation is most frequently observed around the trachea and/or deeply located in the trapezius

muscle. In some cases, calcifications are identified within lipomas<sup>(7)</sup>.

Currently, CT is considered to be the method of choice for the diagnosis, preoperative staging and postoperative follow-up of patients with multiple symmetric lipomatosis. The evaluation of images directly on the CT display screen, on workstations or on computers equipped with visualization software allows a dynamic change of windows and levels parameters, besides the measurement of attenuation coefficients on regions of interest, which may improve the identification and characterization of the adipose material. Despite the absence of comments on this point on recent reviews<sup>(8)</sup>, some of the advantages of CT include multiplanar reformation that, although it is not essential for the diagnosis, this resource may refine the pre-



**Figure 3.** Contrast-enhanced axial image demonstrating diffuse deposition of adipose tissue involving deep structures.

sentation of findings, facilitating the analysis of adipose tissues distribution both by radiologists and nonradiologists (Figure 2).

The utilization of CT allows the investigator to rule out relevant differential diagnosis such as lymphoma and metastatic lymph node involvement. Also, it allows the tumor staging in cases where multiple symmetric lipomatosis is associated with malignant disease, particularly, squamous cell carcinoma of upper airways resulting from alcoholism and smoking whose association is frequently observed<sup>(9)</sup>.

Sonographic findings of lipomatosis are typical. However, despite the frequent utilization of ultrasonography in the investigation of neck masses, measurements of density by CT provide a higher diagnostic fidelity<sup>(6)</sup>.

MRI findings usually overlap with CT findings, but it is important to highlight that MRI is a more expensive and generally less available method<sup>(9)</sup>.

Despite the relatively low incidence of Madelung's disease, radiologists should know the characteristic imaging findings of this condition in order to be able to promptly achieve a correct diagnosis, ruling out overlap with other conditions and providing appropriate information for the required clinical management of the disease.

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