

# Multiple Symmetrical Lipomatosis: Madelung's Disease

Margarida Carvalho<sup>a, c</sup>, Pedro Palma<sup>b</sup>, Filipa Silva<sup>a</sup>, Marta Patacho<sup>a</sup>, Fatima Coelho<sup>a</sup>

## Abstract

Multiple symmetric lipomatosis or Madelung's disease is a rare condition which is characterized by multiple symmetric and disfiguring accumulation of non-capsulated fat tissue depositions mostly in upper arms, neck, and shoulder areas. The disease etiology is unknown, with the highest incidence in the Mediterranean region and an association with alcohol abuse. We present a case of a patient admitted to our internal medicine department for cervical mass study. This patient was already studied by his general practitioner because of thyroid disease suspicion. During our study, diagnosis of Madelung's disease with typical fat depositions was established. This article is a short review of the topic and points out the possible diagnostic difficulties presented in early stages of the disease.

**Keywords:** Madelung's disease; Multiple symmetrical lipomatosis; Etiology

## Introduction

Madelung's disease is a rare condition characterized by the multiple symmetric and disfiguring accumulation of fat depositions. This disease is also known as benign symmetric lipomatosis or the Launois-Bensaude syndrome [1, 2].

## Case Report

A 52-year-old man presented to our internal medicine department for evaluation of a neck mass without associated dyspnea or dysphagia (Fig. 1). Before this hospitalization, he was

examined in his general practitioner because of suspicion on thyroid gland disease. On physical examination, we noticed multiple symmetric mass lesions in the trunk and upper limbs (Figs. 2-4). His previous medical history was significant for a coexisting alcohol abuse and documented alcoholic hepatic disease. His laboratory blood analysis was relevant for a thrombocytopenia ( $91 \times 10^9$  U/L) and a slight increase in aspartate aminotransferase, alanine aminotransferase and gamma glutamyltransferase enzymes, less than three times the reference range. Neck ultrasonography revealed only adipose tissue. Excision biopsy of the neck adipose mass confirmed adipose tissue without malignant transformation.

The clinical aspects, histology and history of alcohol abuse were consistent with the diagnosis of Madelung's disease. The phenotypic appearance was typical but without the disfiguring aspects characteristic of the advanced states of the disease. Possibly for this reason, his general practitioner did not establish the diagnosis earlier. Because the patient was asymptomatic, he was discharged for an alcoholism recovery program and oriented to plastic surgery and internal medicine consults.

## Discussion

Madelung's disease is characterized by diffuse, symmetric, painless, non-encapsulated, and irreversible growth lipomatosis [3]. The etiology of the disease is unknown but recent studies suggest that mitochondrial disorder of brown fat tissue or a defect in the adrenergic stimulated lipolysis, is involved



**Figure 1.** Cervical lipoma mass.

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<sup>a</sup>Internal Medicine Department, Hospital Sao Joao, Alameda Professor Hernani Monteiro, 4200-319 Porto, Portugal

<sup>b</sup>Infectious Diseases Department, Hospital Sao Joao, Alameda Professor Hernani Monteiro, 4200-319 Porto, Portugal

<sup>c</sup>Corresponding Author: Margarida Carvalho, Alameda Professor Hernani Monteiro, 4200-319 Porto, Portugal. Email: amcoes@gmail.com

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**Figure 2.** Multiple lipoma masses in the upper limb and thorax.



**Figure 3.** Lipoma masses in the upper limbs.



**Figure 4.** Multiple lipoma masses in the thorax.

[4, 5]. Also, this disease is more common in the male gender (15:1 ratio), between 30 and 60 years old and in the Mediterranean population [3]. Chronic alcoholism history is present in 60-90% of the cases [2, 6].

Madelung's disease phenotype may vary but most of the cases present with adipose tumefactions in the face, neck, occipital fossa and shoulder areas. Fat deposits around the cervical region form a "buffalo hump" and a "horse collar", while fat deposits around the parotid region may appear as "hamster cheek" [3, 7]. Fat masses are symmetrically distributed, while distal arms and legs are spared [8]. Frequently associated findings include hyperlipidemia, hyperuricemia, gout, diabetes mellitus, hypertension, hypothyroidism, liver disease, and polyneuropathies [9].

The diagnosis is usually made based on clinical presentation, disease history and computed tomography or magnetic resonance. The imaging studies allow the evaluation of the extension and distribution of the mass lesions and facilitate surgery programming [3, 6, 7].

As the masses grow, patients with Madelung's disease may suffer from reduced neck mobility or compression of respiratory or mediastinal structures. These complications are the main surgery indication in this disease [2]. Rarely, some patients present with malignant degeneration to liposarcomas and oropharyngeal tumors [8].

Surgery is the only effective treatment. Alcohol abstinence may delay the disease progression and the post-operative recurrence [2, 8].

### Conclusion

Madelung's disease is very rare but must be remembered in patients with suggestive clinical presentation and history of alcohol abuse.

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