

Multiple symmetric lipomatosis

Lipomatose simétrica múltipla

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ABSTRACT

Multiple symmetric lipomatosis, also called Madelung's syndrome, is a rare disease characterized by the accumulation of rapidly growing benign fat tissue, particularly in the cervical region, back, and more rarely in the extremities and face. It is generally associated with chronic alcoholism. Surgical treatment is palliative, and relapse is common. This paper reports the case of a patient with multiple symmetric lipomatosis followed by a review of the literature addressing the diagnosis and treatment of this condition.

Keywords: Multiple symmetrical lipomatosis. Lipomatosis. Adipose tissue/pathology.

RESUMO

A lipomatose simétrica múltipla ou síndrome de Madelung é uma doença rara, caracterizada por depósitos de tecido adiposo de caráter benigno, de rápido crescimento, principalmente em região cervical, dorso e, mais raramente, membros e face, em geral relacionados ao alcoolismo crônico. O tratamento cirúrgico é paliativo e a recidiva é comum. Neste trabalho é relatado um caso de paciente portador de lipomatose simétrica múltipla, seguido de revisão de literatura enfocando diagnóstico e tratamento dessa afecção.

Descritores: Lipomatose simétrica múltipla. Lipomatose. Tecido adiposo/patologia.

INTRODUCTION

Multiple symmetric lipomatosis (MLS), also called Madelung's syndrome, is a rare entity of unknown etiology characterized by the symmetric and progressive accumulation of non-encapsulated lipomas¹.

Lipomas initially exhibit rapid growth followed by a slow and progressive evolution. MLS mainly affects the neck, arms, thorax, armpits, abdomen, and thighs as well as deeper structures such as the retroperitoneum and mediastinum². This condition is usually reported in adults between 30 and 60 years of age, affecting 15–30 fold more men than women; moreover, there are reported cases of the disease within families^{3–5}.

According to the literature, 60%–90% of patients with MLS have a history of alcoholism^{6,7}. Stopping alcohol consumption is associated with a discrete regression of fat accumulation; meanwhile, increased alcohol consumption appears to accelerate lipoma growth⁸. Hyperuricemia,

gout, liver disease, polyneuropathy, diabetes, and glucose intolerance are occasionally identified in patients with MLS⁶; however, some reports suggest that fat tissue accumulation is metabolically unrelated and is not necessarily accompanied by insulin resistance, possibly preventing lipotoxicity⁹.

This paper reports the case of a patient with MLS who received surgical treatment. In addition, the literature concerning this disease was reviewed.

CASE REPORT

The patient was a 48-year-old man with a sedentary lifestyle, former alcoholism (a usual daily intake of 1 liter distilled spirits for 20 years but abstemious for the last 3 years), a former smoking habit (30 pack-years but abstemious for the last 20 years), and no family history of MLS. The patient reported to the General Surgery Service of Santa Casa de Misericórdia of Itabuna with complaints of cervical

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and thoracic masses that had appeared 15 years ago. He had no complaints regarding other systems.

The nodules initially appeared in the suprasternal region, followed by the cervical region, back, abdomen, upper limbs, and submandibular region with rapid and painless growth but with increasing temperature. The patient had previously undergone 5 lipectomies that were followed by the growth of new masses.

On physical examination, the patient exhibited a good general condition, obesity, and presence of large lipomas in the face, submandibular, cervical, supraclavicular, and infraclavicular regions as well as the thorax, abdomen, back, and upper limbs (Figures 1 to 3).

Abdominal ultrasonography showed a moderate degree of hepatic steatosis, a small calculus in the gallbladder, and an expansive formation in the abdominal wall over the iliac fossa, suggesting large lipomas. Mammary ultrasonography revealed an isodense expansive formation in the fat planes of the costal edges. Meanwhile, thyrocervical ultrasonography revealed a normal-sized thyroid without adenomegaly and normal parotids. Laboratory test results are shown in Table 1.

The patient initially underwent lipectomy of the lower abdomen; 2 pieces of fat and subcutaneous tissue were removed, weighing 10.3 kg and 5.6 kg, respectively. A simple



Figure 2 – Left-sided view of the patient presenting with MLS, showing a scar from a previous lipectomy in the left flank.



Figure 1 – Patient presenting with multiple symmetric lipomatosis, with accumulation of symmetric lipomas in the face, abdominal, supraclavicular, and deltoid regions.



Figure 3 – Posterior view of the patient presenting with multiple symmetric lipomatosis, showing the bilateral symmetry of the lipomas.

Table 1 – Preoperative laboratory data.

Test	Results
Hemoglobin (mg/dl)	9.8
Hematocrit (%)	27.8
Leucocytes ($\times 10^3/\text{mm}^3$)	8,100
Platelets ($\times 10^3$)	274
Sodium (mg/dl)	144
Potassium (mg/dl)	5
Alkaline phosphatase (U/l)	144
Gamma glutamyl transferase (U/l)	28
HDL- cholesterol (mg/dl)	40
LDL- cholesterol (mg/dl)	41
Total cholesterol (mg/dl)	89
Triglycerides (mg/dl)	40
Aspartate aminotransferase (U/l)	16
Alanine aminotransferase (U/l)	11
Urea (mg/dl)	15
Creatinine (mg/dl)	0.53
Fasting glycemia (mg/dl)	103

HDL: high-density lipoprotein; LDL: low-density lipoprotein.

bilateral mastectomy was subsequently performed, and 2.4 kg and 2.8 kg fat and mammary tissue, respectively, were removed. Histopathologic examination yielded evidence of benign subcutaneous lipomas and hyperplasia in the mammary tissue. The patient had an uneventful recovery.

All ethical norms regarding studies involving human beings were in accordance with the revised Declaration of Helsinki. The patient was invited to participate in the case report. He signed the Free Informed Consent Form that guarantees confidentiality of personal data, and provided authorization to use his image.

DISCUSSION

MLS is a rare disease. It first described by Benjamin Brodie in 1846 in St. George Hospital in London and subsequently by Otto Madelung in 1888¹⁰. In 1898, the disease was defined by Launois and Bensaude in a series of patients as a well-defined clinical entity and termed benign symmetric lipomatosis⁸.

MLS is also known as Madelung's disease, Launois–Bensaude disease, symmetric adenolipomatosis, diffuse symmetric lipomatosis, lipomatosis simplex indolens, multiple symmetric lipomatosis, cephalothoracic lipomatosis, and familial benign cervical lipomatosis¹⁰.

It is a rare disease that occurs more frequently in people living in countries near the Mediterranean region². Approximately 385 cases of the disease worldwide have been indexed in the MEDLINE database. Eight cases have been reported in Brazil and published in journals indexed by SciELO.

Although the etiology of the disease remains unknown, it seems to be associated with an alcohol-induced endocrine disorder and a mitochondrial enzyme disorder³.

Enzi et al.⁸ classify MLS into 2 types. In type I MLS, the fat accumulations are limited, forming non-encapsulated masses that are distributed symmetrically in the upper region of the body. Lipomas are formed in the cervical region (“horse collar”), deltoids (pseudoathletic appearance), and back, giving the false impression that the patient has kyphosis of the spine (i.e., a humpback)⁴ like in the present case. In type II MLS, the distribution of lipomas is more diffuse. In many cases, the patient appears to have simple obesity. In addition to the upper part of the body, the inguinal and abdominal regions are frequently involved. However, there is no involvement of the mediastinum or compression of the aero-digestive tract. The body mass index is also increased⁴.

The differential diagnosis includes the following: sarcomatosis processes, angiolioma, lipoblastoma, neurofibromatosis, Dercum syndrome (i.e., painful lipomatosis)¹⁰, Hanhart syndrome, polydysplasia syndrome, Cushing disease, pseudoathletic appearance (in various forms of muscular dystrophy), lymphoproliferative diseases, and thyroid diseases.

Complications of the disease are rarely reported and include compression of the aero-digestive tract, causing dyspnea, dysphagia, and dysphonia. In serious cases, treatment involves surgical resection of the lipomatous tissue with the option of liposuction in the infiltrated areas. The cosmetic and functional benefits of surgical resection are occasionally temporary², and surgical treatment of the disease tends to be associated with increased recurrence⁵.

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