



Multiple symmetric lipomatosis: report of two cases

Lipomatose simétrica múltipla: relato de dois casos

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ABSTRACT

Multiple symmetric lipomatosis is an unusual disease of unknown etiology. It is characterized by symmetrical accumulation of fat tissue deposits in neck, face, trunk and proximal limbs. It is more common in male adult subjects with a history of alcohol intake. Palliative surgery is the main treatment and it can be performed by conventional open excision or liposuction. Recurrence is common. The objective of this article is to report two cases of multiple symmetric lipomatosis treated by liposuction in Regional Hospital of Asa Norte, in Brasilia - Federal District.

Keywords: Multiple Symmetric Lipomatosis. Madelung Disease. Liposuction.

RESUMO

A lipomatose simétrica múltipla é uma doença incomum, de etiologia desconhecida, caracterizada pelo acúmulo simétrico de depósitos de tecido adiposo em face, pescoço, tronco e região proximal dos membros. É mais frequente em indivíduos adultos do sexo masculino, geralmente com história de etilismo. O tratamento é cirúrgico, com ressecção paliativa por via aberta ou por lipoaspiração. A recidiva é comum. O presente artigo tem por objetivo relatar dois casos de lipomatose simétrica múltipla, tratados por lipoaspiração no Hospital Regional da Asa Norte, em Brasília-DF.

Descritores: Lipomatose Simétrica Múltipla. Doença de Madelung, Lipoaspiração.

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INTRODUCTION

Multiple symmetric lipomatosis, also known as benign symmetric lipomatosis, Madelung disease, or Launois-Bensaude syndrome, was first reported by Brodie in 1846. However, the first case

series was described in 1888 by Madelung and in 1898 and by Launois and Bensaude¹. Multiple symmetric lipomatosis is a rare disease that usually affects adult men with a history of chronic alcohol use¹⁻⁶. It is characterized by symmetrical appearance of uncapsulated lipomatous masses

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in the face, neck, trunk, and proximal limbs, characteristically sparing the distal portions⁴. The disease is of great importance to plastic surgeons, as it directly affects body contour, with liposuction as the ideal treatment.

CASE REPORTS

Case 1

A 61-year-old, married, retired, and alcoholic man who consumed 350 mL of spirits per day since 17 years of age and was a former smoker complained of early fatty deposits, about 17 years prior, in the thorax, abdomen, and arms with progressive growth (Figure 1). His personal history revealed two operations, one for peptic ulcer 17 years ago and the other was cholecystectomy 2 years ago. He underwent liposuction in two sessions. In the first session, the posterior regions of the trunk were lipoaspirated. In the second session, 9 months later, the anterior trunk and arms were lipoaspirated. The first operation was uneventful, but the second procedure was complicated by seroma and secondary infection that developed into skin necrosis in the abdominal wall (Figure 2).



Figure 1 - Preoperative photograph .



Figure 2 - Case 1: Skin necrosis in the abdominal wall after liposuction .

Surgical drains were not used in either of the procedures. The complication was successfully treated

with drainage of collections, systemic antibiotics, serial debridement, and delayed primary closure after control of the infection and necrosis .

Case 2

A 35-year-old, married, alcoholic man who was a mason by profession consumed 500 mL of spirits per day and smoked 5 cigarettes per day for 20 years reported early fatty deposits 3 years ago, first in the chest and later in the abdomen and proximal portion of the lower limbs, also with a gradual increase in volume (Figure 3). He underwent liposuction in a single surgical procedure, without complications. Surgical drainage tubes were left in place, under suction, in the lipoaspirated areas.



Figure 3 - Preoperative photograph .

DISCUSSION

Multiple symmetric lipomatosis is a rare disease that usually affects the male sex (approximate male-to-female ratio, 15:1) between the third and sixth decades of life, and is associated with chronic alcohol use in up to 90% of cases¹⁻³. Both of the patients presented in this report present an epidemiological profile in agreement with the literature, including long-term heavy alcohol use.

The etiology of multiple symmetric lipomatosis is unknown¹⁻⁶. However, deficiency in embryonic brown adipose tissue, mitochondrial dysfunction, and defects in the catecholamine-induced lipolytic pathway have been suggested, with alcoholism apparently acting as a cofactor⁵.

The main clinical manifestation of the disease is the appearance of uncapsulated lipom-

atous deposits with symmetrical distribution in the face, neck, trunk, and proximal limbs, characteristically sparing the distal portions⁴. Typical locations include the cervical region ("horse collar"); posterior neck ("buffalo hump"); parotids: submentovertebral and retroauricular ("hamster cheek"); and on the chest, deltoids, and supraclavicular and proximal upper limbs ("pseudoathletic form")². Lipomas in most cases are asymptomatic but may cause difficulty in head movement and affect mediastinal structures such as vessels, nerves, the trachea, bronchi, the pharynx, and the esophagus. Furthermore, they may cause symptoms such as dyspnea, dysphagia, dysphonia, and obstructive sleep apnea^{1,2,4-6}.

Association with metabolic disorders (hyperuricemia, diabetes mellitus, and dyslipidemia), hypertension, liver disease, hypothyroidism, and renal tubular acidosis has also been reported^{1,2,4-6}.

A peculiar aspect of multiple symmetric lipomatosis is a particular form of peripheral neuropathy, which can be motor, sensory, or autonomic, unrelated to alcohol intake^{1,2,4-6}. This form of neuropathy affects up to 90% of patients over the course of the disease and may manifest as paresthesias, cramps, resting tachycardia, postural hypotension, segmental hyperhidrosis, erectile dysfunction, and acrocyanosis, among others⁴.

The clinical course is traditionally reported as benign. The medical literature reports only one case of malignant degeneration to sarcoma^{1,4-6}. However, multiple symmetric lipomatosis presents specific and significant morbidity and mortality, regardless of the benign nature of the deposits⁴. In a long-term longitudinal study, 31 patients with a mean follow-up period of 14.5 years had significantly higher mortality than the general population. Cases of sudden death attributed presumably to a cardiovascular manifestation of autonomic neuropathy were responsible for the inflation of the mortality rate. Moreover, a considerable morbidity was observed during the course of the disease due to the high frequency of mediastinal extension and peripheral neuropathy during long-term follow-up⁴.

In contrast to the literature, we did not observe any of the associated symptoms, the presence of metabolic disorders, or signs of peripheral neuropathy in any of the study patients.

Diagnosis is mainly clinical^{1,2,4} and based on the presence of multiple fatty deposits in a symmetrical distribution, which spare the distal portion of the limbs⁴, as observed in both patients. Computed tomography and magnetic nuclear resonance imaging, however, can be useful in determining the extent of lipomas and their relationship

with the surrounding structures, especially the mediastinum^{1,2}.

Palliative surgical excision, via the open method or liposuction, is the current treatment of choice¹⁻⁶. Lipomas are not encapsulated, have an infiltrative nature, have a fibrous component, and are extensively vascularized, usually preventing their complete removal². Moreover, the basic disorder, which is responsible for fatty deposits, persists, leading to frequent recurrences². Liposuction presents some advantages in that it is less invasive, prevents extensive scars, requires a shorter recovery period, and can be performed multiple times^{2,5}. However, it may present technical difficulties such as resistance to penetration of the cannula (due to the fibrous component of the lesions) and with profuse bleeding (due to the hypervascular nature of the disease)². One can choose, as was decided in case 1, to perform more than one session to minimize blood loss³. Removal of redundant skin may be required after completion of the liposuction³. Conventional surgical excision of lipomas provides a greater possibility of complete resection, despite being hampered by the infiltrative nature of lipomas. A cleavage plane with normal tissues is not easily observed⁵. Hematomas and seromas are common surgical complications^{1,2,5}. Rigorous hemostasis and use of drains are recommended^{1,2,5}.

In the two cases under discussion, we opted for liposuction to improve body contour, and both patients were greatly satisfied with the outcomes. The absence of drains and enhanced liposuction might have contributed to the complications observed in patient 1. After liposuction, the same patient presented with large cutaneous excess in the abdomen, back, and upper limbs but did not desire additional procedures to improve the results. None of the patients had evidence of recurrence until the sixth month of postoperative follow-up (Figures 4 and 5).



Figure 4 - Case 1: Photograph at 6 postoperative months



Figure 5 - Case 2: Photograph at 6 postoperative months .

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