

Benign symmetric lipomatosis*

*Lipomatose simétrica benigna**

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Abstract: Benign symmetric lipomatosis is a rare syndrome characterized by gradual appearance of encapsulated masses of adipose tissue, affecting the upper part of the trunk, neck, and limbs. For the peculiarity of its clinical features, this condition received comparative denominations from its original authors, which are classical and still appropriate. Based on a report of a typical case of the condition, some aspects of the syndrome are exemplified, as the 'horse-collar' and the 'pseudoathletic' appearance.

Keywords: Lipoma; Lipomatosis; Adipose tissue

Resumo: A lipomatose simétrica benigna é síndrome rara, caracterizada pelo acúmulo de massas de tecido adiposo no tronco, pescoço e nos membros que, pela peculiaridade de seus sinais clínicos, recebeu de seus primeiros observadores denominações comparativas que se tornaram clássicas e são até hoje utilizadas de maneira bastante apropriada. Com base no registro de um caso típico dessa condição, são demonstrados alguns desses aspectos, como o "colar de cavalo" e a aparência "pseudoatletica".

Palavras-chave: Lipoma; Lipomatose; Tecido adiposo

INTRODUCTION

From a historic perspective, clinical and laboratorial features of syndromes and diseases have received descriptions according to their similarities to manifestations from other sources. Two classic examples are the "horse collar" and the "pseudoathletic" appearance of benign symmetric lipomatosis. By reporting a typical case of this syndrome, we are able to demonstrate those manifestations through the iconographic record of a 57-year-old white man with a previous history of heavy alcohol consumption and bleeding by esophageal varix, with lipomatous masses on his trunk, neck and shoulders, and other related complications.

COMMENTS

Benign symmetric lipomatosis (BSL) is a rare disorder initially described in 1888 by the German surgeon Otto Wilhelm Madelung, who observed the occurrence of great symmetrical masses of adipose tissue around the neck and shoulders of brewery

workers, with a "horse collar" appearance¹ (Figure 1). Ten years later, Launois and Bensaude² reported 65 similar patients.



FIGURE 1: "Madelung's neck": abnormal deposit of lipomatous masses around the neck creating an aspect similar to a "horse collar"

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FIGURE 2: "Pseudoathletic appearance" due to fat tissue distribution on thorax, abdomen and proximal limbs

BSL occurs preferentially in non-obese white males, from 20 to 65 years of age, generally associated with high ethanol intake. In some patients, fat distribution provides features that resemble a well-developed physique from a professional bodybuilding practitioner, classically named pseudoathletic form of Launois-Bensaude syndrome (Figure 2). In female patients with BSL there is a rather specific morphological aspect, characterized by an obesity-like pattern and a low occurrence of the typical "Madelung's collar".³

The pathogenesis of BSL remains unclear, although fat deposits can be attributed to a neoplastic-like proliferation of functionally defective brown adipocytes.⁴ Another hypothesis is that alcohol could promote lipoma development due to alterations of



FIGURE 3: Dupuytren's contracture

the number and function of beta adrenergic receptors and due to lipolytic and lipogenic activities.

Despite being called benign, BSL can be associated with respiratory symptoms and compression signals due to the infiltration of adipose tissue at mediastinal level.⁵ In the case reported here neural compression by lipomas was responsible for loss of muscular strength, paresthesia, and Dupuytren's contracture (Figure 3).

Treatment options include lipectomy and liposuction, which can give good cosmetic results, although recurrences often occur.⁵ In addition to the surgical treatment, patients should be encouraged to follow a low fat diet, alcoholic abstinence, and to maintain physical activities. □

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