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Case Report

Benign Symmetric Lipomatosis (Launois-Bensaude syndrome) – A Rare Cause of Muscular Weakness

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Abstract
A 76-year-old female patient is presented who suffered from muscular weakness in arms and legs. She was obese and had a symmetric accumulation of fatty tissue with a bumpy structure at both arms which gave the patient a pseudoathletic appearance. Fatty tissue accumulations were present at both shoulders, arms, at both thighs, at the back and the abdomen. She suffered from benign symmetric lipomatosis (BSL), also called Launois-Bensaude syndrome (LBS), which is a rare disorder of unknown origin and poorly understood pathophysiology. It is believed to be a disease of disturbed lipogenesis induced by catecholamines. The syndrome is often associated with features of metabolic syndrome such as diabetes mellitus, hyperuricemia, hyperlipidemia and hypertension and is associated with polyneuropathy which is an integral part of the disease. Therapeutic options are pharmacological treatment with salbutamol and surgical procedures such as lipectomy or liposuction.

Key words: Muscular weakness, benign symmetric lipomatosis, Launois-Bensaude syndrome, neuropathy, lipogenesis

Abbreviations: BSL Benign symmetric lipomatosis, MSL Multiple symmetric lipomatosis, LBS Launois-Bensaude syndrome

Introduction

Benign symmetric lipomatosis (BSL) is a rare disorder which is characterized by symmetric accumulation of unencapsulated lipomata at the neck, the shoulder girdle, upper arms, thorax, abdomens, pelvic girdle and thighs giving the patients a chracteristic pseudoathletic appearance. The syndrome was first described by Brodie in 1846 [4]. Madelung reported 33 cases in 1888 [18]. The classical descripton of the disease, however, is that of Launois and Bensaude [15], who published a detailed account of 65 cases in 1898.

The disease is rare, only around 200 cases are described in the medical literature. It seems to be more frequent in countries around the Mediterranean Sea [10]. There it predominates in adult males with an incidence of 1 in 25,000 and a male-to-female ratio of 15:1 to 30:1. There may be regional differences. Ruzicka et al. [22] describe ten patients from whom half are female. The syndrome is described in adults from 30 to 60 years old. Familial occurance has been reported and an autosomal dominant mode of inheritance has been postulated [17, 20].

The majority of patients with Launois-Bensaude syndrome have features of a metabolic syndrome such as diabetes mellitus, hyperlipidemia and hyperuricemia. In addition, they suffer often from symptoms of peripheral motor and sensory neuropathy [8], which was attributed to accompaning chronic alcohol comsumption by several authors. The examination of sural nerve biospy specimen in patients with BSL [20], howewer, supported the view that chronic distal axonopathy is not due to alcohol consumption but is an intregral part of the disease. Patients complain of numbness, wasting and weakness of arms and legs, sometimes myalgias and arthralgias.

We report on a case of a female patient who presented with myalgias, lack of strength and paresthesias in arms and legs, which could be attributed to peripheral neuropathy as an integral part of the rare Launois-Bensaude syndrome.

Case Report

A 76-year old female patient was referred to rheumatology because of suspected polymyalgia rheumatica. She suffered from myalgias, lack of strength, paresthesias and numbness in arms and legs which had gradually developed within the past five years. She experienced difficulties in getting out of a chair, climbing stairs, walk without a walking stick. She had a history of metabolic syndrome with type 2 insulin-dependent diabetes mellitus since 1984, with hypertension, hypercholesterimia and hyperuricemia.

The physical examination revealed an obese stature. Her weight was 120 kilogram, her length 176 cm correspondidng to a BMI of 39 kg/m^2. Besides this general obesity (Fig. 1) the patient had a symmetrical accumulation of fatty tissue with a bumpy structure at both arms (Fig. 2) which gave the patient a pseudoathletic appearance. Fatty tissue accumulations resembling shoulder pads were present at both shoulders. Fatty tissue masses were also visible at both thighs, at the back and the abdomen.

Muscular weakness was found in arms and more severe in both legs. The tendon reflexes were present except at the ankles. She had a glove and stocking senso-

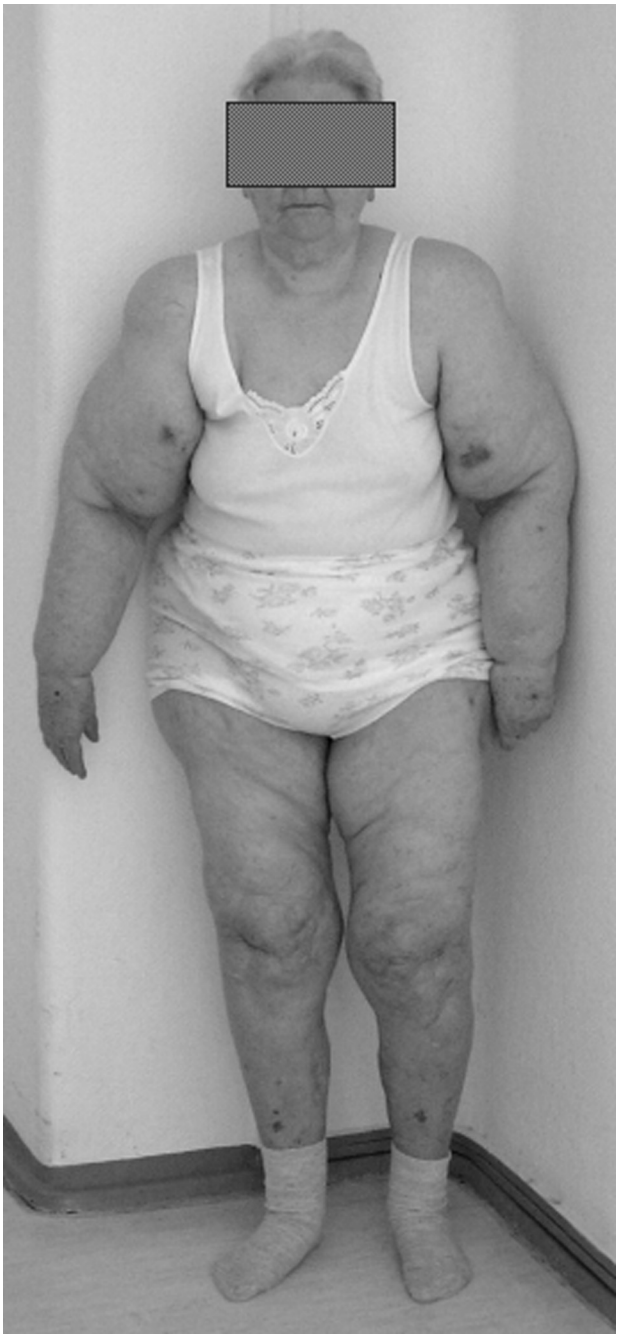

Fig. 1. View of the patient with hypertrophy of fat in the area of upper arms.

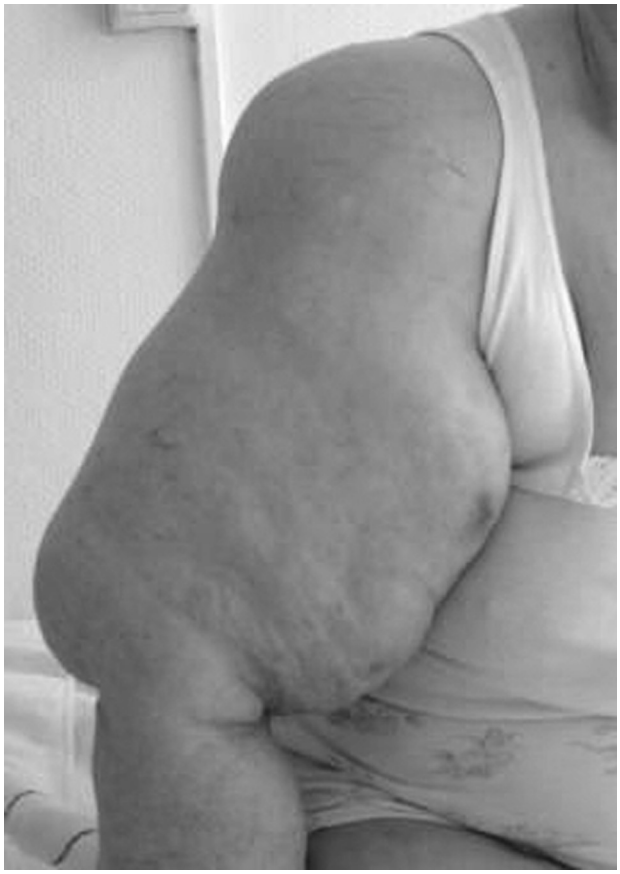

Fig. 2. Right upper arm of the patient with shoulder pad like accumulation of fat around the shoulders and the upper arm resembling puffed sleeves which gives the patient a pseudoathletic appearance. Note the bumpy structure of the skin caused by lipomatas.

ry impairment to pin prick, light touch and temperature shading off to normal at the elbows and at both knee midcalf. Because of distal leg weakness the patient was unable to walk without walking stick more than two or three steps. For longer distances she used a rolling rollator walker or a wheelchair.

Laboratory investigatons revealed a normal ESG, a normal CRP, CK and aldolase. Blood count, liver and kidney functions tests and immunological parameters such as antinuclear antibodies were also normal. Slightly elevated were cholesterin (5.46 mmol/l), uric acid (396 umol/l) and HbA1c (6.8 %). Fasting glucose was normal. Endocrine parameters such as thyrotropin, T3, T4, prolactin, estradiole, testosterone, androstenedione, dehydroepiandrosterone, luteinizing hormone, follicle-stimulating hormone, adrenocorticotropic hormone (ACTH) were normal. Cortisol was suppressed well in a dexamethason suppression test.

Ultrasonography of the abdomen revealed no abnormal finding, so did radiographs of the lung, the arms and legs. Electrodiagnostic studies demonstrated a reduction of motor and sensory conduction velocity of ulnar, tibial, peroneal and sural nerves. Needle electromyography demonstrated a decrease in motor conduction velocity and chronic denervation in the musculus tibialis anterior, rectus femoris and abductor digiti minimi. Biopsy of the fatty tissue revealed normal fat cells.

Based on the unique features of the disease a diagnosis of benign symmetric lipomatosis (Launois-Bensaude syndrome) was established. The weakness could be explained in part by the accompaning polyneuropathy. Other suspected diseases such as polymyalgia and endocrine disorders could be excluded.

Discussion

The patient suffered from benign symmetric lipomatosis (BSL), also called Launois-Bensaude syndrome (BSL). This rare syndrome or disease is characterized by symmetric fat deposits at different parts of the

Table 1. Types of benign symmetric lipomatosis (according to Donhauser et al. [6]).

Type I	Madelung´s disease: Lipomatas are located at the neck (fatty neck, buffula hump, horsecollar lipomata)
Type II	Pseudoathletic type: Lipomatas are located at the shoulder girdle, the upper arms, the thorax, the thighs and sometimes the abdomen and the back
Type III	Gynaecoid type: Lipomatas are located mainly at the pelvic girdle (hips and thighs)

body. Depending on its anatomical locations Donhauser et al. [6] divided BSL into three groups (Table 1). The presented patient has the most prominent fat accumulations on her shoulders and back and may therefore be classified as a type II BSL. Exact diagnostic criteria or classification criteria for this rare disease are not available.

BSL is a rare syndrome which is not very well understood. Patients are described in the literature unter different synonyms. The most frequently used synonyms are Launois-Bensaude syndrome (LBS), multiple symmetric lipomatosis (MSL), diffuse or generalized lipomatosis, lipomatosis simplex, symmetric adenolipomatosis and Madelung´s disease [25].

From a differential diagnostic aspect, BSL must be distinguished from encapsulated lipomas, diseases of the salivary gland, dietary or endocrine forms of obesity, lipodystrophy, Dercum disease, Fröhlich´syndrome and familial lipomatosis. Our patient has been checked for endocrine disorders without any pathological finding.

The aetiopathogenesis of the disease is poorly understood. Some evidence suggests that genetic abnormalities unterlie BSL and that some cases are associated with abnormal mitochondrial DNA and systemic mitochondrial dysfunction [14, 1]. In muscle biopsies, ragged red fibers could be demonstrated supporting the view of mitochondrial dysfunction. Mitochondrial DNA mutations in patients with BSL could be found. BSL fat deposits originate from defective noradrenergic modulation of proliferation and differentiation of brown fat cells [19] accumulating an excess of lipids. So it is considered to be cause by a disturbance of mitochondrial lipid metabolism and is considered a triglyceride storage disease [8]

There are reports of a drug-induced occurance of benign symmetric lipomatosis after being treated with protease inhibitors indinavir or lamivudine [13]. Proteinase inhibitors are known to influence sugar and lipid metabolism.

There are different degrees of severity possible. The disease may be mild [10] and hardly visible or cause severe disfigurement. In rare cases life threatening conditions such as compression of the trachea and thoracic veins requiring surgery are reported [2]. Two cases are reported in the literature, in which the process became malignant. Durand [7] described the occurance of liposarcoma and Tizian [24] the occurance of intramyxoid sarcoma in a patient with BSL.

Patients with BLS have often features of a metabolic syndrome with impaired glucose tolerance of overt diabetes mellitus, hypertension, hyperlipidemia and hyperuricemia. The disease is often associated with sequellae of alcohol abuse such as hepatopathy, peripheral neuropathy or macrocytic anemia.

Neuropathy has been reported in association with BSL [22, 8]. Because alcohol abuse is reported in many cases of patients with BSL, neuropathy was attributed to alcohol usage. Neuropathy has been described in patients who did not drink alcolhol [12]. Pollock et al. [21] and Enzi et al. [8, 9] however, worked out very well, that neuropathy in BSL is an integral part of the disease and not due to alcohol abuse. Pollok et al. [20] did sural nerve biopsies from patients with BSL. They revealed an absense of acute axonal degeneration, a significant shift to the left of myelinated fibre diameter distribtion, reduced indices of axonal and nerve fibre circularity, and an increase in myelin periodicity. This supports the view, that polyneuropathy in BSL is a chronic distal axonopathy and an integral part of the BLS syndrome. Although it is stressed in the literature that patients with BSL are often chronic alcoholics there are reports of patients who do not drink alcohol [12]. The patient presented here does not drink alcohol either.

At the present time point there is no causal therapy available. Dietetic interventions are not successful. There are reports which provide evidence that salbutamol 12 mg per day slows down the disease process [16]. Salbutamol acts on lipolysis via adrenergic stimulation. However, treatment has to be started early in the disease process to be effective. Often the BSL syndrome is recognized only in later stages of the disease.

Removal is the only successful treatment althougth relapses may occur. Surgical resection or lipectomy and liposuction are the two main procedures applied. There are numerous reports in the literature [23, 3, 11, review in 25 and 5] describing successfull surgical interventions especially if the large masses of lipoma cause functional impairment due to compession of important structures such as trachea, larynx or mediastinal organs and for cosmetic or psychologic reasons. Various authors regard liposuction as an effective procedure. This closed subcutaneous technique can be performed under local anesthesia. However, recurrence can only be avoided if exstirpation is radical. Liposuction is the method of choice if smaller lipomas need to be removed.

Our patient was presented to the Department of Plastic Surgery, where liposuction and lipectomy were considered possible forms of treatment. The patient, however, could not decide to undergo an invasive procedure.

Although it was not possible to offer the patient a causative treatment, a diagnosis was established, which may be preventive of futile dietetic intervention, discrimination because of supposed undisciplined eating and provides the patient an understanding of a condition causing physical disfigurement which was unexplicable before.

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