

# Rare case of multiple symmetric lipomatosis – Madelung’s disease

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## **Abstract:**

Multiple symmetric lipomatosis is a rare disease characterized by a symmetrical accumulation of massive adipose tissue on the neck, superior part of the trunk, and limbs<sup>1</sup>. We report an exclusively rare for our practice case of multiple symmetric lipomatosis in a 51-year-old male, who presented with diffuse lipomatosis – face and neck engagement, bilateral breast enlargement, upper parts of the arms and ankle involvement was seen too – typical engagement for Madelung’s disease. There was no serious deviations in his blood tests. No family history was found also. During gathering information alcohol abuse was discovered.

**Key words:** Breast enlargement, multiple symmetric lipomatosis, alcohol abuse, Madelung’s disease

## **Introduction**

Lipomatosis means local or generalized accumulation of fat tissue in the organism. We can distinguish encapsulated and nonencapsulated forms of the disease.

Generally there are two major types of multiple lipomatosis – familial and nonfamilial:

Familial multiple lipomatosis is a rare autosomal disorder characterized by numerous encapsulated lipomas on the trunk and extremities. The lipomas are usually not painful but can cause pain when growing. In rare cases, one lipoma can become painful and progress to multiple painful lipomas; it is then referred to as Dercum’s Disease Type III.

The nonfamilial benign symmetric lipomatosis also called Madelung’s disease is a rare disorder characterized by the presence of multiple, symmetric, nonencapsulated fat masses in the face, neck, and other areas. The disorder was first described by Brodie in 1846. After that, Madelung in 1888 and Launois and Bensaude in 1898 characterized the disease. Typically, this entity has been related to the presence of three anterior bulges in the neck.<sup>2</sup> It is more common in Mediterranean countries and is often associated with alcohol abuse. Benign symmetric lipomatosis is usually described in adults from 30 to 60 years old, with an incidence of about 1 in 25,000 and a male-to-female ratio of 15:1 to 30:1. Most cases have no hereditary pattern. More than 90% of the patients have associated alcoholism. The etiology of benign symmetric lipomatosis remains unknown, but an abnormal lipogenesis



induced by catecholamines has been observed. The transformation of BSL to a malignant tumor is extremely rare.

There are congenital forms of that illness also. The facial congenital lipomatosis is described first by Slavin and colleagues in 1989. It is a rare congenital disorder in which mature lipocytes invade adjacent tissue. The phenotypic features include soft-tissue and skeletal hypertrophy, premature dental eruption, and regional macrodontia. Due to its diffuse infiltration and involvement of important facial structures, complete surgical excision is often impossible. The aetiology, natural history, optimal management, and relationship to other disorders of fatty overgrowth are unclear<sup>3</sup>. Another congenital form of the disease is the Encephalocraniocutaneous lipomatosis (ECCL), also known as Fishman syndrome, it is a rare congenital neurocutaneous disease that commonly involves ectomesodermal tissues, such as eye, skin, and central nervous system<sup>5</sup>.

A rare cases of gastroduodenal lipomatosis, synovial lipomatosis, testicular lipomatosis, lipomatosis

of the ileocaecal valve can also be found in the literature.

No matter what exact type of lipomatosis is, all cases can only be treated by surgery – lipectomy, liposuction or lipectomy combined with liposuction.

## Case report

We present a clinical case of 51-year-old man who was referred to our division for the chief complaints of multiple painless soft masses emerging at his face, neck, breasts, upper and lower extremities. The symptoms started 20 days ago. There were no other complains. The patient showed no family history also.

Clinical exam revealed multiple, painless, subcutaneous lipomas situated symmetrically on his face (Pic №1), especially involving the parotid glands regions, oral cavity bottom and retro auricular spaces (Pic №2). There was total neck involvement (Pic №3). Mild gynecomastia was discovered also.



Picture №1



Picture №2



Picture №3

During the hospitalization period, proximal parts of the upper and lower extremities started to show involvement too. All affected areas were covered with intact tissue.

To put the diagnosis a several tests (blood tests, Computed Tomography of the chest, abdomen,

foots and ankles, MRA of the neck, ultrasound of the thyroid gland and kidneys) were performed.

Blood tests showed no deviations.

Chest CT – lungs, trachea and bronchi – normal images. Multiple enlarged lymph nodes in the mediastinum and axillas (Picture № 4).

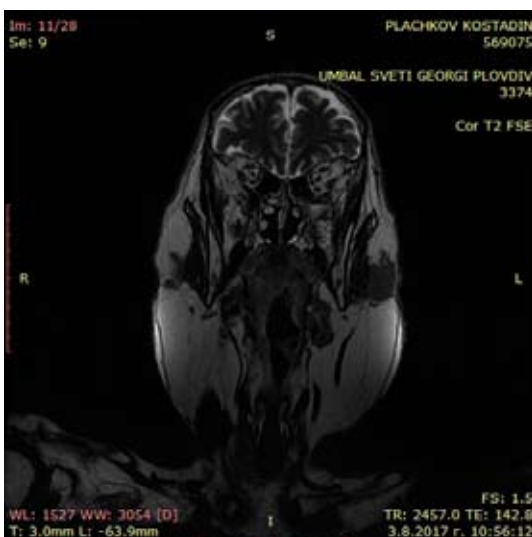
Abdomen CT – liver, gall-bladder, kidneys, bladder, prostate gland and adrenal glands without significant changes.

Feet and ankles CT – old fractures of the left ankle and tibia, as well as osteoporosis of the tarsal bones.



Picture №4

Neck soft tissues MRI – multiple soft tissue lesions, different in shape and size with MRI features of lipomas, engaging subcutaneous and deep adipose neck layers, including periglandular, perivascular spaces and round the muscles which are without adipose degeneration. The lesions present themselves with homogeneous structure and during their evolution they press but do not infiltrate the surrounding structures (Pictures №5-8).



Picture №5



Picture №6



Picture №7



Picture №8



Thyroid gland ultrasound – adipose subcutaneous tissue proliferation in the neck area over than 2 cm., thyroid gland normal (Picture №9).

Kidneys ultrasound – right kidney normal, left kidney with single micro concrements (Picture №10).



Picture №9 Neck ultrasound



Picture №10 Left kidney ultrasound image

Consultations with numerous specialists (specialist in internal diseases, rheumatologist, nephrologist, geneticist) were performed.

At the end, for a histological verification, surgical intervention was performed – partial excision of the neck soft tissue masses. The histology shows mature adipose tissue with thin capsule – lipomas/lipomatosis.

When all test results were analyzed, the diagnosis „*nonfamilial benign symmetric lipomatosis – Madelung’s disease*“ was set.

There was a discussion after all whether to treat the patient by surgery – lipectomy or liposuction, or to leave him without any treatment for a follow up period of a several months. There were no life-threatening symptoms of the disease, so we with his agreement naturally, decided to perform no action at that stage, but only to keep an eye over him.

## Discussion

Multiple symmetric lipomatosis(MSL), is a benign rare disease, which main feature is symmetrical accumulation of adipose tissue at different body parts – neck, the superior part of the trunk, limbs and others. There are multiple synonyms for this disorder, such as Madelung’s disease, Lauenois-Bensaude syndrome and benign symmetrical lipomatosis (BSL). Most cases have no hereditary pattern but according to the literature, more than 90% of the patients with this illness have active or past history of alcohol abuse.

In all cases if treatment is necessary it can only be done by surgery – lipectomy, liposuction or lipectomy combined with liposuction, but before the procedures to be started the patient has to be aware that there are a chance of recurrences and that chance is not very low (according to the literature findings 39% or more)

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