Rare case of multiple symmetric lipomatosis - Madelung's disease

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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. 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.

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 complaints. 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. 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 (Picture №5).

To put the diagnosis a several tests (blood tests, Computed Tomography of the chest, abdomen, feet and ankles, MRA of the neck, ultrasound of the thyroid gland and kidneys) were performed. Chest CT – lungs, trachea and bronchi - normal images. Multiple enlarged lymph nodes in the mediastinum and axillas (Picture №4). The histology shows mature adipose tissue with thin capsule – lipomas/lipomatosis.

After 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 ore liposuction, or to leave him without any treatment for a follow up period of 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 and conclusions: Multiple symmetric lipomatosis(MSL), Madelung’s disease is a benignant rare illness, which main feature is symmetrical accumulation of adipose tissue at different body parts. 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.

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 patent 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)

References: