CLINICAL CASE

THE SURGICAL MANAGEMENT OF CERVICAL LIPOMATOSIS – CASE REPORT

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Abstract

Madelung's disease (MD), also known as multiple symmetric lipomatosis or Launois Bensaude syndrome, is a rare metabolic condition, consisting of fat deposits, located mainly in the cervical, thoracic and abdominal areas. Aetiology of the MD still remains unclear. It affects about one person in 25.000 inhabitants, with no gender distinction. Usually, patients with MD also suffer from other coexisting conditions. For the majority of the patients, most frequent reasons for medical presentation are aesthetic reasons. The unique efficient treatment consists of surgical approach. Considering the rarity of such clinical cases and the lack of operative approaches in the medical literature, the aim of our paper is to present a case of MD of a non smoker and non alcoholic patient and our postoperative results. We report the clinical case of a middle aged male (41 years old), which required the surgical management of his cervical lipomatosis, in order to regain his initial aesthetic aspect. Our surgical result was satisfying both for our surgical team and for the patient. We managed to obtain a positive aesthetic result, with no postoperative complications or negative events.

Keywords: cervical, lipomatosis, Madelung's Disease

Introduction

Madelung's disease (MD), also known as multiple symmetric lipomatosis or Launois Bensaude syndrome, is a disfiguring and rare lipid metabolic condition, consisting of non-encapsulated, benign, symmetric and painless fat deposits, located mainly in the cervical, thoracic and abdominal areas. Aetiology of the MD remains unclear, but there are several theories that suggest the involvement of chronic alcohol consumption in the adipocyte hyperplasia for

genetically susceptible individuals. It affects patients aged between 30-70 years old, about one person in 25.000 inhabitants, with gender distinction (male-female ratio of 15:1).

An enzymatic defect- cytochrome c oxidase or alteration of the adipocyte cell surface could prevent physiological lipid metabolism, which determines the signs and symptoms characteristic of the disease [1]. Some recent data suggest that this impairment of lipid metabolism may also be related to improper activity in the brown fat mitochondria [2].

The genetic factors play an essential role in Madelung's disease. In general, many family members are affected by this condition, although the mode of determination has not been fully established.[3]. It is assumed that mutations in the mitochondrial DNA, mutations expressed in the MT-TK gene.[4],[5]

Usually, patients with MD also suffer from other coexisting conditions, such as metabolic syndrome, dyslipidaemia, hypothyroidism, diabetes, hypertension, epilepsy or alcohol induced cirrhosis.[6],[7] Untreated or mistreated MD tends to grow until it could lead to possible complications, like cervical compressions, peripheral neuropathy, causing hoarseness, dysphagia, odynophagia, or even venous compression or mediastinal syndrome. On the other hand, regression is improbable. Malignant degeneration of MD is very rare described in literature.

For the majority of the patients, most frequent reasons for medical presentation involve aesthetic reasons, such as aesthetical embarrassment. The unique efficient treatment consists of surgical approach. Stopping alcohol consumption, weight loss, and correcting lipid metabolism can complete surgery.

Case presentation

Our case consists of one 41 year old non smoker and non alcoholic male, who sought medical attention due to a painless, symmetrical and bilateral horse collar growing mass of the cervical region and impairment of neck mobility. According to the patient, the mass started growing up progressively 2 years ago. Our patient wasn't suffering from dysphagia, dyspnea or other comorbidities or from genetical susceptibility. He works as a stoker at a local school. Laboratory tests were also within normal limits. The physical examination unveils a soft, mobile growing mass of the entire cervical region (horse collar + buffalo hump) and of the parotid region (hamster cheeks) with an irregular contour. According to Enzi's G. classification of MD (1984), we can include our patient's case to MD type 1. The computed tomography imaging study revealed: "excess of fat in the subcutaneous cervical and supraclavicular region, in the neck posteriorly and in the submental and

submandibular space anteriorly. This investigation is always useful in such cases because it can evaluate the extension of the lipomatous mass, the compression of the trachea, the malignant transformation, the vascularization of the adipose tissue deposition. We hospitalized the patient and he received a preoperative treatment for a few days with antibiotics (Cefuroxime), H2 blockers (Arnetine), NSAIDs.

Figure 1 – Non encapsulated adipose tissue

Figure 2 - Eleventh postoperative day

The patient underwent surgery for cosmetic reasons, in order to regain his self confidence. The surgery consisted of a wide cervicotomy with inspection and radical excision of the lipomatous mass and cervical neck lymph node dissection (Figure 1). Because of the rich vascularization of the lipomatous mass, haemostasis during the operation was quite difficult to perform. At the end of the operation, we placed a drainage tube at the patient's neck.

Postoperatory, our patient received a treatment with hydrocortisone hemisuccinate, NSAIDs and also with other antibiotics, such Metronidazole and Gentamicin. The postoperative care of the cervicotomy was performed daily with betadine and alcohol. Eleven days after the surgery, the drainage tube was removed and the patient was discharged from hospital in the twelfth postoperative day. The postoperative period was uneventful (Figure 2). Histopathological results after the excision biopsy of the lipomatous mass (Figure 3) were communicated after 21 days: adipous tissue of normal histological aspect.

Figure 3 - Resection specimen with massive enlarged adipocytes

Conclusions

MD is a disfiguring condition that results from symmetrical diffuse adipose tissue cumulation of the cervical area. Aetiology of the MD still remains unclear, but there are some theories suggested, such as mitochondrial disorder of fat brown tissue or a defect in the adrenergic stimulated lipolysis. The diagnostic is based mainly by the clinical examination of the patient and facilitated by the computed tomography. An important aspect of the MD is represented by the absence of a capsule and the absence of a strict localization. Concerning the differential diagnosis, MD must be differentiated from

obesity, Cushing's syndrome, neurofibromatosis, lymphoma, encapsulated fibrosis, angiolipomatosis. The unique efficient treatment for the avoidance of the aforementioned complications remains the surgical approach: we used cervicotomy-lipectomy for our paper. In the end, the patient was satisfied with his aesthetic result. Unfortunately, the medical literature mentions a high recurrence rate of approximately 60%. In our case, we will try to monitorise the eventual recurrence of the lipomatous masses by following the patient up two times a year.

Ethics: The patient signed the informed consent and approved the publication of this paper.

Acknowledgement: all authors have contributed equally to this paper.

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