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Case Report

Benign Symmetrical Lipomatosis (Bsl) Of The Head And Neck: Clinical Case Presentation

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Abstract:
Benign symmetrical lipomatosis (BSL) is a rare pathological finding with an alcohol connection. It predominantly affects males in the age between 30 and 60-year-old. Once developed, the disease does not undergo regression. The fatty accumulations have constant and progressive overgrowth that may significantly affect breathing, eating and speech. The most effective method for treatment is the surgical removal of the tumor masses. In the following we present a clinical case report of a 44-year-old male with BSL.

Keywords: Madelung's disease, benign symmetrical lipomatosis, fat tissue accumulations.

Introduction

Benign symmetrical lipomatosis (BSL), also known as Madelung's disease, is an uncommon pathological condition. The disease is characterized by symmetrical overgrowth of non-capsuled adipose tissue around the neck, shoulders, nape, it seldom affects the extremities, the sacral and abdominal regions. It strikes mostly males with (M: F ratio 15:1)[1,2].

BSL is first described in 1846 by Brodie [3]. In 1888, Otto Madelung [4] reported 35 cases of patients with a cervical lipomatosis and gave the name to the disease. At the end of the 19th century Launois and Bensaude[5] describe the main symptoms of BSL- multiple, symmetrical lipomatous masses around the head, neck and shoulders and clarified the typical clinical presentation of BSL.

BSL is a rare condition with the highest incidence reported in the Mediterranean population (1:25 000) [6]. It does not affect people under the age of 20, almost all of the patients are between 30 and 60 years old [7]. The etiology still remains unclear. The most common hypothesis is that there is the strong connection between this condition and the alcohol consumption. The liver dysfunction found in many patients with BSL confirms this hypothesis [8].

There are two types of BSL, depending on the clinical appearance: type 1: the fatty collection is well circumscribed and are distributed on the upper part of the body mostly around the nape, supraclavicular and deltoid regions, and type 2: the lipomatous masses are widespread over the whole body including the extremities and give an appearance of simple obesity. Malignant degeneration of the fatty tissues is extremely rare [9].

Clinical Case

A 44-year-old male patient, diagnosed three years ago with BSL was admitted to our clinic of oral and maxillofacial surgery in May, 2016, complaining of an aesthetic discomfort, functional disturbance and head movement's limitations. The patient reported a rapid growth of the fatty accumulations within the last three months. There was a history of alcohol consumption. On the clinical examination, there were multiple, symmetrical masses, situated in the cervical, submandibular and submental regions. A double chin was observed. On palpation the tumors were soft and fluctuant, with no definite margins. No tenderness or inflammation were observed. A similar collection of 20/20 mm subcutaneous fat is established in the left nape area. Magnetic resonance tomography and CT scan performed, confirmed the clinical diagnosis of BSL. There were no pathological changes in the area of the soft tissues and muscles of the facial skull. The salivary glands were with normal structure. There are no pathological changes in the pharynx and larynx. No significantly enlarged regional lymph nodes and no pathological changes in the mediastinum were found on the imaging methods of examination. All the laboratory tests were in normal ranges. **A)**

B)

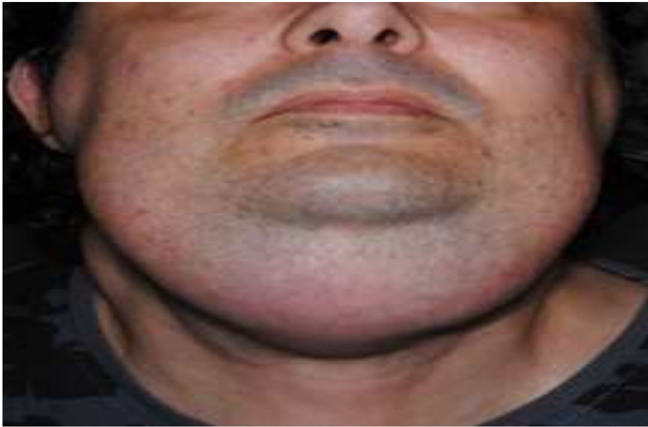

Figure 1 (A- front view Б- side view) : the tumor mass affecting the submandibular and submental spaces

Discussion

BSL is a rare disease. Despite its low frequency, the disease is well known by oral and maxilla-facial surgeons because of its specific symmetrical, non- capsuled fatty enlargements around the head and neck. The differential diagnosis includes angiolipoma, neurofibroma, hybernoma, congenital infiltrative lipomatosis of the face, liposarcoma, lipoblastomatosis, salivary glands disease and etc. It has been reported only one case for malignant degeneration of BSL into mixoid liposarcoma [9]. Chan et al. share the hypothesis upper respiratory tract malignancies in patient with BSL are as a result of the synergic action of smoking and alcohol, which have influence to both the diseases [10]. The most cases confirm the strong connenction between BSL and alcohol consumption in males. The antilipolytic and lipogenic effects of the alcohol disturb the local cateholamine- induced lipolysis [11]. BSL can affect female non- drinker patients too [1]. After a rapid overgrowth during 2 years of the disease, the fatty accumulation can remain relatively constan from then on, eventhough some patients report subjective changes of the tumor sizes. In our case report we did not observe such way of the disease progression. The growth of the tumor-masses was rather slow and constantly progressive, than variable. In many clinical cases, together with all the other clinical symptoms, a polyneuropathy is usually found. It basically starts a several years after the fat-tissue proliferation. In the cases of BSL, polyneuropathy is most commonly described as a sensor, motor and autonomic dysfunction [7].

The treatment recommendations of BSL include surgery and liposuction, and the operative excision of the lipomatous masses remains a preferable method of treatment [1]. In our clinical case the multiple, non- capsuled lipomatous masses were situated in the submandibular and sublingual regions, affecting both the submandibular and sublingual salivary glands. The fatty accumulations were diffusely spreading between the cervical fascias and the submandibular glands. (fig.2, fig.3). After cautios radical excision of the fatty acuumulations the glandulary capsules remained intact. Hemostasis was performed. Drains were placed and the wound got sewed up. The surgical treatment of BSL remains the most- effective technique in patients with BSL [12]. Liposuction is also a choice of treatment, although it is widely accepted that liposuction should be adjuvant to the main surgical therapy after the removal of the main tumor masses [13]. Medical therapy with β2- adrenergic stimulation has also been described in the literature, the main aim of which is to improve the local lipolysis disturbances. Leung et al report an improved health status in their patients after using salbutamol as a main treatment method [14]. Health recommendations such as weight loose, alcohol withdrawal, nutritional habit's improvement are also given to the patient.

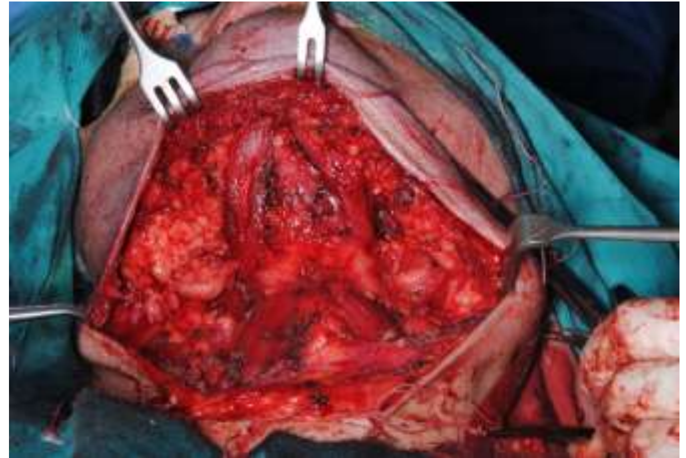

Figure 2. After the incision of the main tumor mass- multiple, non-capsuled, poor defined borders, spreading between the submandibular glands, muscles and soft tissues beneath.

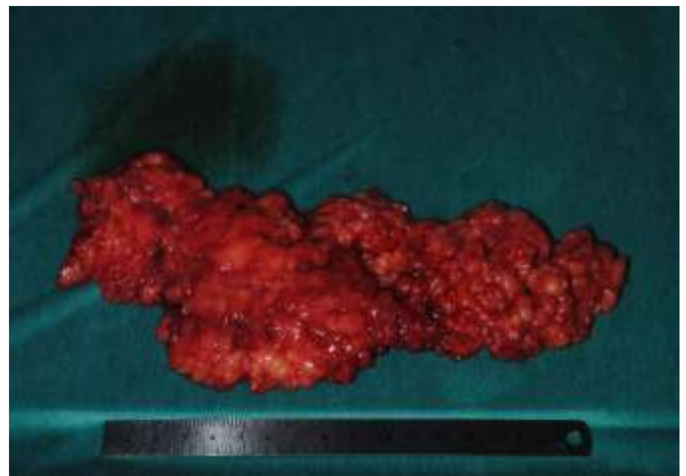

Figure 3. Half of the lipomatous enlargement after the excision.

Conclusion

BSL is a rare, progressive disease with a uniform clinical appearance. It affects predominantly males with Mediterranean origin. The etiology still remains unclear, but there are convincing arguments about the strong connection for the alcohol as a causative factor. Giving up the alcohol consumption and nutrition improvement do not lead to regression of the disease. The head and neck are the main places affected by the disease. Therefore, rogressively enlargement of the tumor masses could lead to disturbances in the breathing, eating and speaking. The operative techniques remain the most effective methods of treatment. The way of growth as well as the absence of a capsule are the main factors

associated with the recurrence of the disease.

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