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CASE REPORT

A Case Report of Multiple Symmetric Lipomatosis (MSL) in an Ethiopian Alcoholic Male

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Abstract

Background: Madelung disease or benign symmetric lipomatosis is an uncommon disorder thatis defined by the presence of symmetrical fatty deposits, which usually involve the upper trunk, neck and head

Case presentation: A 52-year-old male presented with neck masses, which had been growing progressively over a period of two years. His BMI is = 25.64. On examination, there was swelling over the neck and upper trunk.

Conclusions: we reported a black man having typical risk factor.

Keywords: Madelung disease; Lipomatosis; alcoholic; Ethiopian

Introduction

Madelung disease, or benign symmetric lipomatosis, was first described in1846 .it is an uncommon disorder thatis defined by the presence of symmetrical fatty deposits, which usually involve the upper trunk, neck and head (1). This disease predominant affects men of Mediterranean or Eastern European descent, between the ages of 30 and 60 years, with a history of alcohol abuse. [1, 2]

Although the exact pathogenesis of MD is not clear yet, around 90 % of patients with MD had a history of chronic alcohol use. The disease is more common in a middle-aged man of Mediterranean or eastern European descent. Herein, we present a case of 65 years-old Ethiopian male patient, without alcoholism history, presented with a complaint of multiple joint and back pain and progressive fat deposition in his trunk and proximal limb which was misdiagnosed as being obese in the past. [3]

Case Report

A 52-year-old male presented with bilaterally anterior and posterior neck masses, which had been growing progressively over a period of two years. The patient denied any drug use, and his past medical history was unremarkable. Over a period of 20 years, the patient was a heavy alcoholic. On admission, his height was 165cm, and his weight was 69.8 kg (BMI = 25.64, indicating obesity). At the physical examination, regions of localized swelling were observed on the neck and upper trunk, with no sign of systemic edema (Figure 1). There was no tenderness, or palpable mass, in the chest and abdominal areas. The patient had no other neurologic abnormality. His admission laboratory findings were normal white blood cell counts (WBC) 9.7 103/uL (normal; 4.0-11.0 103/uL), normal aminotransferase (AST) 23 IU/L (normal; 10-40 IU/L) and alanine transferase (ALT) 31 IU/L (normal; 6-40 IU/L); other laboratory data were unremarkable. The overall appearance of the patient revealed large symmetrical, protruding mass lesions at the anterior aspects of the bilateral supraclavicular areas and posterior aspect of the neck (Figure 1).

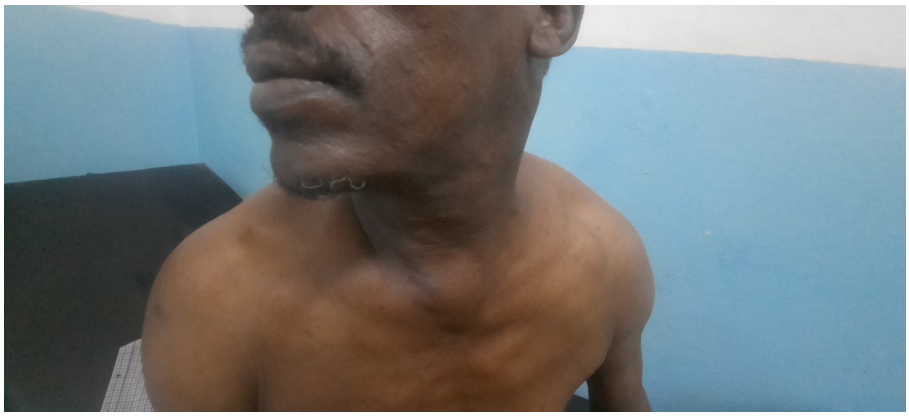

Figure 1: large symmetrical, protruding mass lesions at the anterior aspects of the bilateral supraclavicular areas and posterior aspect of the neck

On ultrasound there is posterior neck and anterior neck supraclavicular region diffuse expanding subcutaneous fat layers extending to the superior mediastinum (Figure 2)

CT scans of the neck and upper chest showed diffuse hypointense lesions, located in the subcutaneous fat layer of mainly the bilaterally supraclavicular areas. These lesions were is intense, with adjacent normal fat tissue, and not separated by a normal subcutaneous fat layer. No solid portions were detected, and there were only normal vascular structures traversing within the lesion and the superficial fasciae. (Figure 3)

Because of no symptoms is appreciated the patient except the cosmotic issue surgical excision of the masses at both neck areas was deferred by the patient after consulting him.

Microscopically, the lesions were composed of normal adipose tissue, without malignant changes and atypical spindle cells, and were diagnosed as lipomas. The diagnosis of Madelung's disease was made, based on the combination of clinical, CT and MRI features.

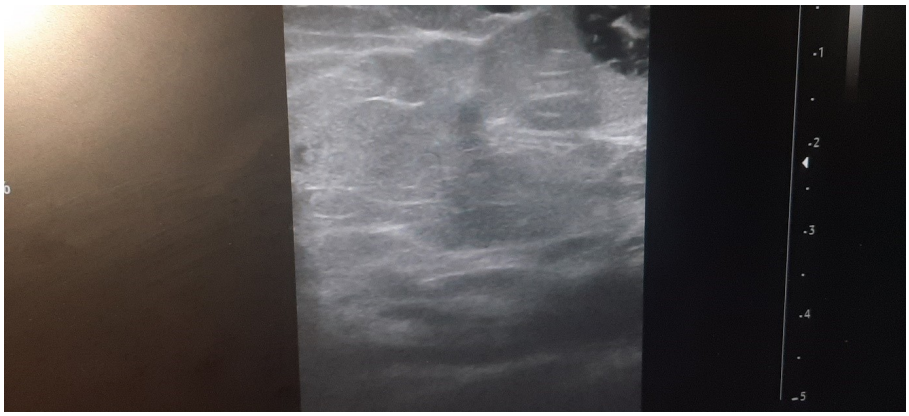

Figure 2: On ultrasound there is posterior neck and anterior neck supraclavicular region diffuse expanding subcutaneous fat layers extending to the superior mediastinum

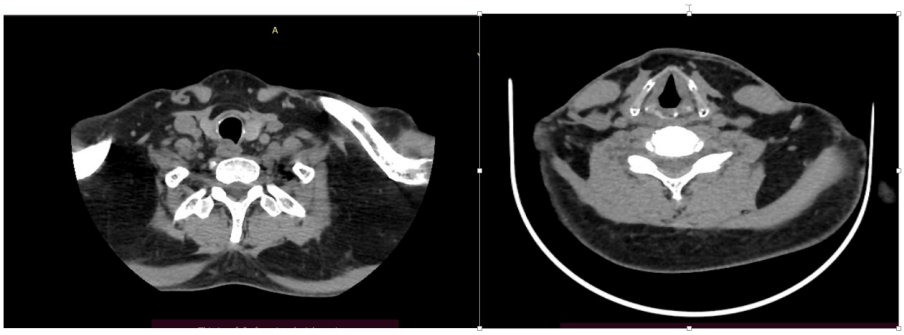

Figure 3: CT scans of the neck and upper chest showed diffuse hypointense lesions, located in the subcutaneous fat layer of mainly the bilaterally supraclavicular areas.

Discussion

Madelung disease (MD) was first described by Benjamin Brodie in 1846 in London hospital and subsequently named by Otto Madelung in 1888. Later in 1898, Launois and Bensaude, characterized the distinct features of the disease and termed benign symmetrical lipomatosis/Launois-Bensaude syndrome .Historically, Madelung disease disease has been mainly encountered in men (male-to-female ratio = 15:1)between 30 and 60 years of age, with the disease being more prevalent among men of Mediterranean or Eastern European descent Although its pathophysiology remains unclear, it is believed that Madelung disease is related to excessive alcohol consumption, particularly that of red wine. However, despite having a history of excessive alcohol consumption, our patient had no history specifically related to red wine consumption. [1]

Based on clinical presentation, there were two types of MD. Type I presents with tumor-like masses around the neck and back, giving the patient a "buffalo hump" and type II MD resulting in "pseudo-athletic appearance" with diffuse fat deposition over the trunk and proximal part of limbs . Our case was diagnosed with type I MD based on the areas involved. Systemic comorbidities such as epilepsy, diabetes, Cushing's disease, primary hypothyroidism, macrocytic anemia, peripheral neuropathy, and some malignancies have been associated with MD. Despite extensive workup none of these conditions identified in our patient. [1, 3]

Diagnosis of MSL is clinical, based on symmetrical distribution of fatty mass. Imaging may assist diagnosis. Chest radiographs may show abnormal symmetrical fatty deposits. Ultrasound and CT may evaluate disease extent however MRI is the best diagnostic tool for evaluating the spread of adipose tissue, presence of tracheal compression and vascular topography within

the fat mass. Biopsy may help to confirm the diagnosis and exclude its mimickers such as angiolipoma, neurofibroma, lipodystrophy, and liposarcoma [1-4]

There is no definite treatment for MD but nonsurgical and surgical options were used in the real world with variable success rate. Clinical management of Madelung disease involves abstinence from alcohol; however, there is only a slight regression in the magnitude of lipomatous deposits surgical Treatment is usually performed for cosmetic reasons or to alleviate symptoms such as dyspnea or dysphagia and it is the only effective treatment. [2, 3, 5]

In conclusion, to the best of our knowledge, this is the first case of Madelung disease reported from Ethiopia and possibly in Africa. Clinicians need to consider MD in their differential diagnosis when encountered with a patient with a slowing growing body fat with a centripetal distribution that can be complicated by metabolic and neurologic conditions. As early diagnosis and timely intervention might prevent serious complications and improves patient quality of life.

References

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