CASE REPORT

MADELUNG'S DISEASE: REPORT OF A CASE

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ABSTRACT

A 52 year old man presented with multiple bilateral symmetric swellings on the neck, back and axilla. A fine needle aspiration done from multiple sites showed that the swellings were composed of mature regular fat cells. The swellings were surgically excised and a histopathologic study revealed an unencapsulated mature fat tissue which confirmed a diagnosis of benign symmetric lipomatosis (BSL) or Madelung's disease. This is a rare disease characterized by the presence of multiple, symmetric, non-encapsulated fat masses in the face, neck and other areas. It commonly occurs in middle aged men of Mediterranean origin but is very rare among Africans. Further history and review of literature with respect to the pathology, clinical presentation and management options are discussed.
Key words: Madelung's disease, Benign symmetric lipomatosis

INTRODUCTION

Madelung's disease or benign symmetric lipomatosis is a very rare disease characterized by the deposition of non-encapsulated fat in the neck, the superior part of the trunk and rarely in the limbs. Since its original description in 1846 by Brodie, about 200 cases have been reported in English literature (1, 2).

Unlike conventional lipoma, these benign fatty masses are not encapsulated within a membraneous capsule with very distinct boundaries.

It commonly occurs in middle-aged males from the Mediterranean European countries and is rare in other regions. It is strongly associated with alcohol abuse and other associated conditions including diabetes mellitus, hyperlipidemia, liver disease, hypothyroidism and polyneuropathy of unknown origin (3).

The commonest complaint of patients is related to cosmetic reasons and rarely compressive symptoms. We report on a case of Madelung's disease in a 52-year old farmer with a history of alcoholism who presented with bilateral symmetrical masses on the neck, axilla and back.

CASE REPORT

A 52-years old farmer from Debark, North-West Ethiopia, was admitted to the Surgical Ward of Gondar University Hospital after presenting with bilateral multiple swellings in the neck, back and axilla of a duration if two years.

The swelling was first noted in the right neck, then progressed to involve the left neck, upper back and axilla bilaterally .He had no history of breathing difficulty, snoring dysphagia or dysphonia.

There was no family history of similar illness. The patient was worried about his appearance; otherwise the swellings were not associated with any pain or discomfort. He has been a heavy drinker of alcohol for 30 years but he doesn't smoke cigarette.

On physical examination, all vital signs were within the normal range; blood pressure 120/70,pulse rate 80 regular, temperature 36.8^0C.There were symmetric and diffuse swellings on both sides of the neck extending medially and measuring about 10 x 10 cm (Fig 1) . Similar bilateral symmetric swellings were present on the upper back and axilla (Fig 2).

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Fig1. *Symmetrical subcutaneous masses involving the anterior cervical region*

Fig 2. *Symmetric upper back fat deposits*

The swellings were soft and non tender. There was no clinical evidence of polyneuropathy although investigations like nerve conduction tests were not done. No abnormality was detected in the examination of the other systems.

Many of the laboratory data were within the normal range; no abnormalities in the blood glucose level, liver and renal function tests except a marginal elevation of uric acid and triglyceride levels (See table 1).

Table 1: Laboratory data

	Patient's data	Normalvalues
CBC:		
WBC count	6800/ml	4000-11000/ml
Neutrophils	56%	
Lymphocytes	35%	
Eosinophils	9%	
Hematocrit	38%	
Urine:		
Ph	6	
Prot	(-)	
Glu	(-)	
Ket	(-)	
Total cholesterol	187 mg/dl	120-250mg/dl
LDL- cholesterol	104 mg/dl	<130mg/dl
Triglycerides	249 mg/dl	54-110mg/dl
Fasting blood glucose	79mg/dl	65-110mg/dl
Blood urea Nitrogen	10.6 mg/dl	7-21mg/dl
Creatinine	0.5 mg/dl	0.8-1.3mg/dl
Uric acid	8.89 mg/dl	2.1-8.5mg/dl
Total bilirubin	0.45 mg/dl	0.1-1.3mg/dl
Bilirubin(Direct)	0.1 mg/dl	<0.4mg/dl
SGOT	22.1 IU/l	8-80IU/l
SGPT	20.7 IU/l	7-56IU/l
Alkaline Phosphatase	88.3 IU/l	50-160IU/l

Ultrasound showed fatty infiltration of the liver. Fine needle aspiration was done from multiple sites and showed clusters of mature regular fat cells consistent with a benign fat growth.

With a diagnosis of benign symmetric lipomatosis, the patient was operated on and unencapsulated fat masses were excised with some difficulty because of infiltration to surrounding tissues (Fig 4).

Histopathologic study was done from the excised tissue to rule out malignant degeneration and showed lobulated mature fat tissue with delicate fibrous septae which confirmed the diagnosis of benign symmetric lipomatosis(Fig 3).

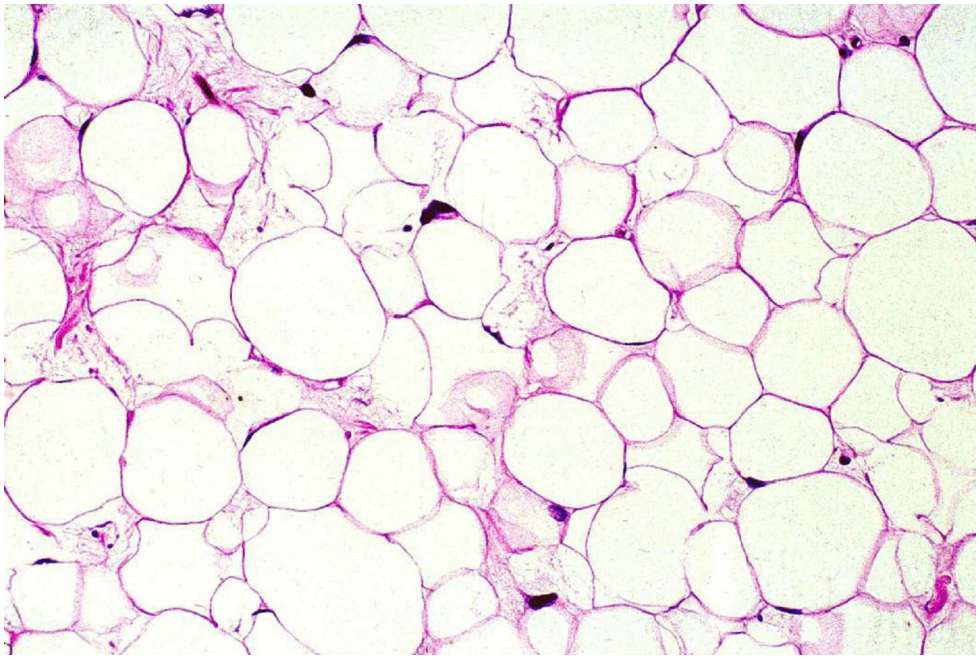

Fig 4: Histologic picture showing normal mature adipose issue (H&E, 400X)

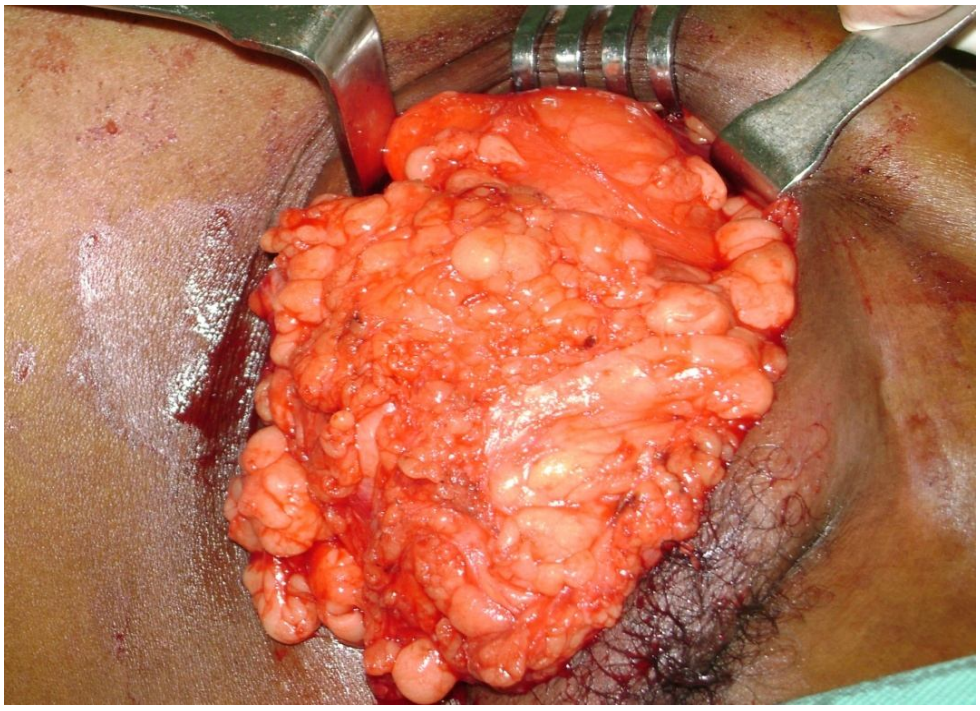

Fig 3: Surgical approach of the fatty mass in the axilla

DISCUSSION

Benign symmetric lipomatosis (BSL) is a rare disease defined by the presence of multiple ,symmetric ,nonencapsulated fat masses in the face, neck and other areas. After the original description by Brodie, it was further characterized by Madelung in 1888 and Launois and Bensuaude in 1898 (4, 5). BSL has been recognized by different names including Madelung's disease, lipomatose circonscrite multiple, symmetric adenolipomatosis, lipomatous simplex indolens, and Launois-Bensaude disease (6).

BSL predominantly affects alcoholic Mediterranean men with reported incidence of 1/25,000 among the Italian population; however, it is very rare among Asians and Africans (6). The disease is usually described in middle aged men from 30-60years with a male-to-female ratio of 15:1 to 30:1 (7)

Two clinical types are recognized. Type I primarily affects men and is characterized by fat tissue accumulation around the neck, nape of the neck, upper back, shoulder and the arms. Type 2 affects both men and women resulting in an exaggerated female fat tissue distribution in the upper back, deltoid region, upper arms, hips and the upper the thigh region (8). Our case has Type 2 clinical phenotype.

The etiology of the disease remains unknown but it is strongly associated with alcohol consumption with 60% to 90% of the patients having history of alcoholism although there are definite reports in patients without a history of alcoholism (7, 9). Other associated metabolic conditions include hyperuricemia, gout, liver disease, polyneuropathy, and diabetes and glucose intolerance (10). The patient described in this report is a heavy alcohol drinker for more than 30 years and his uric acid levels are marginally elevated.

The elevated triglyceride levels seen in our patient can be associated with excessive alcohol intake and fat deposits. Although polyneuropathy was frequently described (~80%) in these patients, we couldn't find any evidence in our patient from history and physical examination. More investigations like nerve conduction test should have been done to confidently exclude polyneuropathy, but it was not possible in our particular setup.

The diagnosis of BSL is clinical and based on the typical fat tissue distribution in the trunk and proximal regions of the extremities. Imaging tests such as CT, ultrasonography or MRI may be useful. Clinical differential diagnoses include entities like angiolipoma, neurofibroma, encapsulated lipomas, hibernoma, congenital infiltrating lipomatosis of the face, liposarcoma ,lipoblastosis and lipodystrophy (11).

Fine needle aspiration cytology also assists in making diagnosis. Histologically, unlike conventional lipoma, the fat tissues in BSL are non-encapsulated with extension to the surrounding structures .The adipocytes are regular normal sized or slightly smaller with some spindling (12).

The treatment of BSL is surgery, and it is indicated in patients who have aesthetic deformities and significant respiratory embarrassment. A complete surgical removal of the tumor may not be possible as these tumors are not well-encapsulated and can infiltrate important structures (13). It is possible to use ultrasound-assisted liposuction for BSL as reported by some authors (14).Liposuction allows surgery to be done under local anesthesia and avoids the use of general anesthesia in patients with chronic alcoholism and possible liver lesion.

However many prefer open excision to liposuction because it offers a safer and more thorough debulking with a better chance to control bleeding (15). Abstinence from alcohol may prevent progression in the size of fat masses, but not in their regression. Other treatment options include ß2 agonists (which acts on lipolysis via adrenergic stimulation) and intralesional injection of enoxaparin although their efficacies have not been widely demonstrated (16, 17).

In our patient, we did excision of the most noticeable neck and axillary masses and told the patient to abstain from alcohol. The patient claimed to have stuck to the order, and we observed no increase in the size of the tumors 6 months after surgery.

Generally, BSL follows a benign course with few major complications and many patients require surgery for cosmetic reasons. Sometimes tongue or mediastinal involvement occurs resulting in compression syndromes of the trachea and superior venacava which can cause dyspnoea, dysphagia and dysphonia (18, 19). There is one report of a case in which a histologically proven lipoma underwent malignant degeneration to a myxoid liposaroma in a patient with Madelung's disease (20).

In conclusion, the occurrence of Madelung's disease in our patient suggests that the disease is not limited to Caucasian Mediterranean men only. Thus, we recommend that it be considered in a patient with a history of alcoholism and multiple symmetric mass deposits in the neck and the trunk.

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