

Multiple Symmetric Lipomatosis with Mediastinal Involvement and Subclinical Hypothyroidism in a Female Patient

MEDİASTİNAL TUTULUM VE SUBKLİNİK HİPOTROİDİ İLE BİRLİKTE SEYREDEN BİR KADIN MULTİPL SİMETRİK LİPOMATOZİS OLGUSU

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Summary

Multiple symmetric lipomatosis is a rare disease of undetermined cause characterized by multiple, diffuse, subcutaneous collections of non-encapsulated mature adipose tissue. A thick and disfiguring deposit of fat is symmetrically distributed on the neck, upper trunk and proximal portions of the upper extremities. The patients are usually middle-aged male alcoholics. The treatment is palliative surgical removal of excess fat from the neck and paracervical regions or any other lesion sites. The report about multiple symmetrical lipomatosis is very rare.

We describe and conclude a female case having hypothyroidism and multiple symmetrical lipomatosis with mediastinal involvement in this article.

Key Words: Multiple symmetric lipomatosis, Mediastinal involvement, Hypothyroidism

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Özet

Multipl simetrik lipomatozis (MSL) kapsülsüz yağ dokusunun multipl ve diffüz bir şekilde subkutan dokuda birikmesiyle karakterize, nedeni bilinmeyen nadir bir hastalıktır. Boyun, gövdenin üst kısmı ve üst ekstremitte proksimal kısımlarında simetrik olarak yerleşmiş, kalın ve şekilsiz bir yağ depolanması vardır. Hastalar genellikle orta yaş alkolik erkek bireylerdir. Tedavisi boyun ve paraservikal bölgeden veya herhangi bir diğer lezyon bölgesinden aşırı yağ dokusunun palyatif olarak cerrahi rezeksiyonudur. Multipl simetrik lipomatozise ait yayınlar oldukça nadirdir.

Biz burada mediastinal tutulumla ve hipotroidi ile birlikte seyreden bir kadın MSL olgusunu, nadir görülmesi nedeniyle tartıştık.

Anahtar Kelimeler: Multipl simetrik lipomatozis, Mediastinal tutulum, Hipotroidi

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Multiple symmetric lipomatosis (MSL), also known as Madelung's disease or Launois-Bensaude syndrome, is a rare disease clinically characterized by the presence of large symmetric lipomas around the neck, shoulders, upper arms or other parts of the trunk. The face, distal extremities, hands, and feet are characteristically spared (1,2). Neurological involvement, particularly peripheral neuropathy, is considered an integral part of the MSL syndrome (3,4).

MSL is predominantly seen in middle-aged male patients. The Mediterranean area seems to present the highest incidence of cases. Familial forms of the disease have been reported, but heredity seems to play a minor role (5). Several associated disorders have been described in MSL such as hyperuricemia, hypertriglyceridemia, liver disease, polineuropathy and hypothyroidism, however the cause of this syndrome remains unknown (1,2). High alcohol consumption is frequently associated and the peripheral neuropathy of MSL is often attributed to alcoholism (4).

Two patterns of distribution of lipomas have been identified. In the type I pattern, lipomatous deposits affect primarily the nape of the neck, the supraclavicular and deltoid regions. In the type II variant, lipomatous tissue diffuses and extends down over the trunk and the proximal part of extremities, giving the patients the appearance of simple obesity. Mediastinal involvement with lipomatous tissue, and superior vena cava syndrome has been described in patients with type I MSL. Despite the cosmetically aspect of the patient with a reduced range of motion of the head and neck, MSL is asymptomatic in most cases (2).

A nonalcoholic female case having MSL with mediastinal involvement, and hypothyroidism was reported in here and we review and update the literature related with this rare entity.

Case Report

A 44-year-old female patient applied with complaints of weakness, constipation and sleeping very much to our



Figure 1.

hospital. In her history, there were multiple operations such as resection of mass from back of the right cervical area 20 years ago and left scapular area 6 years ago and, total salpingohysterectomy 9 years ago. Histopathology of mass resection performed 6 years ago had shown lipomas and it was seen recurrence in same area after 6 months. There was no similiar disease among her relatives and she has never used alcohol in the past. Blood pressure was 140/90 mmHg, pulse rate was 66/min and body mass index was 35-kg/m². In her physical examination, we found multiple masses including in 12x10 cm diameter on the left scapular area, two masses in 3x3 cm diameters on the right scapular area and in 25x30 cm diameter on lateral of right breast (Figure 1). Thyroid was diffuse palpable at grade I and there was a nodule of 0.5 cm diameter in the left lobe. Other examination findings including nervous system were normal.

There were normal routine blood tests including

whole blood tests, biochemical tests and urinalysis except levels of lipids. Total cholesterol and LDL-cholesterol were slightly high. It was found normal free T3, free T4, anti-thyroglobuline (anti-Tg) antibody and anti-microsomal (anti-M) antibody, but level of TSH was found as higher (15.9 µIU/ml). Serum ACTH (adrenocorticotrophic hormone) and cortisol levels were normal (Table 1). Oral glucose tolerance test performed with 75 g glucose was evaluated as normal. Histopathological findings of biopsy taken from mass on the left scapular area showed non-encapsulated lipomatous tissue.

Abdominal ultrasonography resulted in steatohepatitis and thyroid USG revealed diffuse glandular enlargement and nodule of 4 mm diameter in the left lobe. Because of enlargement of the upper mediastinum in the chest radiography, computed tomography of chest was performed. There were multiple masses on the chest wall, multiple mediastinal mass especially on the back madiastinum and significant enlarged vena azygos and vena cava superior in her chest tomography (Figure 2,3). There was no malign degeneration including vessel invasion and density was like as lipomas in these masses. Therefore we considered them as lipomas. In spite of the enlargements of the vena cava superior and vena azygos, there was no clinical symptom belonging to vena caval compression. Electromyography revealed mild sensitive neuropathy in the lower extremities.

The patient was diagnosed as MSL in association with subclinical hypothyroidism by these findings and we started thyroid replacement therapy and antihyperlipemic therapy with simvastatin. There was no complaint related with appearance and mediastinal masses, and so any surgical procedure was not planned. The patient was discharged with these therapy and was invited to regular controls every three months.

Table 1. Laboratory examination in patients with MSL

	Patient	Normal		Patient	Normal
Hemoglobin (g/dl)	11.4	12-14	Total cholesterol (mg/dl)	239*	140-220
Hematocrit (%)	34.7	37-47	Triglyceride (mg/dl)	141	40-160
Leukocyte (mm ³ x10 ³)	7.5	4-10	LDL-Cholesterol (mg/dl)	165*	60-130
Platelet (mm ³ x10 ³)	340	150-400	HDL-Cholesterol (mg/dl)	56	30-85
ESR (mm/hour)	14	0-20	Free T3 (pg/ml)	3.45	1.8-4
Blood glucose (mg/dl)	101	70-110	Free T4 (ng/ml)	0.88	0.8-1.8
BUN (mg/dl)	11	7-18	TSH (µIU/ml)	15.9*	0.4-4
Creatinine (mg/dl)	0.9	0.7-1.3	Anti-Tg antibody	Negative	Negative
Uric acid (mg/dl)	4.3	3.5-7.2	Anti-M antibody	Negative	Negative
ALT (IU/L)	16	15-48	Plasma ACTH (pg/ml)	24.8	0-46
AST (IU/L)	18	13-40	Plasma cortisole (µg/dl)	17.1	5-25
GGT (IU/L)	27	9-50			

* high , Anti-Tg: anti-thyroglobulin antibody Anti-M: anti-microzomal antibody



Figure 2.

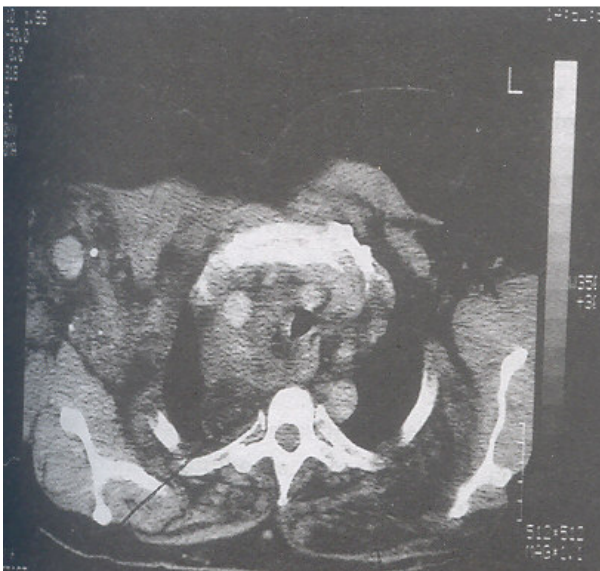


Figure 3.

Discussion

The disease was first described by Brodie in 1846 and subsequently was characterized by Madelung in 1888 when he reported 33 patients with cervical lipomatosis. Ten years later, Launois and Bensaude further defined this syndrome as multiple symmetric fatty accumulations (1,6). Approximately 200 cases have been reported in the literature, since the first report of MSL in 1846. MSL affects adults from 30 to 60 years of age; the male-to-female ratio has been reported as high as 15:1. The disease displays striking clinical features, with accumulation of symmetric deposits of nonencapsulated fat in different areas of the body (2,7,8).

Multiple systemic lipomatosis has been associated with some endocrine and metabolic disorders such as hyperuricemia, abnormal glucose tolerance, hyperlipidemia and hypothyroidism (6,9-12). Up to 90% of patients with Madelung's disease have associated alcoholism, which could explain some of the metabolic abnormalities (1,2,4,8).

There were no metabolic disorders such as hyperuricemia, hyperlipidemia or history of using alcohol, but it was determined hypothyroidism in our patient. In the literature, we found two patients reported MSL in association with hypothyroidism (9,10). However, we cannot find direct relation between MSL and hypothyroidism because we could not perform any molecular study. Some cases reported in the literature were found impaired glucose tolerance (11) or evident diabetes mellitus (7,12), but oral glucose tolerance test was found as normal in our patient.

Biopsy performed from mass on the left scapular area showed non-encapsulated lipomatous tissue and EMG was revealed mild sensitive neuropathy on the lower extremities. These findings were evaluated as harmonious with MSL.

Mediastinal extension of lipomatous tissue with obstructive symptoms caused by compression of the trachea and superior vena cava have been described in patients with type I MSL (1,2). Lipomatous tissue widens the mediastinum and may simulate mass lesions, thus leading to diagnostic errors. The main differential diagnosis lies between MSL and the fat accumulation of Cushing's disease, and liposarcoma. The diagnosis is made by history and physical examination, but when in doubt, CT scans and fine-needle aspiration of the fatty masses may be helpful (6,13).

Up to now, 37 cases reported in the literature and all of them except one had some evidence of clinical primary or iatrogenic Cushing's syndrome (6,14). Nguyen et al reported a male patient with mediastinal lipomatosis who had no history of steroid intake or evidence of primary Cushing's syndrome (14). Our patient had not used steroid and there was no evidence of Cushing's disease by clinical and laboratory examination. According to search of literature, our patient may be the only female patient who had these features. In this article, we report a MSL patient evaluated as a very rare case because of special features such as to be nonalcoholic female, to have mediastinal involvement without history of steroid use and being together with subclinical hypothyroidism.

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