



Case Report

Benign Symmetric Lipomatosis (Madelung's disease); A Case Report

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Abstract

Benign multiple symmetric lipomatosis (Madelung's disease) is a rare disease and characterized by non-capsulated accumulations of fat in a symmetrical manner around the neck and the shoulder, rarely involving the lower limbs and the lower body¹. The prevalence is increased among the mediteranean population² and very rare in the oriental area³. The etiology is still unknown. We have presented with a case of madelung disease type I associated with avascular necrosis at both femoral heads and gynecomastia at the right breast.

Keywords: Benign symmetrical lipomatosis, Madelung's disease.

Introduction

Benign symmetric lipomatosis is a rare disease first described in 1846 by Brodie⁴. Madelung subsequent reported 33 patients with cervical lipomatosis in 1888⁵, the classical description of the disease was published by Launois and Bensuade in 1893⁶ defined this syndrome as presence of multiple symmetric unencapsulated fatty accumulations diffusely involving the cervical and the upper dorsal regions, the abdomen and the groin.

Historically, Madelung's disease has been more frequently observed in men (male to female ratio about 15 : 1 to 30 : 1)⁷ and aged between 30-60 years old. The disease prevalence is increased among the Mediterranean population (incidence in Italy : 1/ 25,000 men)^{2,7}. There is clear relationship between this disease and excessive consumption of alcohol especially red wine^{10,11}. Our case was diagnosed Madelung's disease with avascular necrosis at both hips and right gynecomastia.

Case report

History

A 39-years-old male were referred from the surgical outpatient clinic for searching cause of palpable neck masses. He found that there were some painless lumps at his right posterior neck, both cheeks and the right breast for a year before presentation. He felt that they were slowly growing and he started to worry about their natures.

His past history was that he just had been avascular necrosis at bilateral femoral heads S/P bilateral bipolar hemiarthroplasty. His alcohol intake was excessive past 6 years but he tries to quit now. There is no family history of this condition and no continuous medication.

Physical examination revealed normal vital sign, no pallor or jaundice. There were ill-defined soft consistency mass-like lesions at the bilateral posterior neck and the anterior part of both ears. They measured about 15 cm and 4 cm in diameter; respectively. No sign of inflammatory process was demonstrated. The right breast was enlargement without definite mass or sign of inflammation. The left breast was unremarkable.

The liver and the spleen were not palpable. No sign of chronic liver disease was seen. The cardiovascular systems, the respiratory systems and the nervous systems were unremarkable.

The routine screening blood work up as CBD, BUN, creatinine and electrolytes analysis were unremarkable. Subsequently, He was underwent ultrasonography and computed tomography.

Image findings

The ultrasonography at the neck and both breasts.

The neck study

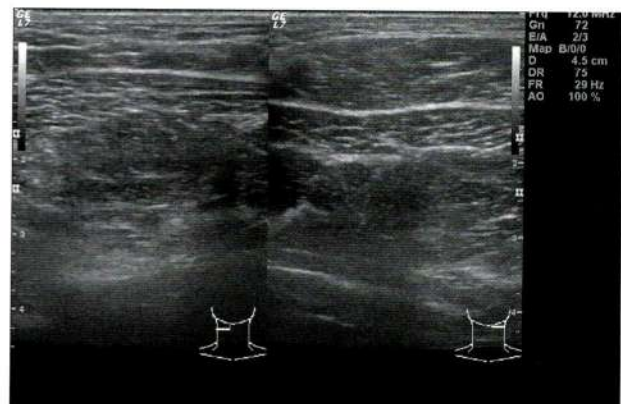


Fig.1 Ultrasound finding showed no definite mass at the posterior necks.

Both breasts study

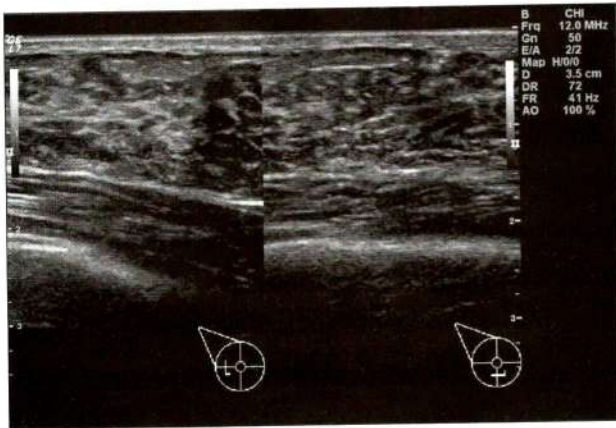


Fig.2a Ultrasound finding show fibroglandular tissue at the retroareolar area of the left breast.

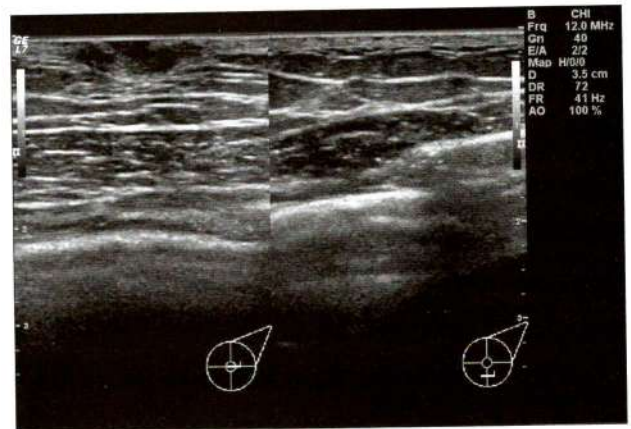


Fig.2b Ultrasound finding show normal nipple without retroglandular tissue of the right breast

The computed tomography of the neck

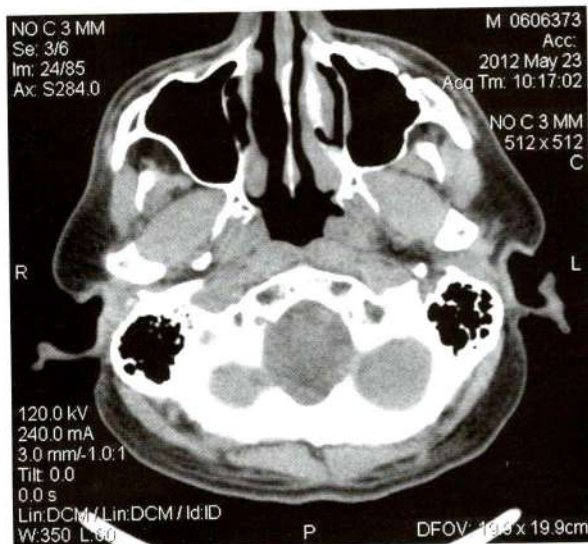


Fig.3a Axial non-contrast CT scan showed excessive fat filtration at bilateral cheeks

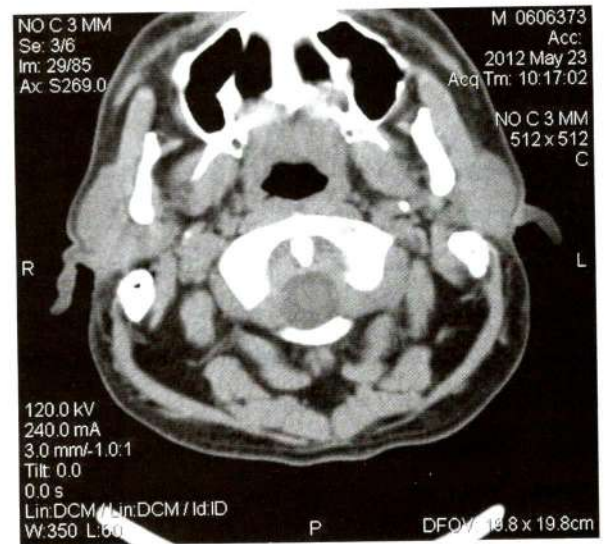


Fig.3b Axial non-contrast CT scan showed excessive fat filtration at posterior cervical space.

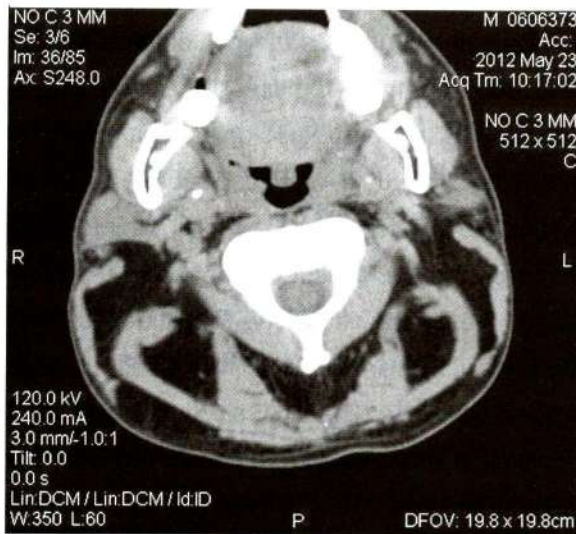


Fig.3c Axial non-contrast CT scan showed thinning of sternocleidomastoid muscles and paraspinal muscles with symmetrical fatty infiltration.



Fig.3d Axial non-contrast CT scan showed excessive fatty infiltration in submental area. No vascular invasion was seen.

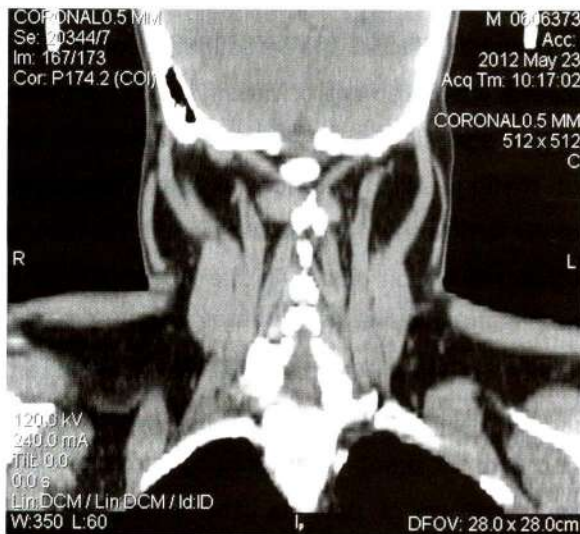


Fig.3e

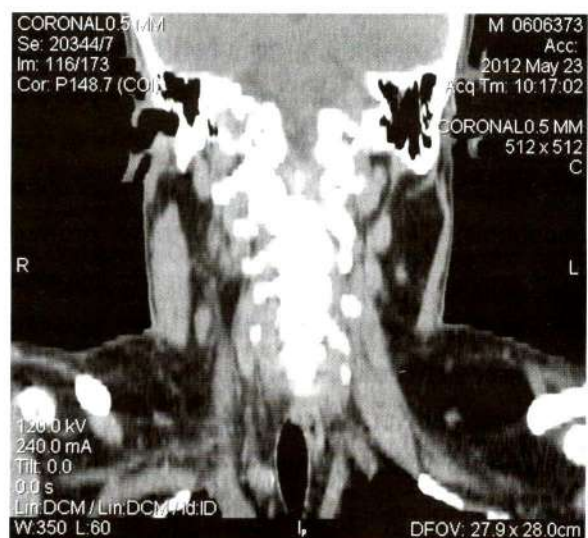


Fig.3f

Fig 3e and 3f Coronal non-contrast CT scan showed thinning of the sternocleidomastoid muscles and the paraspinal muscles with symmetrical fatty infiltration in the posterior cervical spaces, the perivertebral space and bilateral supraclavicular fossa.

He was underwent biopsy and his pathology revealed small piece of fibrofatty tissue as well as striated muscle. No inflammatory reaction or malignant was seen.

He was diagnosed as having multiple symmetric lipomatosis (Madelung disease). Finally; he was referred to Siriraj hospital.

Discussion

Benign symmetric lipomatosis is a rare disease characterized by non-capsulated accumulations of fat in a symmetrical manner around the neck and the shoulder, rarely involving the lower limbs and the lower body¹.

The pathogenesis is still unknown but there are several hypotheses such as defect in the adrenergic-stimulated lipolysis resulting in autonomy and massive proliferation of lipomatous cells, embryogenic brown fat undergoing functional sympathetic denervation, primary defect in brown fat mitochondrial DNA and recently it is found that 80% of patients with HIV -I infection treated with protease inhibitor develop this syndrome⁸.

There is clear relationship between this disease and excessive consumption of alcohol especially red wine^{10,11}. In our case, he had history of excessive alcohol intake in the past 6 years but he tries to quit now. Alcohol is known to have detrimental effects on many organs. In genetically susceptible individuals, alcohol have profound effect on fat metabolism which directly influences blood supply to marginal areas of perfusion such as the femoral heads¹² resulting avascular necrosis of bilateral femoral heads. Ingestion of the alcohol can also decreased in the serum testosterone level by increasing clearance from circulation through enhanced hepatic testosterone A-ring reductase activity resulting in gynecomastia¹³. Asymmetrical gynecomastia is common and unilateral gynecomastia may actually represent a stage in the development of bilateral disease¹⁷. Our case diagnosed of the avascular necrosis of bilateral femoral heads and gynecomastia of the right breast. His physical examination showed no other evidence of chronic liver disease and also the liver study showed

normal LFT. There are also associated of medelung's disease with liver disease, hyperlipidemia including elevated LDL, hypertriglyceride, hypothyroidism, diabetes mellitus and neurological changes^{11,13}.

The symptoms of this disease are variable although the patients may complain of interference with neck motion, difficulty in obtaining a proper fit cloths or respiratory difficulty. The fat deposits never spontaneous disappear and the disease is usually progressive over a period of years¹. Our case seeked for medical help due to progression in size of his palpable masses.

Diagnostic is usually made of history and physical examination and confirmed by sonography, computed tomography (CT) and fine needle aspiration cytology. The sonographic finding of lipomatosis are typical however despite the frequent utilization of the ultrasound in the investigation of the neck masses, measurement of density by CT provide a higher diagnostic fidelity^{11,15,16}. In our cases, we could not distinguish the abnormal fatty tissue from normal fatty tissue by using ultrasound. We detected only some lymph nodes in the deep part of the neck. But when we used the CT, the CT could display the typical character of disease. Currently CT is considered to be the method of choice for the diagnosis; preoperative staging and post operative follow up of patients. The utilization of CT allows the investigator to rule out relevant differential diagnosis such as lymphoma and metastasis lymph nodes involvement. Also it allows the tumor staging in cases of multiple systemic lipomatosis associated with malignant disease¹¹. The MR imaging can also provide the surgeon with adequate information¹⁶.

The multiple symmetric lipomatosis is classified into three types, type I or diffuse lipomatosis of the

neck (horsecollar lipomata) lipomatous deposits maintain the aspect of the circumscribed protruding masses affecting primarily the nape of the neck as well as the supraclavicular and the deltoid regions¹. In advanced cases, there will be mediastinal involvement with trachea and vena cava compressing as well as erodigestive tract compression. Type II or multiple systemetric lipomata of the shoulder girdle, the upper arms, the thorax, the thighs and sometimes the abdomen giving the patients so called pseudoathletic appearance and type III or a rare type with preponderance of the lipomatosis in the pelvic girdle (hips and thighs); gynecomoid type⁹. Our case had characteristic of type I.

At the present time point there is no causal therapy available and dietetic intervention are not successful. Removal therapy is the only successful treatment although relapse may occur. Two main procedures are lipectomy or liposuction⁷. Our case was referred to Siriraj hospital for proper management.

In conclusion; we presented with a case of madelung's disease type I associated with avascular necrosis at both femoral heads and gynecomastia at the right breast resulting from excessive alcoholic consumption without definite evidence of chronic liver disease.

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