CT IMAGING OF MADELUNG DISEASE: A CASE REPORT

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Madelung Disease was first described by Launois and Bensaude in 1898 (1). Its synonyms are Launois-Bensaude disease, Buschke disease, cervical lipomatosis, Madelung neck and multiple symmetrical lipomatosis (2). It is characterized by progressive growth of fat masses which are located symmetrically at the neck, shoulders, chest, abdomen and groin (3). The disease seems to be more frequent in the Mediterranean regions, and approximately 97% of those affected are men.

We report a case of Madelung disease in a 38 years-old Thai male patient.

CASE REPORT

A 38 years-old Thai male patient from Ubolrajthani, a province in the north-eastern part of Thailand, came to the Ramathibodi Hospital due to the presence of neck mass for 2 years. At first, the soft and bulky mass appeared at the anterior chest wall; then it grew slowly up to the anterior and posterior part of the neck. He was not dyspneic. The systemic symptoms did not exist. He drank alcohol for 15 yrs, 2 times a week and smoked for 10 yrs. Non I.V. contrast CT scan of the neck, performed at Subprasithprasong Hospital in Ubolrajathanee (see figure 1) , showed markedly increased fatty tissue around the central structures of the neck without distorting them. The airway was not narrowed. The fatty tumors were unencepsulated, so that the borders between tumors and surrounding tissue are not defined. The histology confirmed the mass to be lipoma.

DISCUSSION

Studies have linked Madelung disease to a specific defect in the regulation of catecholamine-induced lipid mobilization (4,5); thus it could be considered a "triglyceride storage disease" involving adipose tissue. Clinical manifestations are as followings; 1) the onset is in adulthood 2) massive lipomatosis (normal fat that often begins on the back of the neck and extends anteriorly to the submental region and to the thorax in a symmetrical fasion) may spread to the scrotal region 3) respiratory system symptoms related to tracheal compression and recurrent palsy 4) venous stasis of the chest wall in association with mediastinal involvement 5) neuropathy (sensory, motor, autonomic) 6) muscular weakness, tendon areflexia, muscle atrophy, tremor, cramps, loss of vibratory sensation, hypoesthesia, sciatica-like pain, trophic changes, segmental hyperhidrosis, gustatory sweating, impotence, tachycardia at rest etc. 7) metabolic abnormalities, marked increase in adipose tissue lipoprotein lipase activity, plasma hyperalpha-lipoproteinemia, defect in the adrenergic stimulated lipolysis in lipomatous tissue, hyperuricemia, reduced glucose tolerance, renal tubular acidosis 8) red blood cell macrocytosis, macrocytic anemia 9) abnormal liver function test results related to elevated alcohol intake 10) no signs of abdominal or pelvic involvement 11) sudden death.(7-10).

Familial occurrence has been described (11-13). A hyperplastic mechanism has been postulated, with in vitro studies demonstrating a defect

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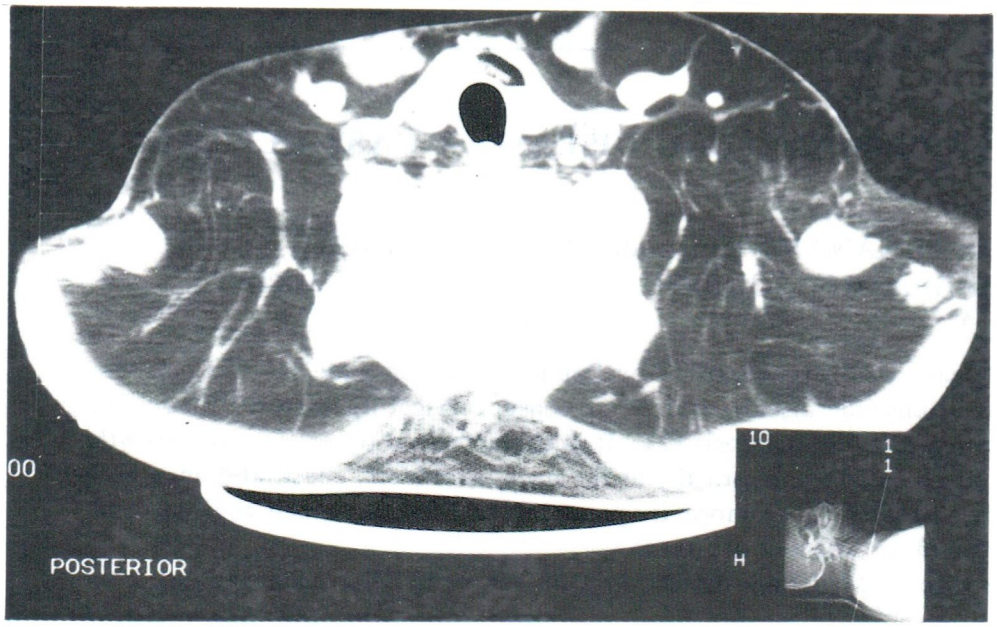

Fig. 1A.
Non I.V. enhanced CT scan of the lower neck showed diffuse fatty tissue infiltration around the central part of the neck.

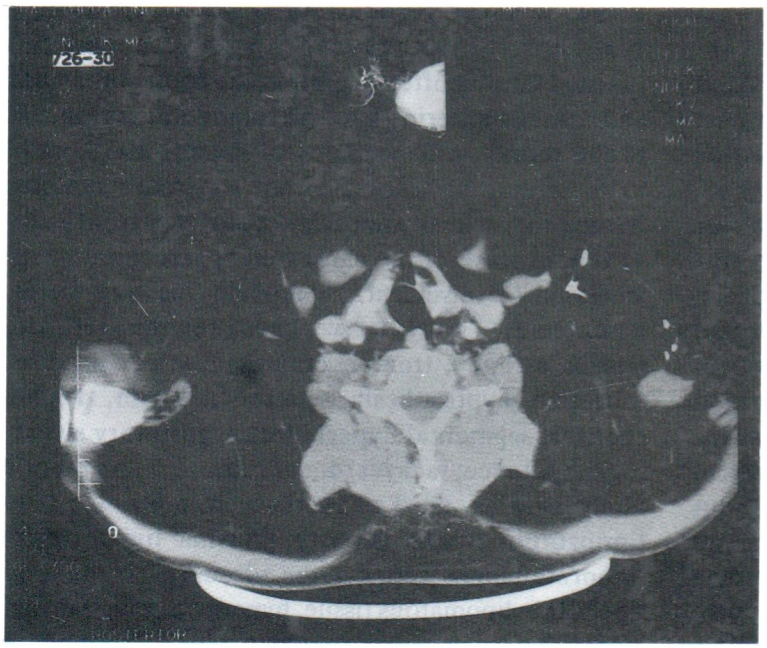

Fig. 1B.
Soft tissue window of the same image as in fig. 1A. showed no distortion of the central structures.

in adrenergic-stimulated lipolysis of lipomatous tissue (4,5). The uninvolved subcutaneous fat is usually very poorly represented of frankly atrophic, and signs of mediastinal compression have often been described (14,15,16). No abnormal deposition of fat in the anterior mediastinal region (17), cardiophrenic angle, or retropleural locations was seen, contrary to observations in patients with long term steroid treatments (18-20), Cushing syndrome (21,22), or obesity (23-25).

Radiologic manifestations in 15 patients with Madelung disease described by Enzi (3) were 1.) Lipomatosis (neck, mediastinum, below the trapezius muscle) 2.) Calcification/ossification within the lipomatous masses 3) tracheal narrowing and deformity 4) venous stasis 5) absence of pericardial, intraabdominal, retroperitoneal and pelvic lipomatosis 6) Large amount of fat at anterior abdominal wall and pubic region.

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