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A case of madelung disease mimicking a superior veina cava syndrome: A challenging diagnosis resolved by CT scan

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Abstract
Madelung's disease or benign symmetric lipomatosis is uncommon, alcohol abuse is frequently associated. Here, we report a moderate form with bilateral swelling over the supraclavicular fossa, cervical and nuchal areas associated with marked dilation of veins on the skin surface misleading to the diagnosis of superior vena cava syndrome. The final diagnosis was confirmed by CT scan by showing important bilateral unencapsulated fat in the subcutaneous fat of neck, in supraclavicular fossa and beneath the trapezius muscle.

Keywords: alcohol abuse; benign symmetric lipomatosis, computed tomography (CT), Madelung's disease

Introduction
Madelung's disease (MD) or benign symmetric lipomatosis is an uncommon disorder which is characterised by the presence of multiple symmetrical fatty accumulations, usually involving the upper trunk, neck and head [1]. The incidence of this disorder is 1:250000, male patients are more affected (the sex ratio is 15:1) [2]. Given the fact that MD is a rare condition, diagnosis was challenging. Here, we report a moderate form which the diagnosis was guided par CT scan.

Case history
A 44-year-old Malagasy man presented to our hospital with a progressive swelling of his neck shoulder and arm. His medical history revealed mild alcoholic hepatitis 2 months prior the visit. Neither dyspnea nor dysphagia was noted. Clinical examination revealed a bilateral swelling with indistinct margins over the supraclavicular fossa, cervical and nuchal areas associated with marked dilation of veins on the skin surface (figure 1). A gynecomastia was also noted. The swelling was soft and fluctuant; neither tenderness nor inflammation was observed. There were no palpable cervical nodes. Moreover, the patient presented a discrete of lower limbs oedema. Laboratory tests findings were as follow: fasting blood glucose level was normal (1.40 g/l), (Alanine aminotransferase was 2 times the upper limit of normal values, and aspartate aminotransferase was 4.5 times the upper limit of normal value, triglycerides were borderline high (2 g/l) and Prothrombin was low (38.5 %). Hepatitis C and hepatitis B serology were negative. Chest CT- scan was performed to rule out superior veina cava syndrome. This latter revealed important bilateral unencapsulated fat in the subcutaneous fat of neck (density -112 HU) occupying supraclavicular fossa and beneath the trapezius muscle (figure 2 and 3). There was no circumscribed mediastinal mass even though fat accumulation in mediastinum was noted. The Diagnosis of Madelung disease was established.

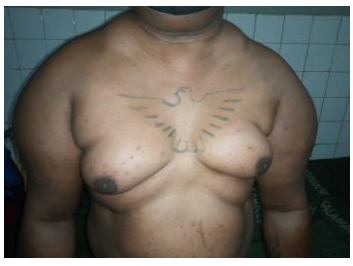

Fig 1: Swelling of the supraclavicular fossa, shoulders, neck and Gynecomastia

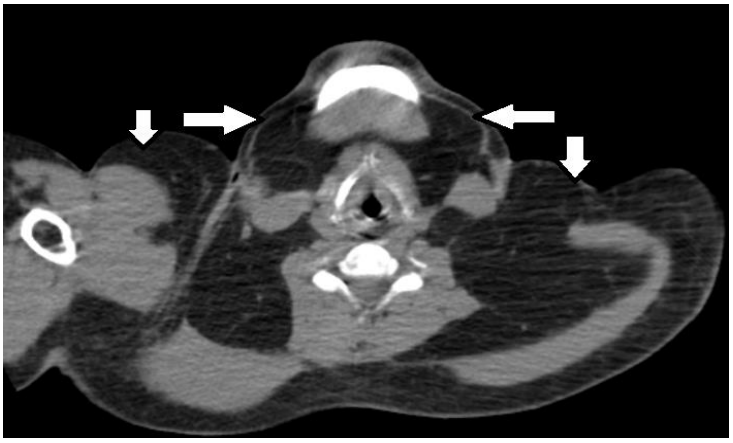

Fig 2: Axial CT scan in Mediastinal window showing an accumulation of unencapsulated fat in subcutaneous tissue of the neck (horizontal arrows) and sub clavicular fossa (vertical arrows)

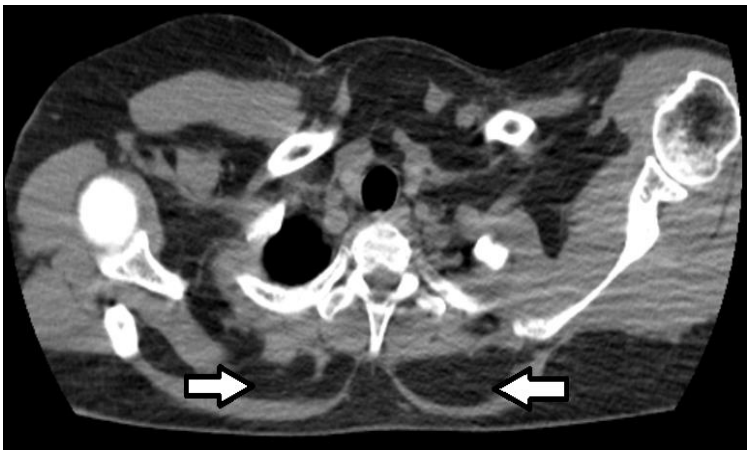

Fig 3: Axial CT scan in mediastinal window showing an accumulation of unencapsulated fat beneath trapezius muscle (arrows)

Discussion

The main feature which characterizes our case in comparison with the previously described forms [3, 4] was marked dilation of veins on the skin surface with the moderate swelling of the neck, shoulder and arm misleading to the diagnosis of superior vena cava syndrome. Indeed the swelling in our patient can be considered as moderate in comparison with others cases reported in literature which described severe forms [5, 6]. The diagnosis of MD is based on history, clinical appearance, and the results of imaging like CT scan [4]. Most of the times as like in our patient MD involves the upper trunk classified as type 1 according to Enzi classification [7]. Clinical manifestations habitually include a symmetric distribution of fat in the submandibular region, over the sub occipital area, the shoulder, and proximal upper extremities, in contrast to the distal upper extremities and body. Differential diagnosis includes: lymphoproliferative diseases, lymph node enlargement, enlarged goiter, cervical cysts and benign tumors but they can be differentiated by computed tomography or biopsy [8]. Most of those MD features were seen in our patients a part from gynecomastia which was rarely reported [9]. Benign symmetric lipomatosis was initially described in 1846 by Brodie [10], and it is more commonly seen in men of Mediterranean descent in the 3rd -6th decade of life who has a history of chronic alcohol use [11]. According to your knowledge this is one of the first Malagasy cases reported, and alcohol abuse was also found. CT scan confirm the diagnosis by showing excess of symmetric and unencapsulated fat deposited at a superficial site such as subcutaneous tissue and at deep sites like under the sternocleidomastoid and trapezius muscles, posterior cervical triangle, around the salivary glands and paraspinal muscles and larynx [12]. Preoperative imaging plays an important role in defining the extent and distribution of fat and the only effective treatment is excision of the masses, liposuction can be done for patients with small masses [13]. Abstinence from alcohol is also recommended in MD [8]. Our patient has stopped alcohol consumption but no surgery of liposuction has been done due to financial issue.

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