

Case Report

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Madelung's Disease

Madelung's disease is a rare disease of undetermined cause characterized by symmetric deposits of painless, diffuse, subcutaneous adipose tissue on the suboccipital area, the cheeks, the neck, the shoulders and the upper trunk. It is commonest in countries bordering the Mediterranean. The patients are usually middle-aged male alcoholics. Although lesions can initially mimic a head and neck malignancy, management is essentially symptomatic, with conservative removals done as indicated clinically or for cosmesis. Oriental reports about Madelung's disease are very rare. Here a case of Madelung's disease in a 59-year-old male is described. We describe his clinical course and review the literature.

Key Words

Launois-Bensaude syndrome;
Madelung's disease;
multiple symmetric lipomatosis

Madelung's disease was first described in 1846.¹ Since then, approximately 200 patients have been reported in the literature.² It is characterized by non-encapsulated accumulations of fat in a symmetric manner around the neck and shoulders, rarely involving the lower limbs and the lower body. The cervical areas of fat enlarge slowly, giving the appearance of a "horse collar". A clear association with alcohol abuse is reported, but the etiology remains uncertain. The clinical course of Madelung's disease is variable, but usually involves an initial period of several years when there is a rapid growth of fat deposits followed by a period of stabilization or slow progression over many years. Spontaneous regression does not occur and the treatment is palliative surgical removal of excess fat from the neck and paracervical regions or any other lesion sites. Oriental reports about Madelung's disease are very rare.^{3,4} Here, we present a typical case and review the literature.

CASE REPORT

A 59-year-old nondrinker presented himself to the

otolaryngological service in the Taipei Veterans General Hospital, complaining of slowly enlarging mass in his posterior neck. He first noticed the appearance of the lesion 3 years prior to this presentation. He did not pay attention to the mass until it bothered his head motion (Fig. 1). There was no dyspnea or dysphagia. Physical examination revealed the swelling mass was situated in the bilateral suboccipital region, sized 7 × 6 cm. It was soft and fluctuant, with no definite margins, and there was no tenderness or inflammation. No cervical nodes were palpable. The patient had no evidence of sensory, motor or autonomic neuropathy.

A computed tomography scan showed a soft tissue mass (about 8.0 × 7.5 × 4.0 cm in size) with irregular contour and chiefly fat content over the inter-muscular space of the posterior neck (Fig. 2), extending from the C₂ dens base to the thyroid cartilage.

Laboratory examination revealed the full blood count, electrolytes, liver function tests and blood glucose were all normal.

The patient underwent surgical excision of the posterior neck tumor under general anesthesia. Everywhere

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Fig. 1. Preoperative and back views of fatty deposits (arrow) in the suboccipital region.

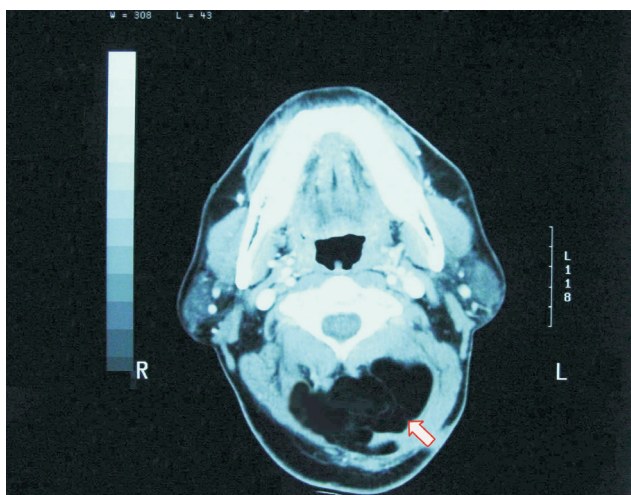


Fig. 2. Computed tomography scan shows the irregular lesion of fat density (arrow) occupying the posterior neck, extending from the level of C2 dens to the thyroid cartilage.

the tumor blended with the surrounding tissues, no capsule was seen. Histological examination revealed fatty tissue. Post-operative course was smooth. The appearance and function of the head motion were normal.

In view of the history, clinical finding and the histology report, a diagnosis of Madelung's disease was made.

DISCUSSION

Madelung's disease, cervical lipomatosis, Launois-Bensaude syndrome, "fat neck", and multiple symmetric

lipomatosis are all terms used to describe the same rare disease. It was first documented by Brodie in 1846,¹ and in 1888, Madelung⁵ described a series of 33 patients with "cervical lipomatosis". Ten years later, Launois and Bensaude⁶ described a further series and defined the syndrome as the presence of multiple symmetrical fatty accumulations involving the head, neck and upper trunk. The deposits not only are cosmetically disfiguring but also may be associated with respiratory symptoms.

The etiology of Madelung's disease is still obscure. There are 2 theories behind the pathogenesis of benign symmetric lipomatosis. First, a defect in the adrenergic-stimulated lipolysis results in autonomy and massive proliferation of lipomatous cells. And second, embryologic brown fat undergoes functional sympathetic denervation, resulting in hypertrophy of these fat cells.²

The disorder predominantly affects white males (M:F ratio 15:1),⁷ with the incidence being highest in the peoples of the Mediterranean (reported incidence in Italy is 1 in 25,000 males).⁸ The fat deposits are commonest between the ages of 30 and 60 years. There is a history of alcohol abuse in nearly all the cases; it is rarely reported in non-drinkers.

The symptoms are primarily those of disfigurement, although the patient may complain of interference with neck motion, difficulty in obtaining a proper fit with clothing, or (in some cases) respiratory difficulties. The fat deposits, once present, never spontaneously disappear, and the disease is usually progressive over a period of years. Although multiple cases have been reported in some families, most cases have been singular, with no evidence of hereditary predisposition.

Two patterns of distribution of lipomatous tissue have been identified.⁸ In the type I pattern, lipomatous deposits maintain the aspect of circumscribed protruding masses affecting primarily the nape of the neck, as well as the supraclavicular and deltoid regions. Mediastinal involvement with tracheal and vena cava compression is possible. The aerodigestive tract may be compressed in advanced cases, causing dyspnea and dysphagia. Occasionally diminished movement of the neck and upper extremities may result. In 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. In the type II, space occupying

mediastinal lesions are not found. Our case had the characteristics of type I.

Diagnosis is usually made by history and physical examination, and confirmed by sonography, computed tomography and fine needle aspiration cytology.⁹

Grossly, the adipose deposits are firm, non-tender, non-encapsulated, and they blend well into the surrounding subcutaneous fat. Microscopically, the fatty tissue is indistinguishable from other lipomatous tissue. Electron microscopy has not shown any significant characteristics.¹⁰

Several associated disorders have been described in Madelung's disease, such as hyperuricemia, liver disease, polyneuropathy, abnormal glucose tolerance, and hyperlipidemia.

There has been only 1 reported case of malignant transformation in Madelung's disease.¹¹ Tizian described the development of a intramyxoid sarcoma. However, in 1987, Ruzicka *et al.*⁷ reported an association with malignant tumors of the aerodigestive tract (carcinoma of the hypopharynx, metastatic oat cell carcinoma and paracecal tumors). This was confirmed by Hermann *et al.*,¹² who reported an increased incidence of carcinoma of the tongue and pharynx in patients with Madelung's disease. Thus, a thorough evaluation to rule out synchronous malignancy in patients with Madelung's disease is mandatory.

The disorder must be distinguished from other lesions of the subcutaneous tissues, including angioliipomas, neurofibromas and hibernomas. Hereditary multiple lipomatosis is fairly common, but usually affects the extremities, the lower chest and the abdomen more predominantly. Dercum's disease is characterized by painful fatty deposits, and it occurs almost exclusively in obese, menopausal females.¹³ The differential diagnosis is usually not difficult because of the grotesque appearance of patients with Madelung's disease.

Successful medical therapy has been reported using a β_2 -agonist (Salbutamol) in patients who maintain normal lipolysis and cAMP accumulation in the presence of adrenergic stimulation.¹⁴ Weight loss, abstinence from alcohol, and reversal of other metabolic abnormalities are recommended despite there being no evidence that the progression of Madelung's disease will be reversed or arrested by these measures.

Conservative surgical therapy is indicated in a pa-

tient with a severe cosmetic deformity, causing psychological stress, and if there is compression of the aerodigestive tract causing dyspnea or dysphagia. The infiltrative nature of the non-encapsulated fatty deposits and low probability of malignant transformation make complete excision of these tumors unlikely, and vital structures should not be compromised in a futile attempt to completely excise these tumors.²

Liposuction techniques may offer a better alternative to standard surgical therapy in selected cases.^{15,16}

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