

CLINICAL PHOTOGRAPHS

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Benign symmetric lipomatosis (Madelung's disease)

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Clinical Note

A 56-year-old Mediterranean man with a history of smoking and alcohol abuse reported progressively enlarging masses in his cheeks, anterior and posterior cervical region, and back for 8 years. The patient had a past medical history significant for alcoholic hepatitis. He denied shortness of breath, dysphagia, odynophagia, or any other problems. The patient stated the cosmetic deformity was unappealing, and he came to our clinic for treatment.

Physical examination revealed a friendly, slender man in no apparent distress. Personal hygiene was poor. Ear, nose, and throat evaluation was completely within normal limits. The face had soft, spongy masses in both parotid regions and the anterior cervical region (Fig. 1). The posterior cervical and upper back regions also had masses that were soft, spongy, and nontender (Fig. 2). The heart, lung, and abdominal examinations were normal. There were no masses on the extremities.

Computed tomography scan revealed extensive infiltrating masses in the neck and back. The density of the lesion was consistent with fat.

The patient underwent a cervical lipectomy. Pathology revealed lipomatous tissue.

DISCUSSION

Benign symmetric lipomatosis was first described by Brodie in 1846. However, it was Madelung in 1888

who reported a series of 33 cases of the disease that now bears his name. Since then about 200 cases have been reported in the world literature.¹

The cause of this disease is still undetermined. However, the disease seems to affect middle-aged alcoholic men of Mediterranean descent, although women and nonalcoholics may be affected.

There are two 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.¹

Most patients with benign symmetric lipomatosis come to the physician because they are displeased with their appearance. However, more serious findings may be apparent. These include difficulties raising the arms or turning the head, difficulties with deglutition, hoarse voice, and more serious problems including respiratory complications caused by compression on the airway.² The physician must be wary that this benign disease is diffuse and infiltrating in nature.

The diagnosis of Madelung's disease is primarily a clinical one. History and physical examination usually reveal alcoholic men with soft, lipomatous-feeling masses in the parotid region ("hamster cheeks"), cervical region ("horse collar") and back ("hump back").³ Computed tomography scan and magnetic resonance imaging can be helpful especially when planning surgical excision. Fine-needle aspiration may also assist in the diagnosis.

Medical treatments have been advocated, including β 2-agonist to impair adrenergic lipolysis. However, surgical excision is still the most effective treatment. Surgical modalities include lipomatous excision¹ and liposuction.⁴ It should be emphasized that conservative surgical management is used because this is a benign disease. Complete surgical removal

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Fig. 1. Front view of patient with Madelung's disease.



Fig. 2. Back view of patient with Madelung's disease.

of this tumor can endanger important anatomic structures because the lipomas can infiltrate or encapsulate these structures.

The prognosis for these patients is good. Recurrence is common because of the inability to completely excise these tumors. Multiple procedures may need to be performed. Abstinence from alcohol may help reduce the rate of recurrence.

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