Madelung’s Disease


Division of Endocrinology and Metabolism (N.-C.Y., C.-Y.Y., C.-W.C., F.-C.Y., S.-Y.L., K.-J.T.), Department of Internal Medicine, Chi Mei Medical Center, Tainan 71004, Taiwan; and Chia Nan University of Pharmacy and Science (K.-J.T.), Tainan 71710, Taiwan

Madelung’s disease is a rare condition characterized by a symmetrical pattern of massive fatty deposits (1). The condition is also known as benign or multiple symmetric lipomatosis, or the Launois-Bensaude syndrome. Two types of Madelung’s disease have been described. In type 1, fat accumulates around the neck and the nape of the neck, shoulders, upper arms, and upper back. In type 2, lipomas are distributed over much of the body, including the hips and thighs. Fatty deposits rarely extend to the lower limbs, mediastinum, and larynx in either type (2). Although the pathophysiology remains elusive, Madelung’s disease may be caused by a local defect in catecholamine-induced lipolysis (3). Most patients have a history of chronic alcoholism. Mediterranean men appear to be at highest risk of acquiring the condition, whereas the disease is remarkably rare in Asian populations (3). The diagnosis of Madelung’s disease is primarily based on physical examination, clinical history, and imaging studies. Diabetes mellitus (DM), lipid disorders, liver disease, and hypothyroidism are frequent comorbidities (4). Neuropathy, including sensory, motor, and autonomic polyneuropathy, is observed in about 85% of patients, and the latter is associated with sudden cardiac death (5).

A 60-yr-old Han Chinese man with a body mass index of 19.3 kg/m² presented with a 12-yr history of soft, progressive swelling masses over the neck and upper back. He had abstained from alcohol for the previous 5 yr. Nine years prior, the patient was diagnosed with DM and dyslipidemia. His family history was unremarkable. Concurrent with many years of poorly controlled DM, the patient’s limbs and abdomen were relatively thin and atrophied. The patient showed no signs of moon face, paper skin, hirsutism, or purple striae.

Lab results revealed a total cholesterol level of 319 mg/dl, a low-density lipoprotein cholesterol level of 218 mg/dl, a high-density lipoprotein cholesterol level of 43 mg/dl, a triglyceride level of 286 mg/dl, and a hemoglobin A1c of 11.0%. Thyroid function and cortisol were within normal limits. Magnetic resonance imaging revealed diffuse, nonencapsulated fatty deposits in the sc and deeper fascial compartments of the neck, upper trunk, and upper back (Fig. 1, A and B). Accordingly, a diagnosis of type 1 Madelung’s disease was made. The patient was prescribed a statin and premixed insulin (total daily dose of insulin, 34 IU). Because this patient was asymptomatic, no surgical treatment was proposed.

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Address all correspondence and requests for reprints to: Dr. Kai-Jen Tien, Division of Endocrinology and Metabolism, Department of Internal Medicine, Chi Mei Medical Center, Tainan 71004, Taiwan. E-mail: cmmctkj@gmail.com.

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References


Abbreviation: DM, Diabetes mellitus.
FIG. 1. T1-Weighted magnetic resonance imaging scan of patient with Madelung’s disease in sagittal (A) and axial (B) views.