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Case report

A Case Repot of Madelung's Disease in a 69 Years Old Man

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A B S T R A C T

Madelung's disease is an extremely rare disorder of unknown etiology. It is characterized by the huge, bilateral, fatty deposits in regions of the neck, shoulders and the upper extremities. A 69-old patient with developed symptoms of Madelung's disease with a 12-years history is described in this study. The patient was initially considered to have a goiter and chronic heart insufficiency, for which he has been treated for three years. Although the Madelung's disease can be diagnosed right after detailed clinical examination, this study pointed out possible diagnostic and therapeutic mistakes when a physician in a differentiation of symmetrical neck and shoulder swellings doesn't consider the possibility of diagnosing a Madelung's disease.

Key words: *Madelung's disease, Madelung's neck, multiple symmetric lipomatosis, diagnosis*

Introduction

Madelung's disease is characterized by massive, symmetrical, neck, shoulder and upper extremities adipose tissue deposits and it was named in honor of Otto Madelung, German physician who first described the series of such patient's in 1888. First description of this clinical entity has appeared in 1846 by Sir Benjamin Brodie[1]. Men are much more frequently affected with this disorder, usually in the age between 30 to 60 years[1]. The biggest incidence is recorded among the inhabitants of the Mediterranean region[1], such as Italy with 1 case *per* 25,000 men[2]. Madelung's disease is benign and with unknown etiology[3]. Most of the patients have longtime history of heavy alcohol intake[4–6], while its association with hypothyroidism, diabetes mellitus, liver disease or polyneuropathy is also well known[1,4,5]. Rare cases of synchronously appearance of Madelung's disease with some of aerodigestive tract cancers were described[7], and just only one case of malignant degeneration inside the fatty deposits[8]. Massive deposits of no encapsulated, adipose tissue, with symmetrical arrangement in the neck, shoulders and upper extremities develop in months or years[1,5,6]. At first, patients usually complain on aesthetic changes and difficulties in finding appropriate clothes because of thick neck[1,9], but in longstanding disease, fatty deposits begin to diminish movements of neck, shoulders and arms, dyspnea and dysphagia as a result of compression to upper mediastinum occur as well[1,10]. It is possible to make the reliable diagnosis of Madelung's disease only after detailed clinical examination. Using imaging diagnostic methods, such as magnetic resonance (MRI), computed tomography (CT) or ultrasound (US) the physician can define disease outspread[2,11,12]. Madelung's disease's unique appearance makes differential diagnosis simple, especially when physicians have it on his mind. Madelung's disease symptoms can be misinterpreted with cysts, lymphomas, sialoadenitis, sarcomas or simple obesity [13]or mixed, as described in this study, with goiter or chronic heart weakness. Surgery is the first choice in Madelung's disease therapy and it is more effective than other known methods. It usually takes few sessions to completely remove bulking deformities[1,9,14]. Inferior expanded face lifting and neck dissection cuts are mostly used to access adipose tissue[9], which can be removed in combination of lypectomy and liposuction techniques[6,14]. Despite of ra-

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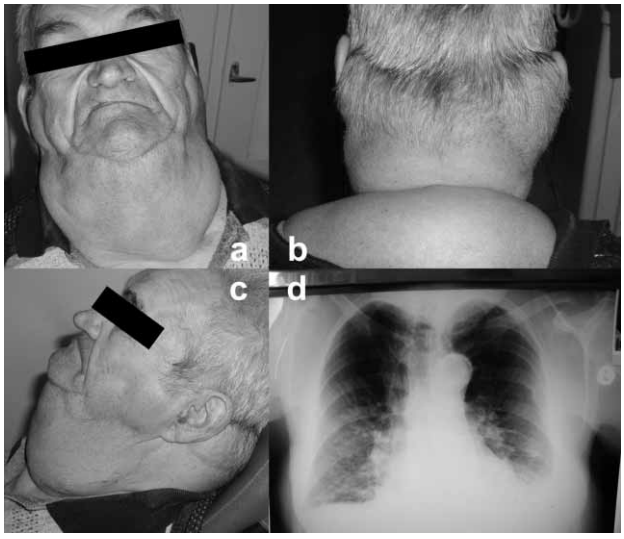

Fig. 1. a) Frontal view, b) Back view, c) Left profile, d) Chest X-ray.

dical procedures recurrences are very frequent, mostly because of impossibility of total fat removal which is non-encapsulated and located between neck envelopes[5,6,14]. Results of Madelung's disease treatment with Salbutamol are still controversial and not finally evaluated[15].

Case Report

This study described a case of a man, 69 years old, retired, non-smoker, with 17 years of non-drinking history. First disfigurement has appeared 12 years before in the anterior neck area which imposed like goiter. Thyroid hormones were in referent ranges at this time and during controls, so patients did not receive any medication. In the meantime anterior neck disfigurement has become conspicuous altogether with new deformities of lateral and back regions of neck and on shoulders (Figure 1a-c). Patient had nocturnal dyspnea in last three years, and the most symptoms occurred during lying on the back. Those symptoms were misinterpreted as signs of chronic heart insufficiency so he was hospitalized for a few times. He had received treatment of cardiac inotrops and diuretics, without obvious clinical improvement. During the last hospitalization the complete diagnostic testing was made. An ECG was changed in meaning of tachyarrhythmia, blood pressure was 130/90 mmHg and pulse was 140 b/min. Except for findings in redistribution of blood flow in upper parts of lungs, atonic core and left side pleural effusion shown by chest X-ray, the big soft tissue shade appeared in upper mediastinum which was characterized as enlarged thyroid gland. (Figure 1d). All laboratory findings were in referent ranges. The ultrasound examination showed signs of hepatic fatty infiltration without any focal lesions which clinically manifested as hepatomegaly, about 8 cm below rib arch. Maxillofacial surgeon recognized Madelung's disease and advised surgical removal of fatty tissue which patient denied.

Discussion

Although the diagnosis of Madelung's disease can be made only after clinical examination, and by one of imaging diagnostic methods such are MRI, CT or ultrasound[2,11,12], this study indicate possibility of diagnostic and treatment mislead, especially in evaluation of neck and shoulder deformities when physician don't know for this rare clinical entity. Madelung's disease progress slowly, in our case it took almost 12 years. Primary it changes appearance of patient, without functional difficulties. Esthetics was not big problem for our patient, until functional difficulties such as nocturnal dyspnea begun, which was the main reason for visiting physician. Although he developed all clinical signs of Madelung's disease[1,4,10], the disease has not been recognized, and patient was mistreated. Patient presented in our study is different than most of other Madelung's disease patients because his alcohol intake was rear almost insignificant[4–6]. Patient did not have any of Madelung's disease associated diseases, only gentle triglycerides and cholesterol elevation and fatty liver infiltration[5,15].

Conclusion

This study indicates that despite the simple diagnosing scheme for Madelung's disease, mistakes could be made if large, symmetrical and bilateral disfigurements in head, neck and shoulders regions are not considered in sense of Madelung's disease especially in geographic regions with higher incidence of this disease.

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MADELUNGOVA BOLEST – PRIKAZ SLUČAJA

S A Ž E T A K

Madelungova bolest je rijetka, nepoznate etiologije, karakteristična po obilnim simetričnim masnim naslagama u području vrata i ramena. Prikaz slučaja 69-godišnjeg bolesnika, čiji su simptomi klinički razvijene Madelungove bolesti zamijenjeni sa simptomima difuzne strume štitne žlijezde i kronične srčane slabosti, pa je tako tri godine liječen. Iako se dijagnoza Madelungove bolesti može postaviti već nakon kliničkog pregleda, cilj ovoga prikaza slučaja je još jednom podsjetiti na moguće dijagnostičke i terapijske pogreške, ako se na ovu rijetku bolest ne misli u postupku diferenciranja simetričnih oteklina u vratu i ramenima.