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Case Report

Madelung's Disease and Upper Airway Involvement: A Case Report

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Abstract

Madelung's disease is a rare condition characterized by multiple and symmetrical deposits of adipose tissue concerning especially neck, shoulders, upper limbs and upper thorax. This lipomatous tissue could cause extrinsic compression or recurrent laryngeal nerve compression because of infiltration of fat [1,2].

In this case, we will describe a 66-year-old man affected by Madelung's disease, with larynx-tracheal anatomical alteration, showed by CT scan and flexible laryngoscopy.

Keywords: Madelung's Disease; Lipomatosis; Upper Airways; Larynx; Videorhinolaryngoscopy

Abbreviations

MD: Madelung's Disease; MLS: Multiple Symmetric Lipomatosis; CT: Computed Tomography; ER: Emergency Room

Introduction

Madelung's disease (MD), also known as Multiple Symmetric Lipomatosis (MSL) or benign symmetric lipomatosis, is a rare disorder, first described by Brodie in 1846 [3].

This condition mainly affects adult patients between 30 and 60 years of age, the ratio of men to women varies from 15/1 to 30/1 [4]. There is a familiar form linked to matrilineal inheritance [5], nevertheless the sporadic onset is the most recurrent disease form.

The pathogenesis of Madelung's disease is unclear, but a correlation with alcohol use disorder has been hypothesized: chronic alcohol abuse has been described in 60-90% of the reported cases [6,7]. Furthermore, a link between MD and other metabolic disorders, such as diabetes mellitus, hyperuricemia, hypothyroidism and liver disease, has been highlighted [8,9].

On clinical examination, we can describe multiple, symmetric, unencapsulated lipomas involving cheeks, neck, upper thorax, shoulder girdle area and arms [10,11].

Madelung's disease could be classified into three types, referring to the distribution of fat overgrowth. Type I is characterized by the accumulation of fat deposits predominantly in the upper body, such as the cervical region, posterior neck, shoulders and proximal upper limbs (or Madelung-Fetthals); type 2 with fatty tissue diffusely deposited in the shoulder area (or pseudo-athletic type); in type 3 the localization is in the pelvic region (or gynaecoid type) [12,13].

Most patients with Madelung's disease are usually asymptomatic, although some tenderness or pressure is present in few patients (13). Otherwise, the fat might accumulate at the trachea or larynx [1], especially for lipomatous distribution of Type 1, which could cause compression of the upper respiratory tract, dyspnea and dysphonia [1,2].

Madelung's disease may be easily diagnosed on the basis of history, clinical appearance, and radiologic examination, such as CT (2), which is useful for disease diagnosis, but mainly for the follow up of the adipose tissue deposition evolution.

Case Report

A 66-year-old man was admitted to the Internal Medicine and Gastroenterology, after ER evaluation for the onset of abdominal pain. He had already been diagnosed with Madelung's disease, type I, with multiple adipose deposits in the neck area and in the proximal upper limbs. His history was significant for alcohol consumption (at admission he referred to two years abstinence), ischemic heart disease, hypertension and HBV infection.

Figure 1: These figures show (A) front and (B) right side views of patient with type I Madelung's disease and lipomatous tissue involving the neck area.

The patient was alert and afebrile and the abdomen was palpable. Abdominal ultrasound scans and x-rays did not show acute injury. Serum tests showed a picture of hyponatremia (Na 10^2 mmol/L), thus Tolvaptan and Sodium (physiological solution 400 ml and 12 NaCl phials in 24 hours) were administered.

At admission, the patient was asymptomatic for dyspnoea and dysphagia; however, a CT scan was executed to exclude a compression of the upper respiratory tract by lipomatous deposits.

Neck and thorax CT scans showed high thickness of the prevertebral adipose tissue, located posteriorly to the oropharynx, the hypopharynx and the supraglottic region with a reduction of the aerial column at this level; the pyriform sinuses appeared obliterated. The trachea had elongated morphology in its proximal and middle tract, and it was slightly laterally deviated to the right with a wide imprint on the left lateral wall, with a modest reduction in caliber of the aerial column.

Figure 2: Sagittal CT scan view of adipose tissue posterior to the oropharynx, the hypopharynx and the supraglottic region.

Figure 3: A-B Coronal CT scan view of adipose tissue deposits diffused in the neck area. The trachea is slightly laterally deviated to the right.

Then, videorhinolaryngoscopy was performed, and it highlighted the patency of the nasal and rhinolaryngeal pathways, with the conservation of the respiratory space and normal pharyngolaryngeal motility. The laryngotracheal axis was deviated clockwise. Macroglossia was also observed, with oropharyngeal space preserved and normal veil motility.

Figure 4: The upper airway respiratory space is conserved at videorhinolaryngoscopy.

A cardiorespiratory polygraphy was performed, due to a likely malposition of the device it did not reveal any air flow records. However, percutaneous nocturnal oximetric monitoring revealed a picture compatible with nocturnal respiratory failure with 92% base saturation. Oxygen therapy was therefore set with VentiMask (FiO_2 at 24%) at night.

Discussion

Madelung's disease, especially Type 1, can occur with obstruction of the upper airways and with OSA, as a combination of different mechanisms, such as mechanical compression from the outside at the trachea and larynx level and the fat infiltration of upper airways structures [14].

In the case examined, the patient was asymptomatic for dyspnoea; however, the CT scan showed a significant increase in the deposition of fat in the posterior region of the neck and, specifically, posteriorly to oropharynx, hypopharynx and supraglottic regions, without highlighting a direct invasion of the previously mentioned structures. There was also a tracheal deviation with a reduction in its caliber. By endoscopic examination, the tracheal axial deviation was confirmed, with clockwise rotation, without a clinically relevant reduction of the respiratory space at the laryngeal level, in accordance with the absence of the patient's symptoms.

The dynamic polygraphy was therefore mandatory, allowing through percutaneous oximetric monitoring to highlight the picture of nocturnal respiratory insufficiency with the presence of pathological desaturations in pathological numbers. According to the evidences of Oliven., *et al.* [15] the main mechanism that determines the laryngotracheal collapse is the increase of the surrounding pressure, which, in the case reported, is due to the presence of the plenty lipomatous tissue described by the CT scan.

There is no drug treatment for the reduction of adipose tissue excess; the surgical treatment of extensive lipectomy remains the standard treatment of Madelung's disease.

However, the intervention to remove adipose tissue, even massive, only rarely improves nocturnal respiratory symptoms, probably due to the difficulty in reaching the parapharyngeal space [14].

In asymptomatic patients, follow-up by imaging and functional tests could be considered to evaluate lipomas growth, upper airway compression and the appearance of symptoms.

In symptomatic patients, first line treatment is the application of continuous positive airway pressure (CPAP). In patients who cannot tolerate ventilatory therapy, a specific surgical intervention such as Functional expansion pharyngoplasty could be considered [15].

Conclusion

The possibility of the upper airways obstruction should always be considered in patients with Madelung's disease, especially in Type 1. Also, in asymptomatic patients, a functional and imaging evaluation should always be carried out to identify patients at risk or those who already have obstructive problems, and also to implement the necessary measures for effective treatment. In this regard, CPAP remains the first line treatment, without excluding the possibility of specific surgical treatment in patients who cannot tolerate functional therapy [14].

Conflict of Interest

There are no conflicts of interest.

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