CASE STUDY

Laryngeal Lipoma Associated With Madelung’s Disease: A Case Report

Guillermo Arturo Landínez-Cepeda,* Emilio V. Alarcos-Tamayo, Teresa Millás-Gómez, Darío Morais-Pérez

Servicio de Otorrinolaringología y Patología Cervico-Facial, Hospital Clínico Universitario de Valladolid, Spain

Abstract  Multiple symmetric lipomatosis is an alteration in the neck, upper trunk and upper extremities fat deposits. It produces an aesthetic problem and sometimes upper airway obstruction when the larynx is infiltrated by the mass. We report the case of a male with Madelung’s disease, which began with acute dyspnoea caused by laryngeal fat deposits and obstructive lipoma. © 2010 Elsevier España, S.L. All rights reserved.

Lipoma laringeo asociado a enfermedad de Madelung: a propósito de un caso

Resumen  La lipomatosis cervical múltiple es una alteración del depósito graso cervical, parte alta de tórax y extremidades superiores que genera un problema estético y eventualmente cuadros obstructivos de la vía respiratoria en aquellos casos de infiltración grasa de la laringe. Presentamos el caso de un hombre con enfermedad de Madelung y cuadro disneico agudo por infiltración grasa laringea y lipoma obstructivo. © 2010 Elsevier España, S.L. Todos los derechos reservados.

Introduction

The association of multiple cervical lipomatosis and laryngeal infiltration with intrinsic lipomas is rare, with only 3 cases reported in the medical literature. We draw attention to this fact and the possibility of dyspnoea symptoms by laryngeal obstruction due to lipomas associated with Madelung’s disease.

Clinical Case

We present the case of 48-year-old male with a drinking habit, who smoked over 20 cigarettes/day and who attended the emergency service due to a worsening in his “usual fatigue”. The patient presented inspiratory dyspnoea with stridor, supraclavicular retractions and a marked thickening of the neck which led us to request an emergency CT scan (Figs. 1 and 2). The scan revealed the presence of multiple tumours, scattered throughout the neck, with fat density and infiltrating deep tissues, compressing the upper airway up to the sternal notch. Laryngeal examination by fibroscopy found a reddish-yellow mass of 3 cm in diameter, which pedicled on the glottic lumen from the right arytenoepiglottic fold and obstructed...
it intermittently, according to breathing movements. We performed an emergency tracheotomy and subsequently removed the laryngeal tumour by microlaryngoscopy. The anatomopathological report was of lipoma. The patient recovered uneventfully and the tracheostoma closed a few days later. At 4 months we conducted a bilateral cervicotomy to remove the cervical lipomas, which improved the physical appearance and reduced the sensation of cervical compression.

Discussion

Madelung’s disease, Launois-Bensaude syndrome or multiple symmetric lipomatosis is an increasingly rare entity, with over 300 cases reported in the medical literature. It consists in the infiltration by unencapsulated fat of the anterior region of the neck, the proximal regions of the upper limbs, chest and facial region. It can cause cosmetic deformity, extrinsic vascular compression symptoms, upper airway compression symptoms and, more rarely, compression of the recurrent laryngeal nerve by fatty infiltration, leading to dyspnoea and dysphonia. There are 2 types; type I is more localised and manifests in middle-aged Mediterranean men with a history of alcohol intake in 60%-90% of cases, metabolic disorders such as hyperuricemia, diabetes, goitre, and hypertension, whereas type II is more diffuse and more common among women with a high body mass index. Presumably, its aetiology involves a local defect in lipolysis induced by catecholamines and the lipogenic and antilipolytic effect of alcohol. It displays greater susceptibility towards developing squamous cell carcinomas of the upper aerodigestive tract in alcoholics and smokers, related to these risk factors.

At present, there have been about 115 reported cases of isolated fat infiltration in the larynx, representing 0.6% of benign tumours of the head and neck. However, the association of Madelung’s disease with the appearance of intrinsic laryngeal lipomas is more uncommon, with only 3 reported cases in the specialised literature. The diagnostic approach is based on examination and palpation of the fatty deformity. A CT or MRI imaging study may show densities and intensities similar to those of normal fat infiltrating cervical spaces and structures. Laryngeal infiltration and the possibility of developing a lipoma which occludes the lumen should be suspected upon observing symptoms of inspiratory dyspnoea with stridor. In such cases, airway preservation and diagnostic laryngeal endoscopy, along with excision of the tumour, should be the attitude to follow with urgency. Surgical resection of cervical lipomas by cervicotomy would be indicated in cases of extrinsic vascular compression and upper aerodigestive tract compression with visceral infiltration, while liposuction should be reserved for cases of strict cosmetic deformity without infiltration of important structures.

Conclusion

We wish to draw attention to Madelung’s disease and its possible association with laryngeal lipomas. When this happens, symptoms of subacute dyspnoea may appear which may require, as in our case, rapid action including tracheotomy and subsequent excision of the lipoma obstructing the laryngeal lumen.

Conflict of Interests

The authors have no conflicts of interest to declare.

References