CASE REPORT

Symptomatic retropharyngeal space lipoma. A patient with Madelung disease

Chrysovitsiotis G, Papanikolaou V, Kyrodimos E, Giotakis E

1st E.N.T. Department of Faculty of Medicine, National and Kapodistrian University of Athens, Athens, Greece

Abstract

Background: Retropharyngeal space lipomas (RSL) are rare benign tumors of the head and neck region. They can, sporadically, occur as part of syndromic lipomatosis, such as Madelung disease. Symptoms are caused due to increasing pressure on surrounding structures.

Description of case: We present a 64-year-old male patient with symptomatic RSL and symmetric lipomatosis, who was treated surgically.

Conclusion: RSL can grow to a large size before becoming symptomatic. Their diagnosis and treatment can be challenging due to their anatomical site, diverse symptomatology, and diffuse growth pattern. Imaging is necessary for diagnosis. In the vast majority of cases, RSLs are treated surgically with a favorable outcome. HIPPOKRATIA 2020, 24(2): 91-93.

Keywords: Lipoma, retropharyngeal space, transoral excision, Madelung's disease

Corresponding Author: Georgios Chrysovitsiotis, Otorhinolaryngologist-Head and Neck Surgeon, 114 Vassilissis Sofias Avenue, 11527, Athens, Greece, tel: +302132088030, fax: +302132088402, e-mail: chrysovi@gmail.com

Introduction

Lipomas are benign tumors of mesenchymal origin, which develop slowly and demonstrate a non-infiltrating behavior. About 13 % of these occur in the head and neck region, most commonly subcutaneously at the posterior neck[1]. Of these posterior neck lipomas, the retropharyngeal space lipomas (SPL) are very rare. Clinical presentation is rather atypical; symptoms are caused due to increasing pressure on surrounding structures[2].

Lipomas of the head and neck region may occur as part of syndromic lipomatosis[3]. One of these lipomatous syndromes is multiple symmetrical lipomatosis, also called Madelung's disease or Launois-Bensaude syndrome. This condition consists of subcutaneous fatty deposits mainly in the head, anterior and posterior neck, upper torso, and upper extremities. Patients are usually males in their third to seventh decades of life[4]. Prevalence of the disease seems to be higher in the Mediterranean countries[5]. Furthermore, a strong connection is established in the literature between Madelung's disease and moderate to heavy alcohol consumption[4-6].

Description of case

A 64-year-old man was hospitalized and subsequently intubated due to acute respiratory failure. The attending doctors reported a low-pitched stertor before intubation, which was also present after extubating the patient. The patient reported a history of nocturnal snoring over the preceding ten years and mild hoarseness for the latest three years. These symptoms were reported as stable, without worsening or exacerbations. Furthermore, the patient suffered from mild daytime somnolence over the preceding two years and progressive difficulty swallowing solid food for six months before hospitalization. The patient's medical history consisted of hypertension, chronic restrictive lung disease, and alcohol abuse of approximately five Units per day.

During flexible rhinopharyngolaryngoscopy, a protrusion of the posterior pharyngeal wall was noted, with its maximum extent at the vallecula level. The rest of the hypopharynx and larynx appeared normal. On palpation of the neck, multiple symmetrical, soft, mobile, subcutaneous masses were found. Similar lesions on the patient's torso and upper extremities were also noted on further examination. The patient reported that he was diagnosed with lipomatosis before approximately 20 years. Given his alcohol consumption history and the clinical findings, benign symmetrical lipomatosis (Madelung's disease) was suspected.

Further investigation of the neck masses was performed initially by computed tomography with intravenous contrast and subsequently by magnetic resonance imaging with paramagnetic contrast agent. The scans revealed various fatty-tissue density masses in the neck. Furthermore, a large mass, partially obstructing the upper aerodigestive tract, was located slightly on the midline right, extending from the level of the soft palate to the vallecula (Figure 1).

Given the tumor's size and location, a transoral excision was suggested. The clinical condition, including the possibility of an emergency tracheotomy, was explained to the patient and informed written consent was obtained.

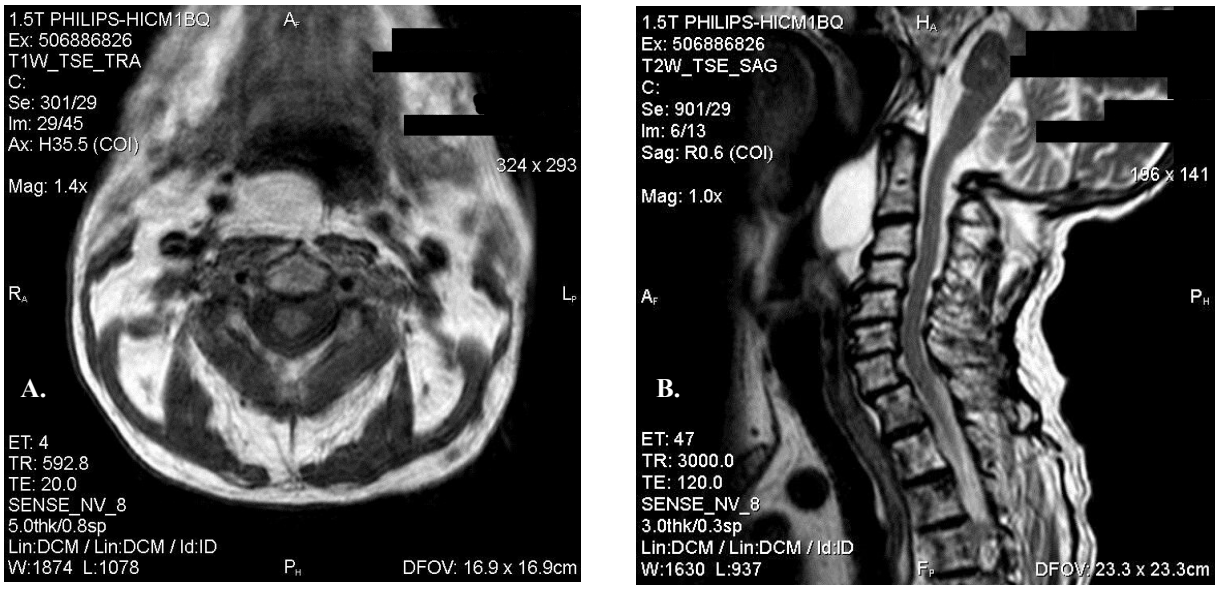

Figure 1: Preoperative magnetic resonance imaging of the reported case, A) axial T1-weighted image at the level of hypopharynx showing a lesion at the retropharyngeal space slightly on the midline right, B) sagittal T1-weighted image of the neck showing a high signal lesion at the retropharyngeal space extending from the level of the soft palate to the vallecula.

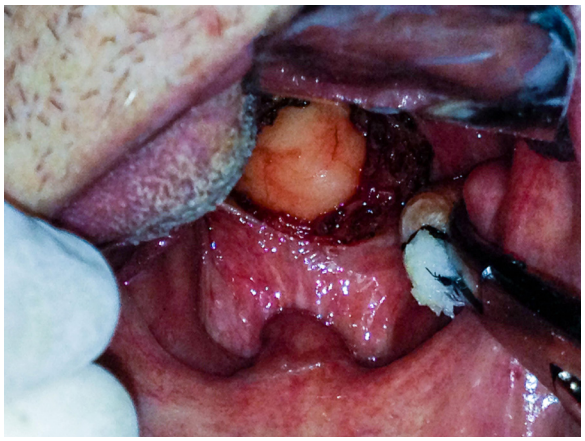

Figure 2: Intraoperative image showing adequate exposure of the yellowish tumor through a vertical incision of the posterior pharyngeal wall.

Figure 3: Intraoperative image showing the surgical wound's primary closure with interrupted sutures in layers, after *in toto* excision of the tumor.

In the operating theatre, the patient was placed in Rose's tonsillectomy position; a Boyle-Davis gag was inserted in the oral cavity facilitating the posterior pharyngeal wall's adequate exposure. A vertical incision of approximately 4 cm was performed, and the constrictor muscle was divided, providing easy access to the tumor, which was then bluntly dissected from the surrounding tissues (Figure 2). The tumor was excised *in toto*, and the wound was primarily closed with Vicryl 4-0 interrupted suture in layers (Figure 3). The patient had an uneventful postoperative recovery. Histopathologic examination of the excised tumor confirmed the preoperative diagnosis of a non-infiltrating lipoma, sized 4.5 x 3 x 1.5 cm.

His stertor presented marked improvement in the postoperative period, and he was fed the second postop day orally. The patient was discharged on the third postop day. During follow-up examination in one week, two weeks, and six months, further improvement of his swallowing, stertor, and voice were noted. Despite a slight improvement in snoring, no changes regarding daytime somnolence were reported.

Discussion

Lipomas of the head and neck region can uncommonly occur due to syndromic causes of lipomatosis and exhibit symptoms resulting from a cumulative pressure effect. In the literature, laryngeal or pharyngeal involvement was described in 19 cases of Madelung disease[7], with only six cases of RSL reported[8]. Furthermore, it should be noted that only in one case of a patient with Madelung disease,

the symptoms were caused by the RSL itself, rather than the symmetrical lipomatosis in the neck[9].

RSLs, solely or as part of syndromic lipomatosis, are a challenging entity mainly because they remain asymptomatic until they obtain a considerable size. The symptoms reported by the patients are attributed to pressure caused by the lipomas, and they tend to overlap. The most common symptom reported in the literature is dysphagia, while dyspnea, dysphonia, snoring, and stertor have also been reported.

A high level of suspicion is required to include this entity in the differential diagnosis. Malignancy, abscess, hematoma, and other benign tumors of the retropharyngeal space must be excluded. Imaging is an indispensable method in order to differentiate between potential diagnoses. Moreover, it provides valuable information regarding preoperative planning. Surgical excision is almost always selected as the treatment modality, and full recovery is expected.

Conflict of interest

Authors declare no conflicts of interest.

Acknowledgment

This case was reported in abstract form, during the 19th Panhellenic Congress of Otolaryngology – Head and Neck Surgery, Athens, November 2017.

References

1. Som PM, Scherl MP, Rao VM, Biller HF. Rare presentations of ordinary lipomas of the head and neck: a review. AJNR Am J Neuroradiol. 1986; 7: 657-664.
2. Haddad FS, Zaytoun G, Haddad FF. Retropharyngeal lipoma, a benign yet potentially lethal condition: Case presentation and review of the literature. Neurosurg Q. 2005; 15: 145-154.
3. Tadisina KK, Mlynek KS, Hwang LK, Riazi H, Papay FA, Zins JE. Syndromic lipomatosis of the head and neck: a review of the literature. Aesthetic Plast Surg. 2015; 39: 440-448.
4. Ramos S, Pinheiro S, Diogo C, Cabral L, Cruzeiro C. Madelung disease: a not-so-rare disorder. Ann Plast Surg. 2010; 64: 122-124.
5. Meningaud JP, Pitak-Arnnop P, Bertrand JC. Multiple symmetric lipomatosis: case report and review of the literature. J Oral Maxillofac Surg. 2007; 65: 1365-1369.
6. Ruzicka T, Vieluf D, Landthaler M, Braun-Falco O. Benign symmetric lipomatosis Launois-Bensaude. Report of ten cases and review of the literature. J Am Acad Dermatol. 1987; 17: 663-674.
7. Ojo B, Jang DW, Gurudutt VV. Case Report of Madelung's Disease with Upper Airway Obstruction. Laryngoscope. 2011; 121: S177.
8. Zhang XY, Li NY, Xiao WL. Madelung disease: manifestations of CT and MR imaging. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2008; 105: e57-e64.
9. Ali S, Kishore A. Dysphagia and obstructive sleep apnoea in Madelung's disease. J Laryngol Otol. 2007; 121: 398-400.