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Parapharyngeal space lipomatosis with secondary dyspnea, disphagia and disphonia

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ABSTRACT

INTRODUCTION: Parapharyngeal space tumors are a small percentage of all head and neck neoplasms. Due to their anatomic location, they represent a therapeutic challenge. To our knowledge, 11 cases of parapharyngeal lipomatous tumors have been reported in the literature.
CASE: A 48 year old male with chief complaints of dyspnea, dysphagia and dysphonia was found to have a parapharyngeal space tumor. He was scheduled to undergo lumpectomy and neck exploration.
DISCUSSION: Benign tumors represented 70% of all cases. Open neck surgery is considered the gold-standard of treatment.
CONCLUSION: It is important to bear in mind the lipomas of the parapharyngeal space to establish an accurate diagnosis and implement timely, appropriate treatment in order to avoid future complications and reduce morbidity and mortality.

1. Introduction

The parapharyngeal space is a region with complex anatomical relationships. It can give rise to a large variety of benign and malignant neplasms. Tumors arising in the parapharyngeal space are uncommon, and account for only 0.5% of all neoplasms of the head and neck. Tumors arising in the parapharyngeal space are often asymptomatic. Symtomatology presents often after the tumor has reached a size of about 2.5 cm or more. At this point, the associated symptoms largely depend on the affected specific location; hence, the pre-styloid location can be associated with serous otitis media, voice change, nasal obstruction, aspiration, or dyspnea. This last symptom appears to have been abated in our patient, once the operation had been carried out. In the poststyloid compartment, the tumor may compress the 9th, 10th, 11th, or 12th cranial nerve, causing hoarseness, dysphagia, dysarthria, or Horner's syndrome, as the result of tumor pressure on the superior cervical sympathetic ganglia [1].

Parapharyngeal space tumors are usually benign neoplasms (70%), with the rest (30%) being malignancies. Salivary gland tumors are the most common origin, usually arising from the deep lobe of the parotid gland and from minor salivary glands; neurogenic tumors are the second most common neoplasms. In our experience in 21 patients, 8 men and 13 women, average age of 41 years (range, 20–70 years) in ten years, the neurogenic tumors represented 57%, salivary gland tumors 33%, and sarcomas 10% [1]. However, the most common vascular tumor in the parapharyngeal space continues to be the carotid body tumor [2].

For this case, two entities should be considered: (1) tumors of adipose tissue origin, the most common variant being lipoma, a benign tumor most often found in soft tissue, but which is rare in the head and neck region (less than 10%) [3–5] and (2) benign symmetric lipomatosis, also known as Madelung–Launois disease [6].

2. Case report

A 48 year old man with a past medical history of chronic alcohol abuse and recently diagnosed hypertension had a chief complaint of progressive increase in volume of the right side of the neck, which started 8 months before. Associated symptoms included dyspnea and dysphagia (dysphonia is mentioned earlier, add it if pertinent). A physical examination confirmed an increase in volume in the right neck (Fig. 1). A nasofibrolaryngoscopy showed decrease in the secondary upper airway due to extrinsic compression of the pharynx and hypopharynx, thus hindering proper observation of the larynx and causing an obvious obstruction of the airway. A poorly defined echogenic image was documented by ultrasound (US) through the entire course of the right sternocleidomastoid muscle, and some minor lymph nodes under 1 cm were observed. A fine needle aspiration(FNA) cytology, revealed scarce fragments of adipose and fibrous connective tissue on a

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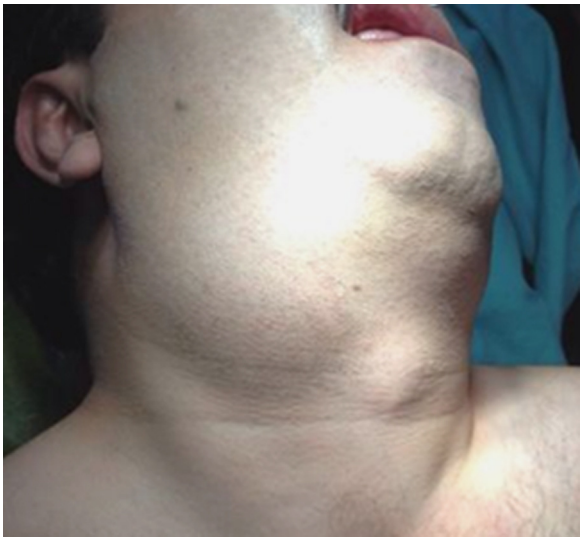

Fig. 1. An increased neck girth (right side) is noticed on this 48-year old man. The mass was associated with dyspnea, dysphonia, and dysphagia.

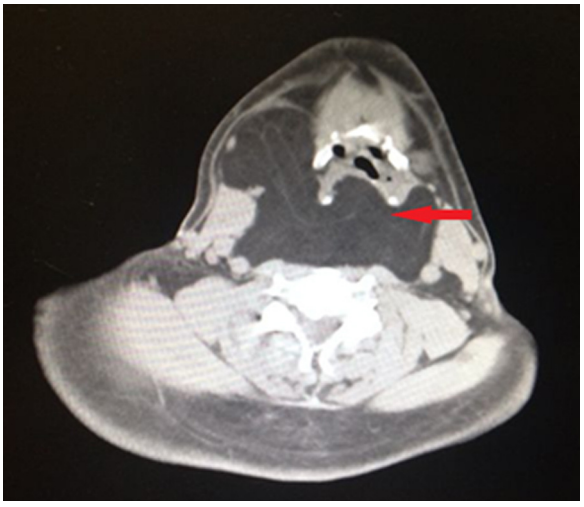

Fig. 2. Computed tomography (CT) showing hypodense image with -112HU, in the left and right parapharyngeal region, with displacement of the pharyngeal wall, causing less light in the upper airway, as far as the hypopharynx.

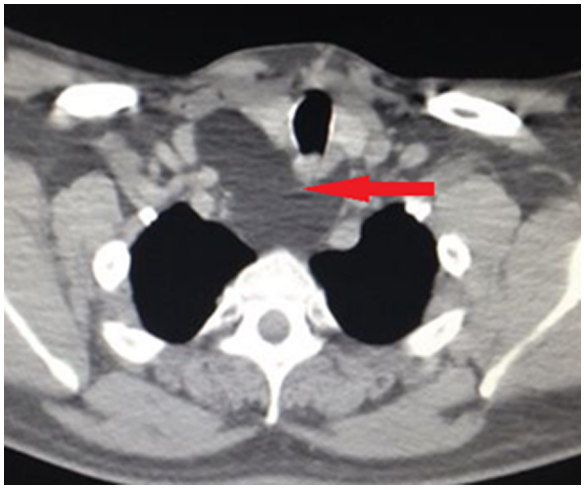

Fig. 3. CT showing the extent of the lesion in both upper mediastinum and towards both supraclavicular regions.

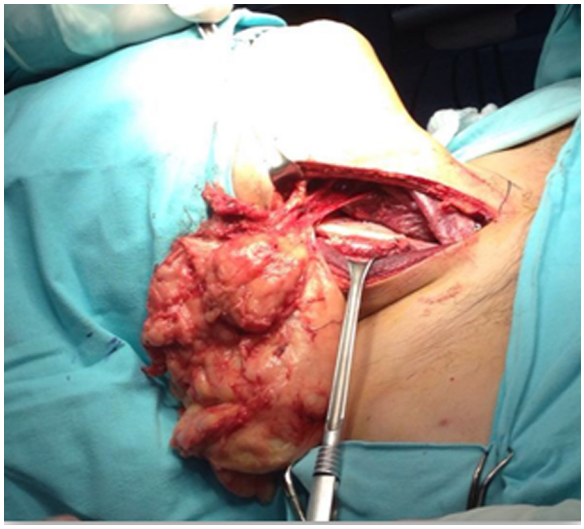

Fig. 4. Lateral cervical focus with view of lipomatoous lesion with areas of soft tissue arranged in lobules (10 ×8 cm), right parapharyngeal space, extending to the left side, displacing the posterior wall of the pharynx and hypopharynx towards the upper mediastinum.

hemorrhagic background were observed. A computed tomographic (CT) of the neck showed a hypodense image, with a coefficient of attenuation of -112HU, in the right parapharyngeal region extending to the hypopharynx (Fig. 2), superior mediastinum, and far as both supraclavicular regions (Fig. 3), with anterior displacement of the submandibular gland and displacement of the extrathoracic portion of the left-sided airway.

A lumpectomy with cervical exploration was carried out, revealing a subplatysmal lobulated mass, suggestive of lipomatous origin (10 × 8 cm), in close contacts with the right and left parapharyngeal regions and extending as far as the hypopharynx and superior mediastinum, as well as to both supraclavicular regions (Fig. 4); together, with the carotid vessels and the vagus nerve, it was necessary to ligate the right anterior and external jugular veins and the thyrolinguofacial trunk in order to resect the neoplasm. The post-surgical progress of the patient was uneventful; the dyspnea/dysphagia were immediately resolved, and he continued follow up every 24 h in our outpatient clinic.

Final pathology was consistent with a capsulated 160 × 120 × 25 mm lipoma showing some steatonecrotic area in its context but no sign of malignancy (Fig. 5). The diagnosis of benign lipoma was confirmed also by immune-histochemical analysis (S100 protein positive, p53 and MDM2 negative, Ki67 1%).

3. Discussion

The majority of parapharyngeal space tumors are benign neoplasms (70%), including: neurogenic tumors, chemodectomas, branchial cysts, goiter, carcinomas, cervical cysts, lifoproliferativas diseases, multiple familial lipomatosis, painful lipomatosis (Dercum syndrome), Touraine and Renault's Lipomatosis, Verneuil and Potain's Pseudolipomatosis, hibernoma, and angiolipoma. In this case, the differential diagnosis in this case included two entities: Madelung–Launois disease (benign symmetric lipomatosis) [6], and lipoma. Benign symmetric lipomatosis is an entity of unknown etiology, Kodish [7] stated that it was the result of hypertrophy of brown adipose tissue; it was first described in 1846 by Benjamin Brodie [8], and subsequently by Otto Madelung in 1888 and a decade later Launois and Besaunde [8] presented two case studies of 35 and 30 patients respectively; it predominantly affected males with reports emerging that refer to a ratio of 15:1, with a range of age of onset between 20 and 60 years; a pattern for family

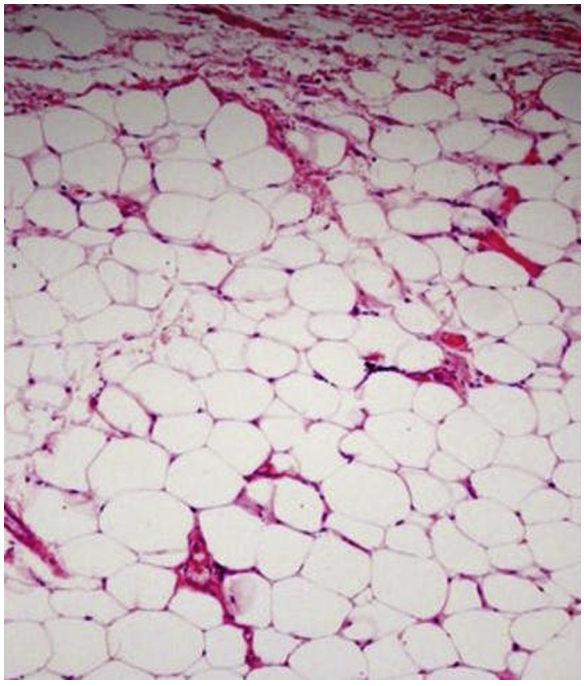

Fig. 5. Histological pattern showing well differentiated adipocytes with no signs of infiltration.

history has not yet been discovered and it is considered to be a sporadically occurring entity; there are some reports of family history, with dominant autosomal patterns of inheritance [6].

Lipomas constitute a very rare tumor in the parapharengeal space, as previously only 11 cases had been reported in the literature, presenting features which may include symptoms of pressure, with signs such as obstructive dysphagia and airway obstruction, sleep apnea manifested as dyspnea,similar to the present case [3–5,9,10]. Associated symptoms for both conditions are variable and depend on their anatomical location, although most are asymptomatic.

Open neck surgery is considered the gold standard for treatment, however recent reports of new technologies such as TORS (Transoral Robotic Surgery) [11] are beginning to be recognized as apt for particular tumor cases in the parapharyngeal space; it is curious that until the arrival of TORS, transoral approaches were considered a poor choice for the management of these tumors [1,9], thus this has still not surpassed open surgery, because experience for TORS is limited and does not appear to be adequate for tumor treatment, even in a case like the one presented here. Diagnosis of these ongoing lesions continues to be carried out applying computed tomography and/or magnetic resonance, with no differentiation between the two; definitive diagnosis is always confirmed by a study using FNA cytology; in our case we carried out computed tomography and confirmation with FNA was considered sufficient for treatment to be implemented. As in this case, many soft tissue tumors present a slow and gradual phase, followed by rapid growth, which in many cases is associated with a particular trauma and/or surgical procedure.

Most lesions are asymptomatic, the presence of dyspnea and dysphagia is extraordinarily rare and is secondary to the effect of mass on aerodigestive tracts, but should always be suspected when associated with some other neoplasia such as thyroid or tongue cancer among others; transformation to liposarcoma is extremely rare.

4. Conclusion

It is important to keep the different differential diagnoses regarding lipomatous tumors of the parapharyngeal space to implement the adequate, timely treatment, which in turns prevents future complications and reduces morbidity and mortality.

Conflicts of interest

There is no conflict of interest.

Funding

There is no sponsor.

Ethical approval

No this is a research study.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request".

Author contribution

Lead author together with co-authors performed surgical intervention and review of the literature.

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