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Case Report

Hypopharyngeal liposarcoma, a Rare and Uncommon Entity, a case report

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Abstract

Liposarcoma is the most common form of soft-tissue sarcoma, only 3-8% have a head and neck location. They are commonly found in the retroperitoneum and on the limbs, laryngeal and hypopharyngeal presentation are extremely rare and less than 40 cases have been reported in literature. We present the case of a 50-year-old man with no prior history, who developed two contiguous hypopharyngeal liposarcomas.

keywords: liposarcoma, hypopharynx, dysphagia, rare entity

Introduction

Liposarcoma is the most common form of soft-tissue sarcoma, only 3-8% have a head and neck location. Liposarcomas are malignant tumors of adipose tissue that rarely have a metastatic evolution but can recur locally [1, 2].

They are commonly found in the retroperitoneum and on the limbs, laryngeal and hypopharyngeal presentation are extremely rare [3] and less than 40 cases have been reported in literature [4]. Their evolution is slow and painless with progressive difficulty swallowing, dyspnea and, or dyspnea. They mainly occur in men between with a median of 64 years old[5]. The diagnosis is suspected based on the endoscopic and radiographic aspects of a polypoid, pedunculated mass with adipocytic density. The final diagnosis is histological with FISH amplification or immunochemistry detecting the abnormal amplification of MDM2 and CDK4, which are located on chromosome 12q13-15. The reference treatment is the surgical excision ad integrum with tumor free margins. Adjuvant radiation is controvertes[3].

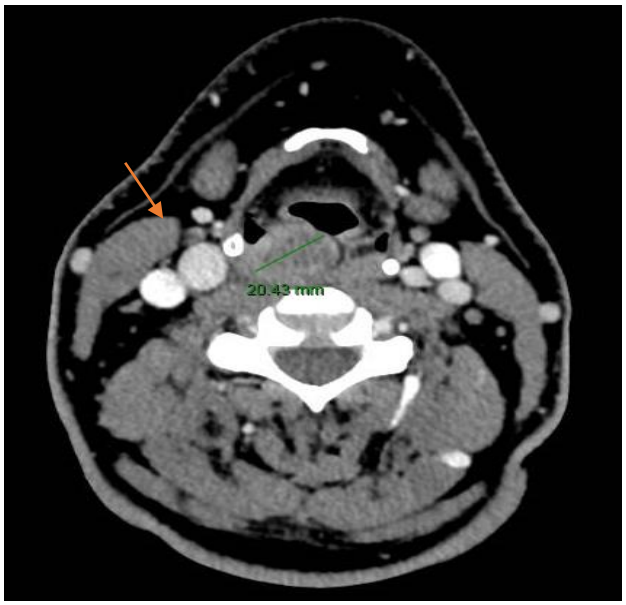
We report a case of well-differentiated hypopharyngeal liposarcoma, in which a diagnosis of a simple lipoma was initially proposed.

Case report

A 50 years old man presented with chronic dysphagia and regurgitations. He had no prior history of chronic disease, no treatments and no history of tobacco or alcoholic intoxication either. He was in an excellent general condition with no recent loss of weight. He denied throat pain or dyspnea but endorsed frequent oral regurgitation of solids, food blockage and frequent coughing spells.

The nasofibroscopy examination revealed a benign looking ovoid mass measuring 4cm, attached to the posterior pharyngeal wall, overhanging the esophagus mouth, non-hemorrhagic, flexible, with normal mucosa aspect. The cervical examination showed no lymphadenopathies.

The contrast-enhanced neck and chest computed tomography revealed a 2.5cm circumscribed adipose tumor of the hypopharynx, without reaching the larynx and the retro-pharyngeal spaces, no lymph node was associated (Figure 1)



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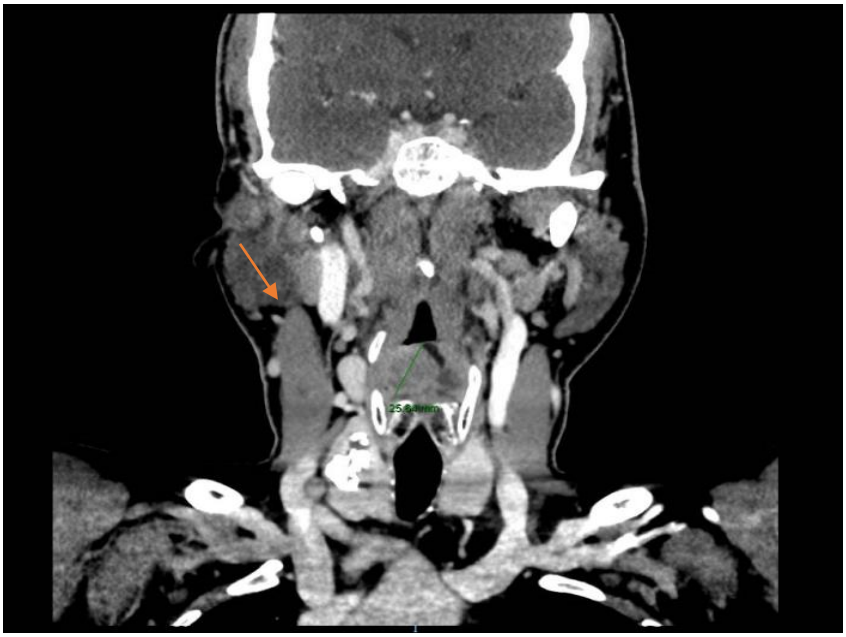

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Figure 1: *Contrast-enhanced CT workup. Axial (A) and coronal (B) views show a well delineated fat-delineated mass arising from the posterior wall of the pharynx*

A panendoscopic examination under general anesthesia with biopsies was performed. The anatomopathologic results were in favor of a lipoma. Unfortunately, the patient kept experiencing dysphagia with an aggravation of the regurgitations.

We decided to perform an endoscopic surgical resection. The surgery consisted of a complete resection of the mass by LASER dissection, performed off of the tumor (Figure 2). A second tumor adjacent to the first one was removed during the same operation.

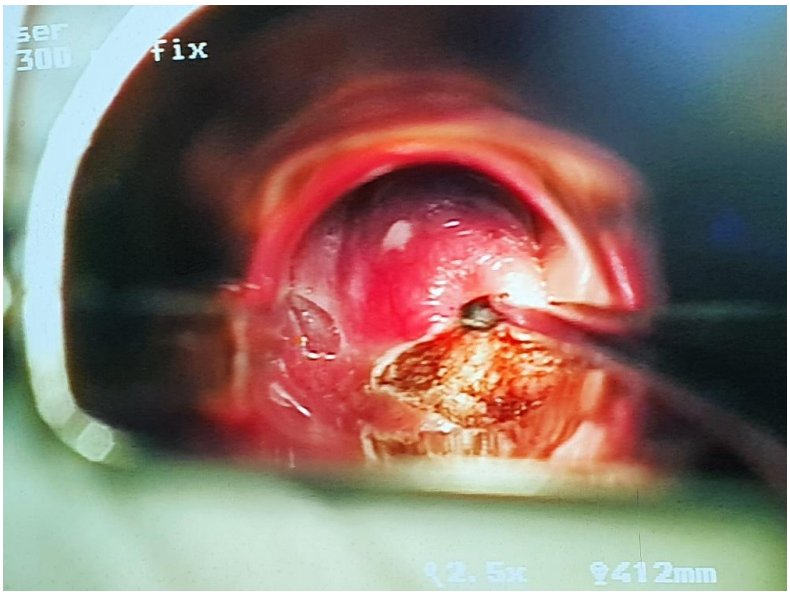

Figure 2: *Endoscopic surgical laser resection of the liposarcoma*

Histological examination concluded on two well-differenciated sclerosing liposarcomas, composed of mature adipocytes and CD34+, HMGA2+ spindle cells with positive MDM2 gene amplification. The resection was complete with tumor free margins (Figure 3).

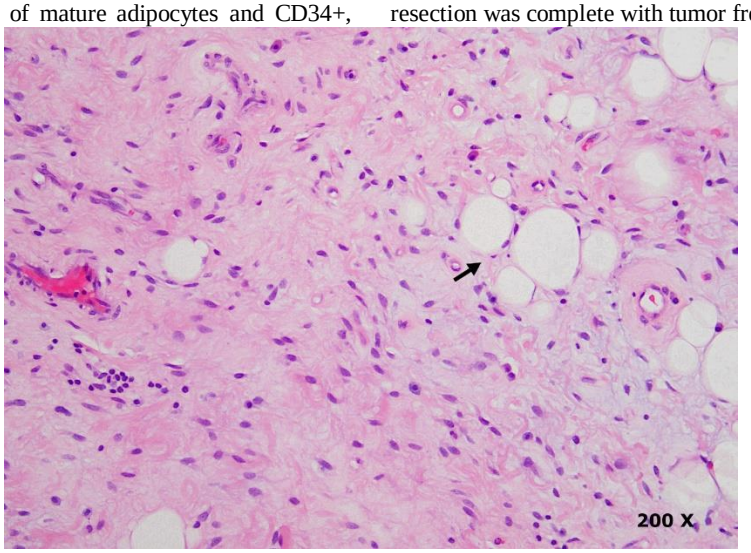

Figure 3: *Nodular but non-encapsulated neoplasm with low to middle cellularity. The cells are spindled with bland nuclear features. The stroma is collagenic, sometimes myxoïd, with lymphocytic infiltrate. Note the mature adipocytes (arrow).*

The one-month and the six months follow-up the patient was asymptomatic. The endoscopies revealed a good healing of the mucosa with no sign of recurrence. The cervical neck palpation was free of lymphadenopathies. The CT-scan at 6 months showed no signs of recurrence either.

The case was reported to a specialized multidisciplinary reunion, which decided on a biannual clinical and CT surveillance.

Discussion

Liposarcomas are malignant [1] tumors of adipose tissue that are relatively common in adults, they account as 15-18% of all sarcomas.

The hypopharyngeal localization is extremely rare, with only 28 case reports referenced in Pubmed in 2017, and less than a hundred cases have been reported in literature [6].

Clinically, the symptoms are related to the localization adjacent to the pharyngolaryngeal crossroad, with organic dysphagia, sensation of foreign body, dysphonia or dyspnea due to airway obstruction. A nasal endoscopy performed at the consultation shows a sub-mucosal tumor, with respect of the mucosa, which might obliterate the pharyngolaryngeal space [7].

CT or MRI examination reveals one or more well-limited tumor, with fat density. The MRI is more accurate than the CT-scan to delineate tumor boundaries before surgical work-up [4]. The liposarcoma can be easily confused with a benign or malignant tumor of the pharyngolaryngeal space, such as lipoma, fibroma, carcinoma, sarcoma, and lymphoma. The MRI can help to distinguish lipomas from liposarcomas by showing a mosaic heterogeneity in liposarcomas whereas lipomas are preferentially homogenous [1].

The histological staging proposed by Ezinger and Winslow in 1962 [8] is considered as the reference for liposarcomas. Four histological types of liposarcoma are described: well-differentiated, mixoid or round cell, pleomorphic, and dedifferentiated liposarcoma [6]. The most common (40-45%) is the well differentiated, a low-grade lesion with three subtypes: the lipoma-like liposarcoma, the inflammatory and the sclerosing liposarcoma [3]. Liposarcomas do not develop from lipomas but rather from primitive mesenchymal cells[1]. FISH detection of MDM2 and CDK4 amplification distinguishes welldifferentiated and dedifferenciated liposarcomas from lipomas. If FISH technic is not availabale immunohistochemistry is a reasonable alternative [5].

The gold standard treatment is the surgical resection with a cervical or an endoscopic approach, to obtain free margins of resection [3,6]. The second option allows a minimal post-operative morbidity: rapid resumption of feeding, no need for a tracheotomy and decreased length of hospital stay. Systematic neck dissection is not indicated if there are no lymphadenopathies, the rate of nodal metastase is low [7].

The diagnosis established, the case has to be discussed in a specialized multidisciplinary meeting to decide of a potential adjuvant therapy. Adjuvant radiation might be discussed in cases of dedifferenciated liposarcomas or in cases of inoperable tulmors [3, 7].

Metastasis has not been reported and the long-term prognosis is excellent (with an over 90% 5-year survival) [5, 7]. The mortality rate depend on the histological type and location. However the main risk is the recurrence, it is frequent and can occur at long term [6]. The main risk factor is the incomplete resection [9]. For all these reasons a long term follow up with clinical, CT or endoscopic surveillance is essential.

Compliance with Ethical Standards

There are no conflicts of interest for the authors in this study. The patient gave an informed consent.

Conclusion

The hypopharyngeal liposarcoma is a rare entity encountered in clinical practice. Despite this, the clinician has to think about this etiology and have recourse to a specialist opinion concerning the surgical management of this tumor and the follow up.

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