Cellular angiofibroma in the hypopharynx
A case report
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Abstract
Rationale: Cellular angiofibroma is a rare benign mesenchymal tumor which mostly occurs in the superficial soft tissues of the genital region. Occurrence in the head and neck region is extremely rare. To our knowledge, this is the first case of cellular angiofibroma arising in the hypopharynx.

Patient’s concerns: A 54-year-old male complained of a mass moving through his throat for 1 month. A tumor was found in the right lateral wall of the pharyngeal at the level of the epiglottis by laryngoscope. Magnetic resonance imaging confirmed the presence of a mass in the right lateral pharyngeal area. A benign tumor was suspected based on the clinical symptoms and imaging findings.

Diagnosis: A supporting laryngoscope was performed under general anesthesia and the lesion was resected. Immunohistochemical analysis revealed cellular angiofibroma.

Interventions: The patient underwent surgical excision of the lesion.

Outcomes: Thus far, no recurrence has been observed 6 months after excision.

Lessons: Cellular angiofibroma located in the lateral pharyngeal is rare; however, immunohistochemical staining is helpful for its diagnosis. Treatment is relatively simple and requires local excision and follow-up.

Abbreviations: ER = estrogen receptor, MRI = magnetic resonance imaging, T1WI = T1-weighted images.

Keywords: benign tumor, cellular angiofibroma, hypopharynx

1. Introduction

Cellular angiofibroma was first described by Nucci et al in 1997 as a benign mesenchymal tumor commonly occurring in the vulvovaginal region of women and the inguinoscrotal region of men,[1] with very few occurring in the nasopharynx or retroperitoneal regions.[2–3] Cellular angiofibroma is rarely located in the head or neck, but has been reported previously in the nasopharynx.[1] Generally, the tumor presents as a slowly enlarging mass and tends to be asymptomatic, however, it may cause pain.[4] Chen inferred that cellular angiofibroma behaves in a benign fashion and has a tendency to recur after simple excision.[4]

Here, we present the case of a 54-year-old man who had a cellular angiofibroma attached to the lateral pharyngeal wall. In this report, we presented the characteristics, histopathologic features, and treatment. This is the reported first case of cellular angiofibroma in the hypopharyngeal region.

2. Case report

This study was approved by the institutional review board of the First Affiliated Hospital, College of Medicine, Zhejiang University, Hangzhou City, China, and the patient provided written informed consent for publication of the case.

A 54-year-old Chinese man was referred to our outpatient clinic with a 1-month history of an abnormal throat sensation, and denied any history of bleeding, pharyngeal pain, eating obstruction, and dyspnea. He had no medical, family, psychosocial history (including co-morbidities), or relevant genetic information, and had not been treated previously for this condition. Laryngoscopy revealed a smooth, pale oval mass whose pedicle adhered to the lateral wall of the right pharynx at the level of the epiglottis. Magnetic resonance imaging (MRI) showed that the lesion was in the right lateral pharyngeal wall. The lesion was hypointense on T1-weighted images (T1WI) (Fig. 1) and hyperintense on T2-weighted images (Fig. 2). It then underwent enhancement with contrast administration using T1WI (Fig. 3).

Under general anesthesia, the lesion was found at the right lateral pharyngeal wall via the supporting laryngoscope. The pedicle was small and adhered to the lateral pharyngeal wall. Then, the tumor was completely removed by coblation, and a histological examination of the resected mass was performed. The white tumor which was surrounded by a pale pink fibrous capsule had a smooth surface, mobility and a size of about 2.2 × 3.3 cm (Fig. 4). The consistency of the tumor was firm to rubbery.
The specimen showed which spindle cells were diffusely distributed and differentiated with loose extracellular interstitium and scattered fibrovascular tissue (Fig. 5). Immunohistochemical staining revealed a strong positivity for CD34 and estrogen receptors (ER), and negative staining for S-100 protein and smooth muscle actin. The final pathology report showed that the specimen was consistent with a cellular angiofibroma of the hypopharynx. The patient was asymptomatic and did not show recurrence at the 3-month follow-up. Thus far, no recurrence has been observed 6 months after excision.

3. Discussion

Cellular angiofibroma is a benign mesenchymal tumor that is usually found in the vulvovaginal and inguinoscrotal regions,[5] and is characterized by 2 main features:

(1) small to medium-sized vessels with mural hyalinization, and
(2) bland spindle cells.[1,6,7]
Genetically, cellular angiofibroma is similar to spindle cell lipomas, as both have abnormal karyotypes that entail the heterozygous loss of material from the long arm of chromosome 13[8,9]. Cases outside the vulvovaginal and inguinoscrotal regions have also been reported, including the oral mucosa, male pelvis, subcutaneous tissue of the chest wall, nasopharynx, and retroperitoneum.[2,3,10,11] Cellular angiofibromas are well-circumscribed benign mesenchymal tumors, and most are located in the superficial soft tissue of the trunk. Most cellular angiofibromas present as an asymptomatic subcutaneous mass, except for nasopharyngeal and retroperitoneal tumors.[2,3,10,11] In the nasopharynx, the tumor presents with frequent epistaxis and persisting nasal obstruction.[2] whereas retroperitoneal tumors present with pain in the right iliac fossa.[3] This paper is the first report of cellular angiofibroma arising in the hypopharynx, which shows proves that the tumor can occur not only in the urinary and respiratory tract, but also in the digestive tract.

**Figure 4.** The operative region after excision, which had a smooth surface, good activity, and a size of about 2.2 x 3.3 cm.

**Figure 5.** (A) Higher magnification showing spindle cells with admixed adipose tissue and blood vessels (hematoxylin-eosin staining, 200×). (B) CD34 immunoreactivity in both endothelial cells and fibroblasts (immunohistochemistry, 200×).
The major histopathologic differential diagnoses of cellular angiofibroma include vessel-rich lesions with myofibroblastic or fibroblastic elements (eg, such as solitary fibrous tumor), spindle cell lipoma, angiomyxofibrolastoma, and superficial angiomyxoma.\(^{12,13}\) Solitary fibrous tumors are either found incidentally via imaging or produce local compression on nearby structures. Both computed tomography and MRI scans reveal homogeneously enhanced masses with distinct borders.\(^{14}\) Hematoxylin-eosin staining examination of the lesions reveal in patternless, haphazardly arranged spindle cells and amorphous areas of collagen, a lack of prominent endothelium,\(^{15}\) a significant presence of CD34, CD99, and bcl-2.\(^{14}\) Spindle cell lipomas consist of small, cylindric spindle cells and collagen fibers, as well as mixed mature adipocytes within a matrix containing mucinous material, which shows immunoreactivity for CD34, vimentin, and bcl-2, but not S-100.\(^{16}\) The angiomyxofibrolastoma is a subcutaneous, well-circumscribed, vulvovaginal tumor composed of epithelioid and spindle mesenchymal cells arranged in cords and nests preferentially around numerous small to medium-sized vessels.\(^{17}\) Superficial angiomyxoma presents as individual nodules which are sparsely to moderately cellular, with copious basophilic interstitial material. The tumor contains scattered spindle and stellate-shaped cells and a scarce vascular network.\(^{18}\)

Preoperative diagnosis of cellular angiofibroma is challenging based on clinical symptoms and imaging findings alone; the exact diagnosis depends on pathological and immunohistochemical results. Microscopically, cellular angiofibroma is a cellular neoplasm, composed of bland spindle-shaped cells, numerous small- to medium-sized thick-walled and often hyalinized vessels, and a minor component of adipose tissue.\(^{19}\) Studies have found that cellular angiofibroma is consistently positive for CD34, and consistently negative for S-100 protein, h-Caldesmon, and Keratin. Further, some studies have reported that cellular angiofibroma shows immunoreactivity for Vimentin and ERs and/or progesterone receptors.\(^{20,21}\)

In conclusion cellular angiofibroma appears to behave in a benign fashion, and treatment without atypia is easy to excise and requires close follow-up. To date, follow-up studies have shown that it has a low recurrence rate with no chance of metastasization. The same phenomenon was also found in cases of atypia or sarcomatous transformations.\(^{22}\) The limitations in this case is the short follow-up period.

**Author contributions**

Conceptualization: Ya Liu, Qinying Wang, Qiongqiong Chen.

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Investigation: Ya Liu.

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