Clinical Case Report

Medicine®

OPEN

Cellular angiofibroma in the hypopharynx
A case report

Ya Liu, Bachelor's Degree, Yaping Xu, PhD*, Qinying Wang, MD, Qiongqiong Chen, MD

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.[2] 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.

Editor: N/A.

The authors have no conflicts of interest to disclose.

Department of Otolaryngology, The First Affiliated Hospital, College of Medicine, Zhejiang University, Hangzhou, Zhejiang, China.

* Correspondence: Yaping Xu, Department of Otolaryngology, The First Affiliated Hospital, College of Medicine, Zhejiang University, 310003 Hangzhou, Zhejiang, PR China (e-mail: 2185033@zju.edu.cn).

How to cite this article: Liu Y, Xu Y, Wang Q, Chen Q. Cellular angiofibroma in the hypopharynx: a case report. Medicine 2019;98:50(e18385).

Received: 9 July 2019 / Received in final form: 31 October 2019 / Accepted: 14 November 2019

http://dx.doi.org/10.1097/MD.0000000000018385

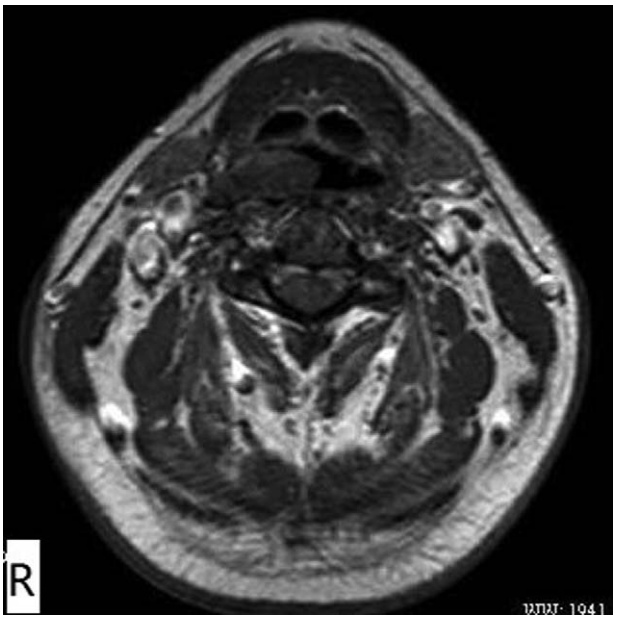

Figure 1. The lesion shows a hypointense, well-defined, oval mass in which the pedicle adhered to the lateral wall of the right pharynx on T1-weighted images.

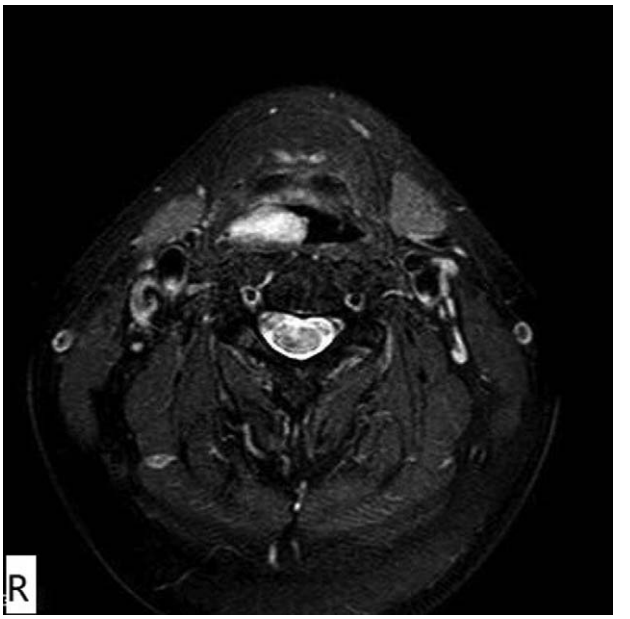

Figure 2. The lesion presented as hyperintense on T2-weighted images.

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]

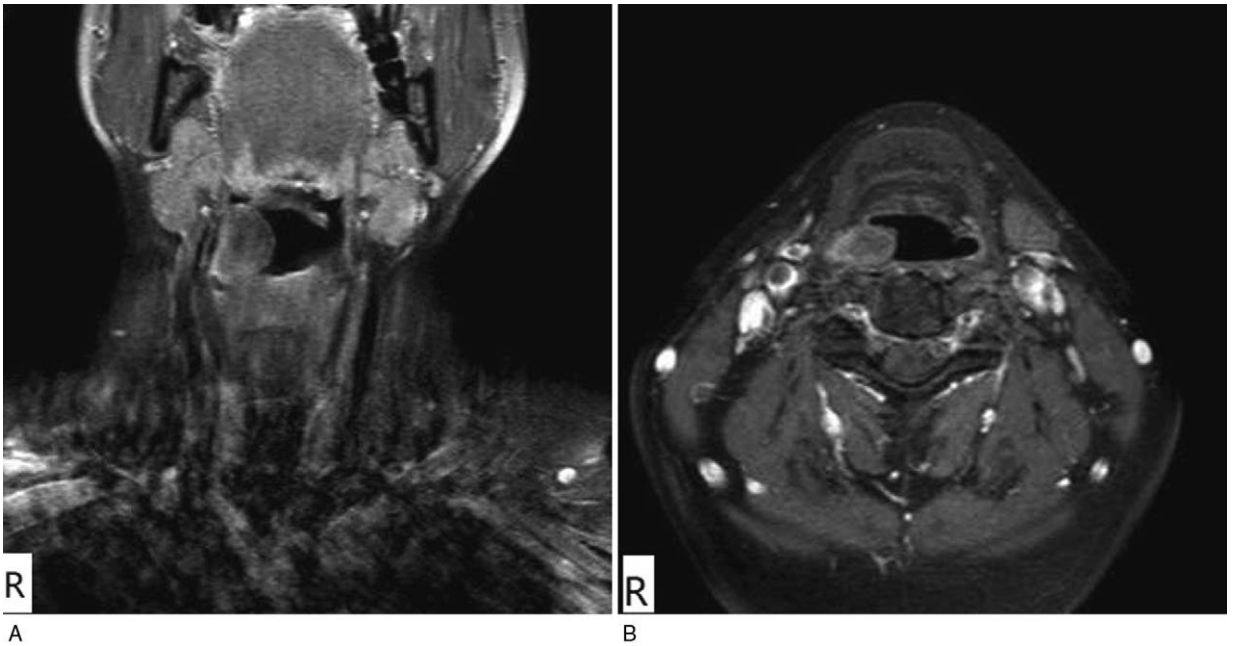

Figure 3. The lesion with enhancement via contrast administration on T1-weighted images.

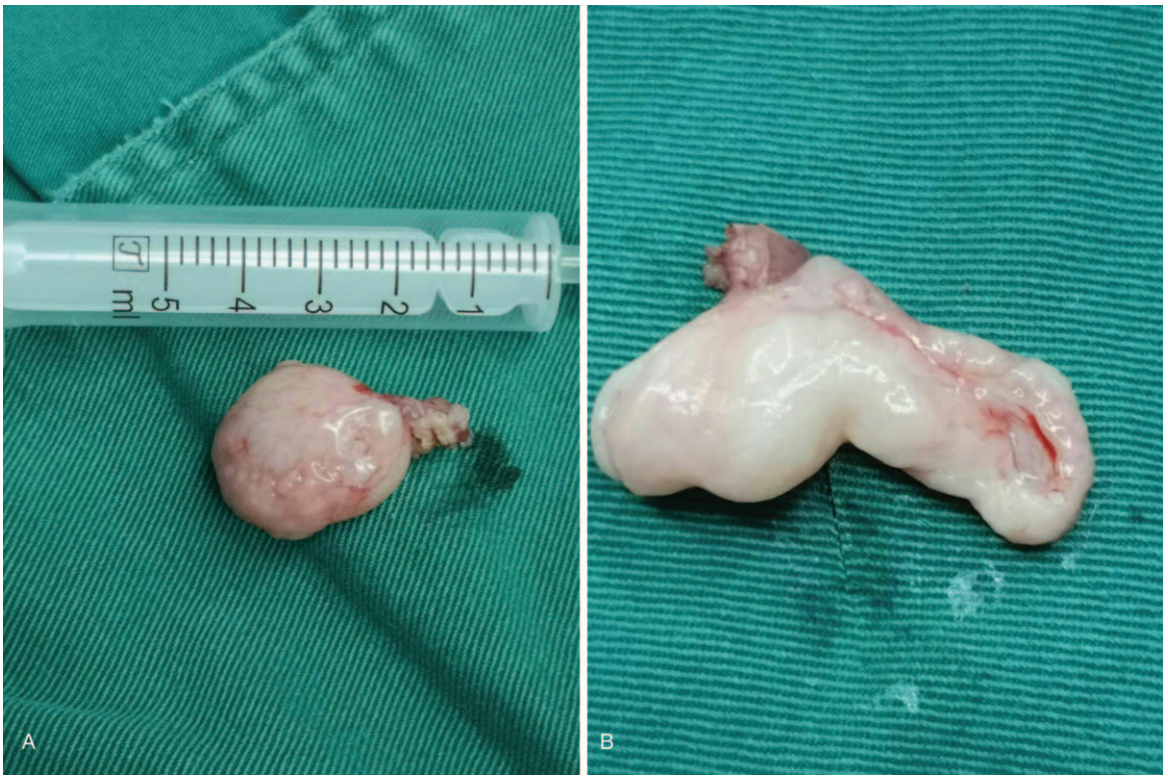

Figure 4. The operative region after excision, which had a smooth surface, good activity, and a size of about 2.2 × 3.3 cm.

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.

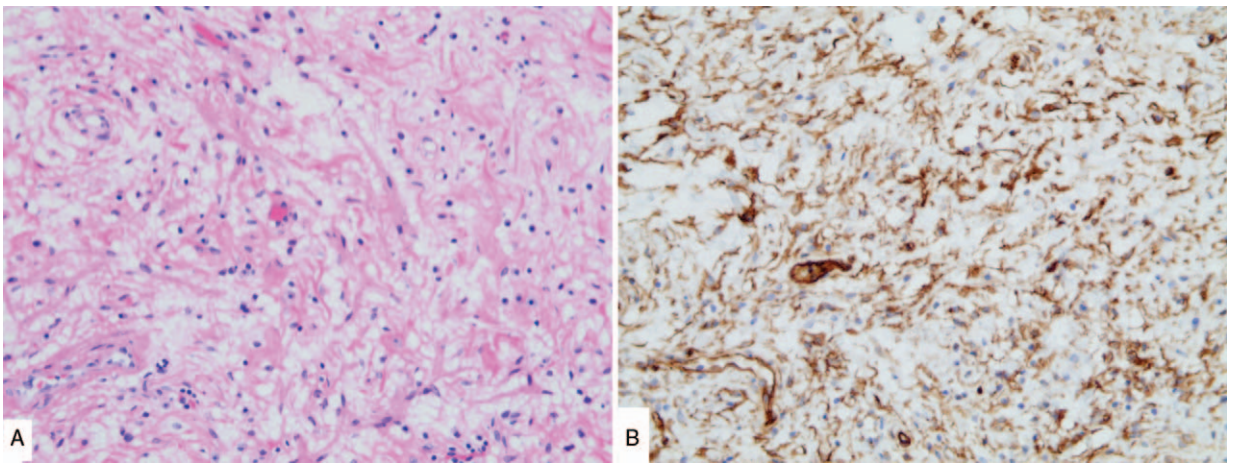

Figure 5. (A) Higher magnification showing spindle cells with admixed adipose tissue and blood vessels (hematoxylin-eosin staining, 200×). (B) CD34 immunoreactivity in both endothelial cells and fibroblasts (immunohistochemistry, 200×).

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, angiomyofibroblastoma, 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 homogenously enhanced masses with distinct borders.[14] Hematoxylin-eosin staining examination of the lesions reveal in pattern-less, 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 immunoreactiveity for CD34, vimentin, and bcl-2, but not S-100.[16] The angiomyofibroblastoma is a subcutaneous, well-circumscribed, vulvovaginal tumor composed of epithelioid and spindled mesenchymal cells arranged in cords and nesting 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 spindled 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.
Data curation: Ya Liu, Qinying Wang.
Investigation: Ya Liu.
Methodology: Ya Liu.
Project administration: Yaping Xu, Qinying Wang.
Supervision: Yaping Xu, Qiongqiong Chen.
Visualization: Qiongqiong Chen.
Writing – original draft: Ya Liu.
Writing – review and editing: Ya Liu, Yaping Xu, Qinying Wang.

References

[1] Nucci Mr GS, Fletcher C. Cellular angiofibroma: a benign neoplasm distinct from angiomyofibroblastoma and spindle cell lipoma. Am J Surg Pathol 1997;21:636–44.
[2] Erdur ZB, Yener HM, Yilmaz M, et al. Cellular angiofibroma of the nasopharynx. J Craniofac Surg 2017;28:720–2.
[3] Brandao A, Campos S, Fraga J, et al. Retroperitoneal cellular angiofibroma: a rare gynecological entity. Acta Med Port 2017; 30:882–6.
[4] Chen E, Fletcher CD. Cellular angiofibroma with atypia or sarcomatous transformation: clinicopathologic analysis of 13 cases. Am J Surg Pathol 2010;34:707–14.
[5] Iwasa Y. Cellular angiofibromaclinicopathologic and immunohistochemical analysis of 51 cases. Am J Surg Pathol 2004;28:1426–35.
[6] Flucke U, Van Krieken JH, Mentzel T. Cellular angiofibroma: analysis of 25 cases emphasizing its relationship to spindle cell lipoma and mammary-type myofibroblastoma. Mod Pathol 2011;24:82–9.
[7] Iwasa Y, Fletcher CD. Cellular angiofibroma: clinicopathologic and immunohistochemical analysis of 51 cases. Am J Surg Pathol 2004; 28:1426–35.
[8] Panagopoulos I, Gorunova L, Bjerkehagen B, et al. Loss of chromosome 13 material in cellular angiofibromas indicates pathogenetic similarity with spindle cell lipomas. Diagn Pathol 2017;12:17.
[9] Hameed M, Clarke K, Amer HZ, et al. Cellular angiofibroma is genetically similar to spindle cell lipoma: a case report. Cancer Genet Cytogenet 2007;177:131–4.
[10] Garijo MF, Val-Bernal JF. Extravulvar subcutaneous cellular angiofibroma. J Cutan Pathol 1998;25:327–32.
[11] Eversole LR. Cellular angiofibroma of oral mucosa: report of two cases. Head Neck Pathol 2009;3:136–9.
[12] Canales Bk WD, Hoffman N, Slaton J, et al. Angiomyofibroblastoma-like tumors (cellular angiofibroma). Int J Urol 2006;13:177–9.
[13] Fetsch JF, Tavassoli FA. Superficial angiomyxoma (cutaneous myxoma) a clinicopathologic study of 17 cases arising in the genital region. Int J Gynecol Pathol 1997;16:325–34.
[14] Wignall OJ, Moskovic EC, Thway K, et al. Solitary fibrous tumors of the soft tissues: review of the imaging and clinical features with histopathologic correlation. AJR Am J Roentgenol 2010;195:W55–62.
[15] Gengler C, Guillou L. Solitary fibrous tumour and haemangiopericytoma: evolution of a concept. Histopathology 2006;48:63–74.
[16] Evcimik MF, Ozkurt FE, Sapci T, et al. Spindle Cell Lipoma of the Hypopharynx. Int J Med Sci 2011;8:479–81.
[17] Laskin WB, Fetsch JF, Tavassoli FA. Angiomyofibroblastoma of the female genital tract: analysis of 17 cases including a lipomatous variant. Hum Pathol 1997;28:1046–55.
[18] Allen PW, Dymock RB, Maccormac LB. Superficial angiomyxomas with and without epithelial components. Report of 30 tumors in 28 patients. Am J Surg Pathol 1988;12:519–30.
[19] Nucci F. Vulvovaginal soft tissue tumours: update and review. Histopatholog 2010;36:97–108.
[20] Val-Bernal JF, Garijo MF, et al. Extragenital subcutaneous cellular angiofibroma: case report. APMIS 2007;115:254–8.
[21] Mccluggage WG, Ganesan R, Hirschowitz L, et al. Cellular angiofibroma and related fibromatous lesions of the vulva: report of a series of cases with a morphological spectrum wider than previously described. Histopathology 2010;45:360–8.
[22] Mandato VD, Santagni S, Cavazza A, et al. Cellular angiofibroma in women: a review of the literature. Diagn Pathol 2015;10: 1746-1596.