

## Case Report

# A case of laryngeal angioleiomyoma and review of literature

Xue Zhao, Dan Yu, Yin Zhao, Yan Liu, Xinmeng Qi, Chunshun Jin

*Department of Otolaryngology Head and Neck Surgery, The Second Hospital, Jilin University, 218 Ziqiang Street, Changchun 130041, Jilin Province, China*

Received November 3, 2014; Accepted November 23, 2014; Epub January 15, 2015; Published January 30, 2015

**Abstract:** Angioleiomyoma is a rare benign vascular smooth muscle tumor that arise from the tunica media of veins and arteries. Here a case of laryngeal angioleiomyoma in a 57-year-old Chinese man is reported. The patient presented with dysphagia for one and half-month and dyspnea during the previous one week, was hospitalized for treatment with a tracheotomy and laryngofissure with the unblock mass excision. Final pathological evaluation of the neoplasm confirmed a diagnosis of laryngeal angioleiomyoma. The patient had been followed up 4 years with no recurrence of disease. This study demonstrated the clinical feature, pathology, treatment and outcome of the rare disease of laryngeal angioleiomyoma.

**Keywords:** Angioleiomyoma, laryngeal tumor, treatment

### Introduction

Angioleiomyomas (also named vascular leiomyoma or angioleiomoma) are benign vascular smooth muscle tumors that arise from the tunica media of veins and arteries [1, 2]. It's commonly seen in uterus, gastrointestinal system, extremities and skin, but rarely in the head and neck region, especially in larynx. Laryngeal angioleiomyoma mainly presents between 40-60 year olds and frequently in male [3]. We present a case of laryngeal angioleiomyoma with an emphasis on clinical data, pathologic findings, and surgical approach.

### Case report

A 57-year-old Chinese man presented to the Department of Otolaryngology/Head and Neck Surgery, Second Hospital, Jilin University, Changchun, Jilin Province, China, in December 2010, complained about a one and half-month history of dysphagia and dyspnea during the previous one week, with breathing difficulties that more severe after exercise. However, there was no fever or other systemic symptoms and the patient didn't report any hoarseness, laryngeal pain, odynophagia, cough or hemoptysis.

Electronic laryngoscopic (**Figure 1**) and esophagoscopy examination revealed no abnormali-

ties within the nasopharynx, oropharynx or hypopharynx. Laryngeal evaluation revealed a smooth submucosal eminentia neoplasm covered with normal mucosa in the left aryepiglottic fold and extending over arytenoid cartilage. The mass was tenacious. Both true vocal cords were mobile. No other abnormalities were found in the remainder of the head and neck examination.

The patient was hospitalized. Computed tomography (CT) (**Figure 4**) showed a solid lesion that was about 2.0 cm in diameter with a well-defined margin in the left aryepiglottic fold. There was no evidence of adjacent thyroid cartilage destruction, extra-laryngeal extension, or cervical lymph node enlargement.

Magnetic resonance imaging (MRI) (**Figure 5**) presented a well-defined oval mass that was slightly hypointense to skeletal muscle on T1-weighted and hyperintense to skeletal muscle and heterogeneous on T2-weighted, with strong enhancement after IV injection of contrast material.

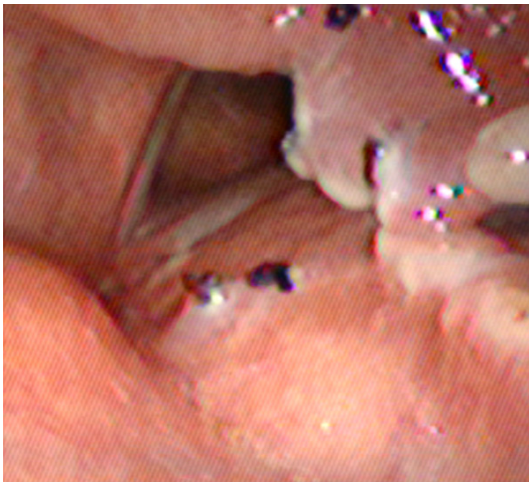
As the tumor was covered with normal mucosa in the left aryepiglottic fold, the apparent tissue would be reported to be non-diagnosable, so a laryngoscopy-guided biopsy was not performed. Biopsy was performed through direct laryngos-



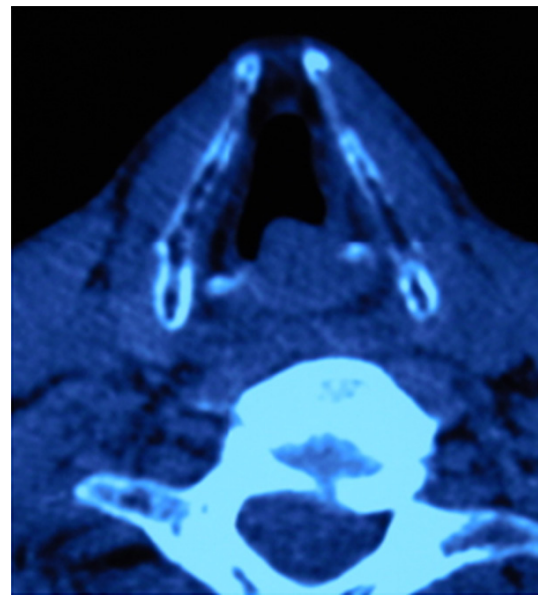
**Figure 1.** The electronic laryngoscopy picture before operation; Laryngeal evaluation revealed a smooth submucosal eminentia neoplasm covered with normal mucosa in the left aryepiglottic fold and extending over arytenoid cartilage.



**Figure 3.** The electronic laryngoscopy picture of excision site of the larynx, with no recurrence one year later after operation.



**Figure 2.** The electronic laryngoscopy picture of excision site of the larynx, with visual black knots after operation.

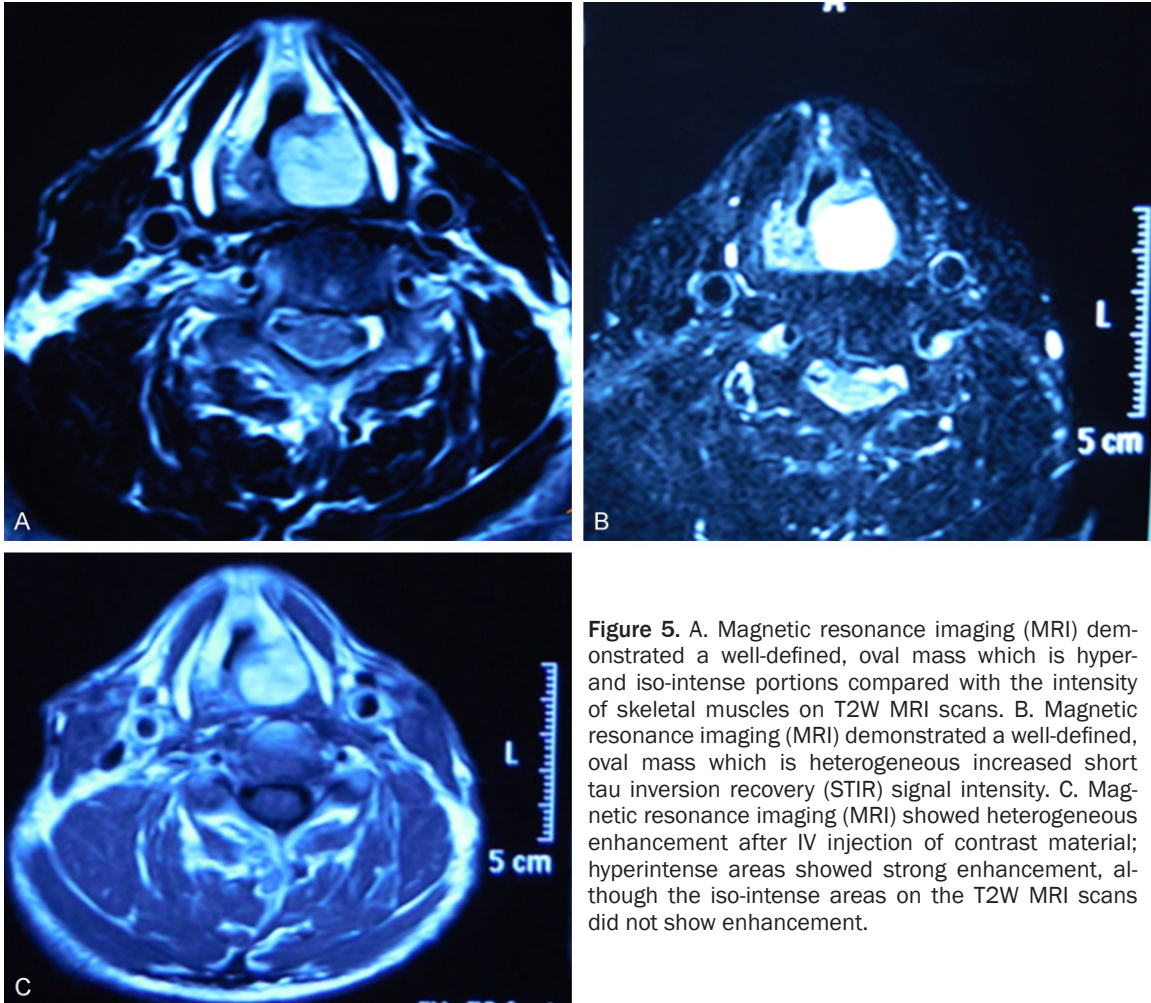


**Figure 4.** Computed tomography (CT) showed a solid lesion that was about 2.0 cm in diameter with a well-defined margin in the left aryepiglottic fold.

copy under general anesthesia. Intraoperative frozen section examination revealed the lesion comprised of rich vascular channels with thick vessel walls and smooth muscle bundles with elongated nuclei. As bleeding was encountered by the transoral approach, the pre-planned surgical procedure was, therefore, modified to include a tracheotomy and laryngofissure with unblock mass excision. During the operation, the fascia, perichondrium and the thyroid cartilage were excised in the middle. There were no abnormalities within the false vocal cords, laryngeal ventricles and the true vocal cords

under direct vision. The mass covered with normal mucosa was in the left aryepiglottic fold and extended over arytenoid cartilage. After the division of the aryepiglottic fold mucosa, the encapsulated tumor (**Figure 6**) was exposed and removed completely. All the laryngeal functions were preserved.

On gross examination, the tumor was a solid, dense grayish, well-defined, encapsulated mass measuring approximately 2.0 cm × 1.5



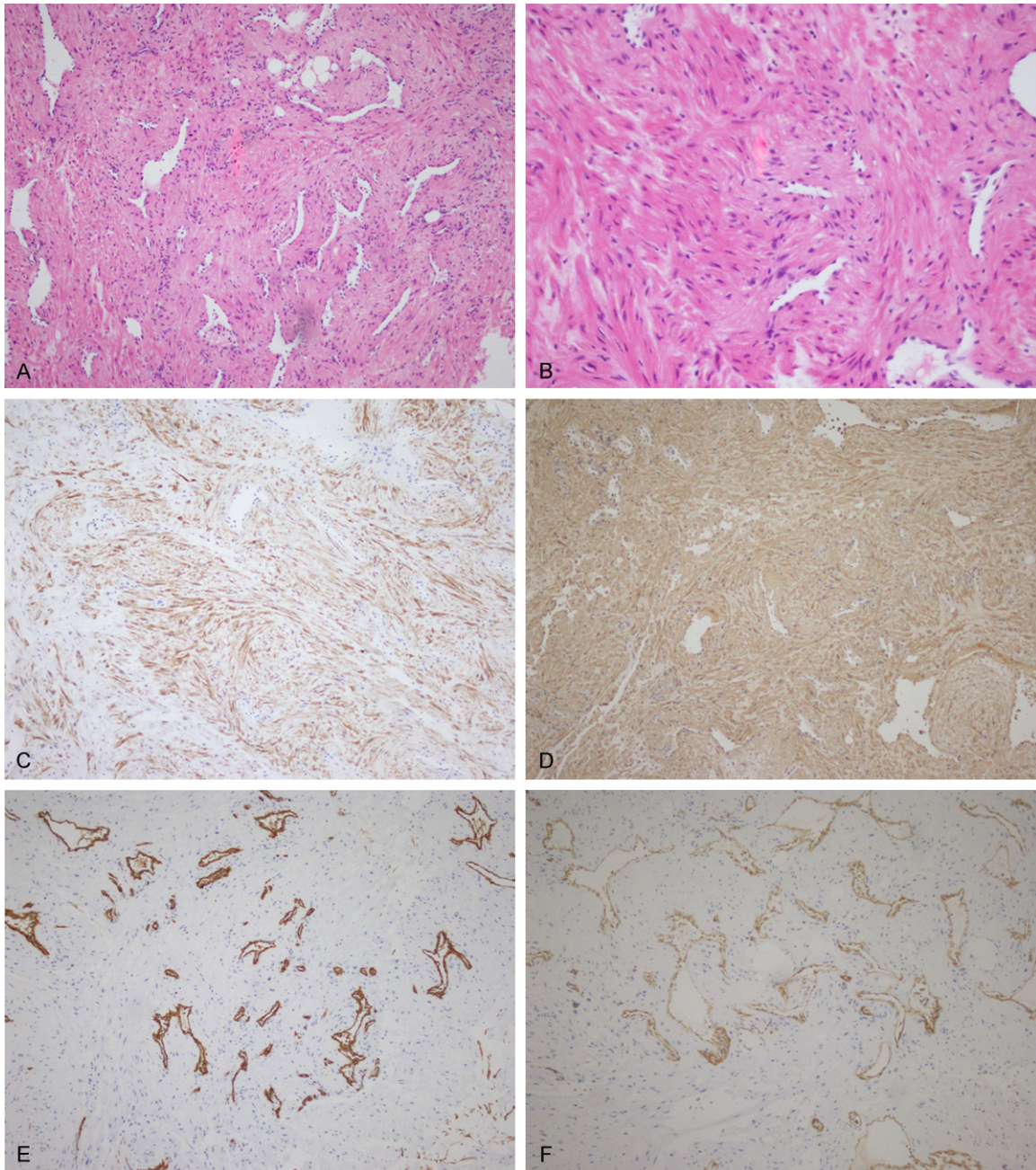
**Figure 5.** A. Magnetic resonance imaging (MRI) demonstrated a well-defined, oval mass which is hyper- and iso-intense portions compared with the intensity of skeletal muscles on T2W MRI scans. B. Magnetic resonance imaging (MRI) demonstrated a well-defined, oval mass which is heterogeneous increased short tau inversion recovery (STIR) signal intensity. C. Magnetic resonance imaging (MRI) showed heterogeneous enhancement after IV injection of contrast material; hyperintense areas showed strong enhancement, although the iso-intense areas on the T2W MRI scans did not show enhancement.



**Figure 6.** The encapsulated tumor was removed completely; It was a solid, dense grayish, well-defined, encapsulated mass measuring approximately 2.0 cm × 1.5 cm × 1.0 cm.

cm × 1.0 cm. Histologically, the lesion consisted of rich vascular channels with thick vessel walls and smooth muscle bundles with elongated nuclei. Immunohistochemistry was used to evaluate further the cellular components of the mass. Some cells labelled for smooth muscle actin and desmin. Endothelial cells in tumor vessels stained positively for CD34 and CD31 (**Figure 7**). Based on these histological data, the tumor was pathologically diagnosed as angioleiomyoma.

There were no complications during or after operation. Nutrition was provided for 7 days post-surgery by means of a nasogastric feeding tube positioned during the surgical procedure. The nasogastric feeding tube and the tracheotomy tube were removed separately on day 7 and day 10 post operatively. The speech and swallowing functions were completely restored. The patient was followed up for 4 years with no recurrence.



**Figure 7.** Histopathological findings as following: (A and B) Laryngeal Angioleiomyoma: composed of blood vessels and smooth muscle fibers, which are seen surrounding the vessels (HE, original magnification  $\times 100$  (A) and  $\times 200$  (B)); (C) Laryngeal Angioleiomyoma: Desmin (+); (D) Laryngeal Angioleiomyoma: SMA (+); (E) Laryngeal Angioleiomyoma: CD34 (+); (F) Laryngeal Angioleiomyoma: CD31 (+).

### Discussion

Angioleiomyoma (also named vascular leiomyoma or angiomyoma) is a benign vascular smooth muscle tumor arising from the tunica media of veins and arteries [1, 2]. It's infrequent in the head and neck region, accounting for 10% of cases in the published literature. Some studies have reported the buccal muco-

sa, the parotid gland and palate are the common sites for angioleiomyomas [4]. However, it is especially rare in larynx. The majority of angioleiomyomas are  $< 2$  cm in diameter on presentation and rarely exceed 4-5 cm [4].

Some karyotypic abnormalities of angioleiomyoma from different anatomic sites have been reported in the literatures [5], such as the

## A case of laryngeal angioleiomyoma and review of literature

translocation 45,XX,t(4;5)(p12;q33), der(13;15)(q10;q10)/46,XX in the extremity angioleiomyoma. However, there were none descriptions about Laryngeal angioleiomyoma.

The etiology of angioleiomyoma is unknown. In addition to minor trauma and venous stasis, the effects of hormone have been proposed for angioleiomyoma. Masaru Hayashi [6] inferred that sex hormones possibly have influence on uterine angioleiomyoma. However, Hirakwa [7] reported estrogens appeared to be unrelated to angioleiomyoma outside uterus. While Naito et al [8] believes that parathyroid-hormone-related peptide contributes to the formation of characteristic microenvironment of angioleiomyoma cell composition by an autocrine and/or paracrine mechanism.

Histologically, Angioleiomyoma is composed of smooth muscle bundles and vascular channels as like hematoma, encapsulated roundly. Morimoro classified the 241 cases of studied angioleiomyoma into three histological types: one was the solid type, closely composed of smooth muscle and many small, slit-like vascular channel. Second was the venous type with thick, easily identifiable muscular walls characteristically. The last was the cavernous type with dilated vascular channels and less smooth muscle. Also, Morimoro grouped them into 2 groups. One was the larger group of extremity tumors, where the tumors are mainly of the solid type and painful, with high prevalence among women. The other was the smaller group of head tumors, where the tumors are usually of the venous type. On the contrary, they are often painless and more common in males. Recently, Cheng-ping Wang et al [9] reviewed the clinical records of 21 patients with angioleiomyoma of the head and neck and showed the solid type was the most common, whereas the venous type was the least common. There was another opinion that the histological classification was not essential in angioleiomyoma of head and neck, for it had no effect on the treatment of choice. However, Histological and Immunohistochemistry examination following resection remains the most reliable method for diagnosis. The positive expression of desmin and  $\alpha$ -actin smooth muscle antibodies demonstrated the presence of smooth muscle cells, and the positive expression of CD34 and CD31 demonstrated the

presence of vascular endothelium. Based on these histological data, a diagnosis of laryngeal angioleiomyoma should be confirmed by the exclusion of vascular tumors (such as hemangiomas and lymphangiomas), benign mesenchymal tumors (including lipomas, schwannomas and neurofibromas), spindle cell tumor (such as leiomyoma, myofibroma), malignant mesenchymal tumors (including leiomyosarcoma) et al. [4].

Although MRI has no characteristic findings for preoperative diagnosis, it is helpful in the differential diagnosis of angioleiomyoma, such as fibrous nodules (low signal intensity on T1-and T2-weighted images) and lipomas (high signal intensity on T1-weighted images). It is also effective to prevent the likelihood of profuse bleeding after blind biopsy or operation, especially for the laryngeal angioleiomyoma. MRI of angioleiomyoma typically demonstrates a well-defined, oval mass which commonly is isotense to muscle on T1-weighted images with heterogeneous increased internal T2W/short tau inversion recovery (STIR) signal intensity, resulted from the varied proportions of the different components. The well-defined peripheral hypointense area on the T2-weighted images is corresponded to the fibrous capsule. And enhancement after IV gadolinium ranged from diffuse to heterogeneous [10]. After the intravenous administration of a contrast material, hyperintense areas showed strong enhancement, although the iso-intense areas on the T2W MRI scans did not show enhancement. The hyperintense areas on the T2W MRI scans correspond to the smooth muscle bundle cells, and the iso-intense areas correlate with the fibrous tissue or intravascular thrombi [11]. Because enhancement of a varying intensity is consistent with the vascular nature of the lesion, MRI should be performed rebiopsied or preoperatively, due to the considerable hemorrhage.

The best treatment is to surgical excision completely via laryngomicrosurgery or external approach. The surgical modality option depends on the site of tumor occurrence, morphology, tumor size, the expected amount of blood loss and comorbidities the patient may have. Shibata et al [12] reported two cases of laryngeal angioleiomyoma found separately in the supraglottic area of the larynx and the subglot-

tic space were removed through direct laryngoscopy. Fuse et al [13] reported resection of angioleiomyoma of the retropharyngeal space via a lateral pharyngotomy approach under general anesthesia. Laryngomicrosurgery takes the advantage of less tissue damage and rapid recovery, while with the high risk of hemorrhage during and after surgery. As the CO<sub>2</sub> laser technology and COBLATION technology have been developed enormously, the surgery with them has proven to reduce effectively profuse bleeding and other complications. Anderson et al [14] reported a CO<sub>2</sub> laser was used successfully to completely excise the laryngeal angioleiomyoma and to arrest blood. Although preoperative embolization [15] of the tumor to minimize perioperative hemorrhage is suggested, no cases have been applied until now. Larger angioleiomyoma may require external approach, so as to the tumor that is not clearly visualized. Coagulating wounded surface by the electrosurgical unit protects from intraoperative and postoperative bleeding. Owing to a fibrous capsule, the tumor could be resected completely and the normal mucosa covered is proposed to be reserved as more as possible with the benefit of repairment of the wound and laryngeal function reconstruction.

The recurrence is rare. At the time of writing, the patient had been followed up 4 years with no recurrence of disease. However two case has been reported in the literature [14, 16], suggesting that it is significant for the complete excision of the tumor along the capsule. Malignant transformation of laryngeal angioleiomyoma is highly unlikely [9]. However, there are only two reports about the malignant transformation of the benign angioleiomyoma in the reviewed literature, separately in the index finger of a 17-year-old case and in the forearm of a 67-year-old woman [17]. So patients should be monitored after surgery.

In summary, angioleiomyoma is a rare, benign, local disease. In spite of the rarity of this disease in the larynx, it should be included in the differential diagnosis for laryngeal swellings. The diagnosis of angioleiomyoma is confirmed by histological and immunohistochemistry examination. Complete resection via laryngomicrosurgery or external approach is the best choice of treatment. The long term follow-up is advised despite of the rare recurrence and malignant transformation of laryngeal angioleiomyoma.

### Disclosure of conflict of interest

None.

**Address correspondence to:** Dr. Chunshun Jin or Dr. Dan Yu, Department of Otolaryngology Head and Neck Surgery, The Second Hospital, Jilin University, 218 Ziqiang Street, Changchun 130041, Jilin Province, China. E-mail: chunshunjindan@163.com (CSJ); yudan19792003@163.com (DY)

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## A case of laryngeal angioleiomyoma and review of literature

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