ANGIOMYXOMA OF THE EPIGLOTTIS WITH A SINGLE INSTITUTION CASE SERIES

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Introduction

Objectives: We describe the first case, to our knowledge, of an angiomyxoma occuring on the epiglottis. We also present a case series of head and neck angiomyxomas at Mayo Clinic. Lastly, we provide an updated review of the literature regarding these rare tumors.

Study Design: Case report; case series; literature review.

Methods: We report the case of a patient presenting with globus sensation caused by an angiomyxoma of the epiglottis. Included is a case series at our institution, established by examining the pathology database, as well as the BSI systems database for patients with a diagnosed angiomyxoma of the head or neck. We also performed a review of the available biomedical literature and summarized by site and frequency those tumors previously documented.

Results: We discovered 6 patients in our database that had been histologically diagnosed with angiomyxomas of the head and neck, the largest single institution series to our knowledge. One tumor was located on the posterior neck, two on the lower lip, one on the upper eyelid, one on the nasal tip and the most recent located on the epiglottis. A review of the current literature regarding these tumors was also completed and, aside from those reported in our case series, at least 44 have been reported thus far in the head and neck.

Case Report

A 60 year old female with an otherwise benign past medical history presented to our Laryngology clinic with a persistent globus sensation present for about one year. She had an upper respiratory tract infection just prior to becoming aware of this sensation. She noticed progressive dysphagia, feeling as if food was “stuck” in her throat, as well as a change in the quality of her voice. A flexible fiberoptic exam demonstrated a submucosal mass on the epiglottis and a CT scan was obtained and can be viewed in Figure 1.

She was taken to the operating room and underwent microdirect laryngoscopy and C02 LASER excision of the mass pictured in Figure 2, histopathologic diagnosis of which revealed angiomyxoma.

Figure 1: Contrasted CT scan

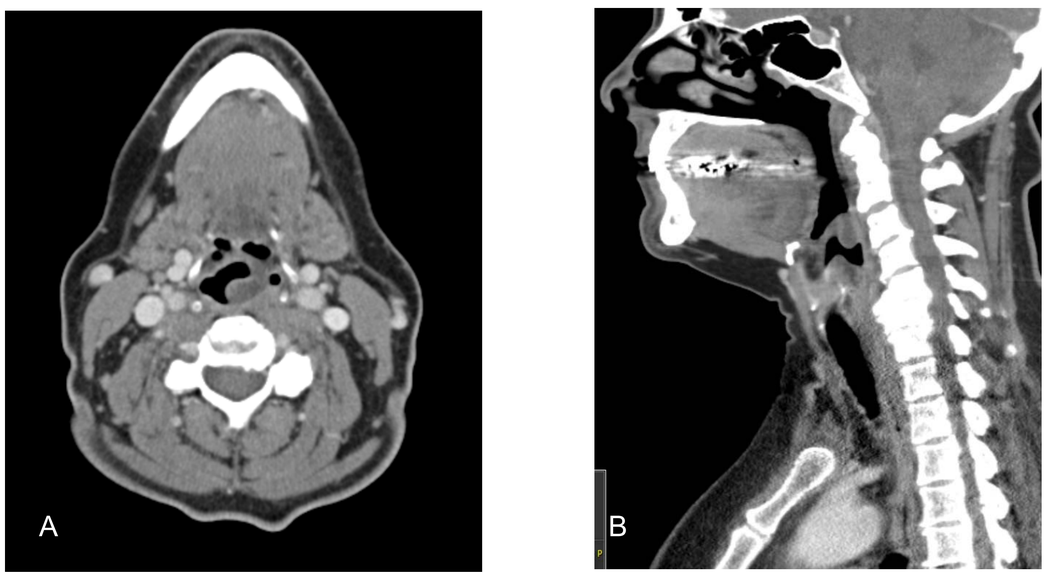

Figure 1: Contrast- enhanced axial (A) and sagittal (B) CT images demonstrating an approximately 1.8 x 1.6 x 1.3 cm cystic mass pedicled on the epiglottis and left aryepiglottic fold

Figure 2: Intraoperative photographs

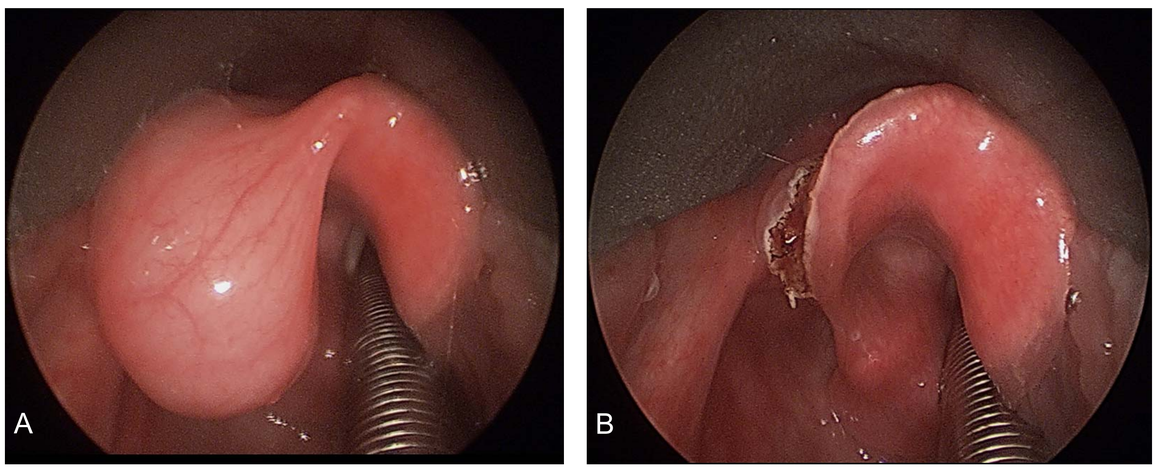

Figure 2: Intraoperative, endoscopic photographs of the epiglottic lesion (A) and the larynx after surgical excision (B). The Lindholm laryngoscope can be seen at the upper aspect of both images with a LASER-safe endotracheal tube passing through the larynx.

Table 1: Case Series at Mayo Clinic

Location	Size (maximal diameter)	Age/ Sex	Presentation	Treatment	Outcome	Follow up (months)
Skin, Posterior neck	2.3 cm	27 M	Painless neck mas	Excision	Local recurence, re-excised	72
Nasal tip	3 mm	10 F	Painless nasal papule	Excision	NED	120
Epiglottis	2.8 cm	60 F	Globus	C02 LASER excision	NED	21
Lower lip	1.3 cm	20 M	Submucosal lesion	Excision	NED	60
Lower lip	0.5cm	30 F	Lower lip lesion after trauma	Excision	Lost to follow up	NA
Upper eyelid	0.3 cm	83 M	Rapidly growing, tender nodule	Excision	NED	46

Table 1. Cases of angiomyxoma of the head and neck at Mayo Clinic from 2000-2016. Age ranged from 10- 83 years (mean=38). Length of follow up was 21-120 months (mean=63) with one patient last to follow up. All were surgically excised with negative margins. There was one episode of local recurrence. NED= no evidence of disease.

Table 2: Literature Review

Location	Incidence
Eye	**9**
Retropharynx	**2**
Skin	**22**
- **Eyelid**	2
- **Neck**	5
- **Nose**	2
- **Face**	11
- **Ear**	2
Oral cavity	**6**
Parotid	**1**
Larynx	**2**
Sinuses	**1**
Deep neck	**1**
Total	**44**

Table 2. Literature review designating incidence of angiomyxoma occuring in the head and neck by subsite. A total of 44 tumors were encountered in the literature.

Conclusion

- Angiomyxomas are a rare subset of tumors of the myxoid, or connective tissue, family. They are infrequently found in the head and neck and encountered by the otorhinolaryngologist.

- Most lesions in the head and neck present as painless cutaneous or subcutaneous nodules or papules with varying gross appearance

- Histologically, they demonstrate numerous small blood vessls of varying size, mucin, spindle cells and stellate cells among a myxoid stroma

- Can be isolated or in the setting of Carney’s complex

- No evidence of malignant potential but have been shown to invade and recur locally

- Treatment of choice is excision with clear histopathologic margins and long term follow up

References

- Attaran S, Ainsborough N, Helliwell T, Shackcloth M. Retropharyngeal angiomyxoma presenting with transoral intussusception. Interact Cardiovasc Thorac Surg. 2011;13(1):96-7.
- Liu W, Lin J, Yu X. Clinical and histopathological features of conjunctival myxoma and angiomyxoma. Chinese Journal of Ophthalmology. 2012 Nov; 48)11): 981-4.
- Toth A, Nemeth T, Szucs A, Szollosi Z, Sziklai I. Retropharyngeal superficial angiomyxoma. J Laryngol Otol. 2010;124(9):1017-20.
- Ali N, Child CS, Michaelides M, Olver JM. Recurrence of a rare skin tumour: superficial angiomyxoma in the eyelid. Can J Ophthalmol. 2011;46(2):205-6.
- Yuen HK, Cheuk W, Luk FO, Wat CS, Auyeung KC, Lam DS. Solitary superficial angiomyxoma in the eyelid. Am J Ophthalmol. 2005;139(6):1141-2.
- Chen YK, Lin LM, Lin CC, Yan YH. Myxoid tumor of the oral cavity with features of superficial angiomyxoma: report of a case. J Oral Maxillofac Surg. 1998;56(3):379-82.
- Meer S, Beavon I. Intraoral superficial angiomyxoma. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2008;106(5):e20-3.
- Gardner AW. Superficial angiomyxoma of the floor of the mouth--a case report. Br J Oral Maxillofac Surg. 2007;45(5):418-9.
- Mokhtar SM, Shahardin R, Abidin MZ, Rahman RA. Superficial Angiomyxoma of the Floor of the Mouth in an Infant. Asian J of Oral and Maxillofacial Surg. 2008 Mar; 20 (1): 26-28.
- S v R, Raju MS, J d S, et al. Intraoral superficial angiomyxoma of the upper alveolus: report of a unique case. Case Rep Med. 2012;2012:859021.
- Bajaj MS, Mehta M, Kashyap S, et al. Clinical and pathologic profile of angiomyxomas of the orbit. Ophthal Plast Reconstr Surg. 2011;27(2):76-80.
- Rosado rodríguez P, De vicente JC, De villalaín L, Blanco V. [Superficial angiomyxoma of the parotid region and review of the literature]. Acta Otorrinolaringol Esp. 2012;63(2):147-9.
- 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(7):519-30.
- Calonje E, Guerin D, Mccormick D, Fletcher CD. Superficial angiomyxoma: clinicopathologic analysis of a series of distinctive but poorly recognized cutaneous tumors with tendency for recurrence. Am J Surg Pathol. 1999;23(8):910-7.
- Bedlow AJ, Sampson SA, Holden CA. Congenital superficial angiomyxoma. Clin Exp Dermatol. 1997;22(5):237-9.
- Khadilkar UN, Khadilkar NP, Rao PS, Chakravorty S, Goel G. Superficial angiomyxoma of the external ear not associated with Carney's complex: a case report. Kathmandu Univ Med J (KUMJ). 2007;5(4):546-9.
- Rodríguez-vázquez M, García-arpa M, Delgado M, Cortina P, Vera E, Romero G. [Superficial angiomyxoma]. Actas Dermosifiliogr. 2005;96(5):311-4.
- Sylvester DC, Kortequee S, Moor JW, Woodhead CJ, Maclennan KA. Aggressive angiomyxoma of larynx: case report and literature review. J Laryngol Otol. 2010;124(7):793-5.
- Xiao P, Liu D, Peng J, et al. Aggressive angiomyxoma of the sphenoidal sinus. Pathology. 2011;43(1):79-81.
- Pai CY, Nieh S, Lee JC, Lo CP, Lee HS. Aggressive angiomyxoma of supraclavicular fossa: a case report. Head Neck. 2008;30(6):821-4.