



## CASUISTIC PAPER

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# Atypical extra nasopharyngeal angiofibroma in an unusual location: tonsil posterior pillar (oropharynx)

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## ABSTRACT

**Introduction.** Angiofibroma is a benign, locally aggressive highly vascular tumor that typically affects young adolescent males and has a pathognomonic epicenter of origin in the nasopharynx. The atypical angiofibromas share the same histological features as that of Juvenile nasopharyngeal angiofibroma, however they differ significantly in their clinical features.

**Aim.** Here we are reporting a very rare case of atypical angiofibroma in an adult male presenting as a non-vascular mass in the oropharynx with posterior pillar as the subsite.

**Description of the case.** A 26-years old male patient presented with chief complaints of foreign body sensation and mass in throat for one and a half months which was gradually progressive in size. Histopathological examination revealed a lesion with an intricate mixture of blood vessels, irregular fibrous stroma with loose edematous and fibrous area along with multinucleated stromal cells which was suggestive of angiofibroma.

**Conclusion.** Oropharyngeal angiofibroma, being an atypical angiofibroma in terms of site and presenting complaints, presents a diagnostic challenge. Though rare, it should always be kept as a differential diagnosis in any oropharyngeal mass irrespective of its vascularity, typical age or sex of the patient.

**Keywords.** atypical angiofibroma, extra nasopharyngeal (ENA), oropharynx

## Introduction

Angiofibroma is a benign, locally aggressive highly vascular tumor that usually arises from the lateral wall of the sphenopalatine foramen and pterygoid base.<sup>1</sup> But it has an early submucosal spread to nasopharynx and it mainly affects young adolescent males thus also known as Juvenile Nasopharyngeal Angiofibroma (JNA). However, it has also been reported at very young ages, el-

derly, and female patients and at the sites other than the nasopharynx. These rare variants have been termed as atypical angiofibromas.<sup>2</sup> The atypical angiofibromas share the same histological features as that of JNA, however they differ significantly in their clinical features.

Angiofibromas rarely originate outside the nasopharynx. Reports of primary extra nasopharyngeal angiofibromas (ENA) have appeared sporadically in the

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**Participation of co-authors:** A – Author of the concept and objectives of paper; B – collection of data; C – implementation of research; D – elaborate, analysis and interpretation of data; E – statistical analysis; F – preparation of a manuscript; G – working out the literature; H – obtaining funds

Received: 25.09.2019 | Accepted: 28.01.2020

Publication date: March 2020

literature. The most common site for atypical angiofibromas is the nasal septum.<sup>3</sup> Other less frequently involved sites are the maxilla, inferior turbinate, middle turbinate, ethmoid sinus, sphenoid sinus, oral cavity, oropharynx, larynx, ear, trachea, larynx, middle cranial fossa, infratemporal fossa, tonsil, retromolar region and conjunctiva.<sup>4,5</sup> To date, there have been only 10 cases reported of angiofibromas originating from the oropharynx in the literature.

### Aim

Here we report another rare case of atypical angiofibroma in an adult male patient originating from the oropharynx with posterior pillar as its subsite and this being the second case report in PubMed and google scholar literature.

### Description of the case

A 26 years old male patient presented in the O.P.D. of ENT department with the chief complaints of foreign body sensation and mass in the throat for one and a half months which was gradually progressive in size. He also complained of difficulty in breathing on lying down for one month. He had no history of any oral bleed or pain in the throat.

On examination, a single, pale, multilobulated mass with smooth surface was seen in the oropharynx arising from the right side posterior tonsillar pillar inferiorly reaching till right laryngopharynx (Figure 1). The mass was non pulsatile, soft on palpation, and did not bleed on touch. The rest of the ENT examination was normal. A contrast enhanced MRI was done which showed a polypoidal heterogeneously enhancing mass of size 8×17×20mm



Fig. 1. Intraoral mass from the right side posterior tonsillar pillar inferiorly reaching till right laryngopharynx

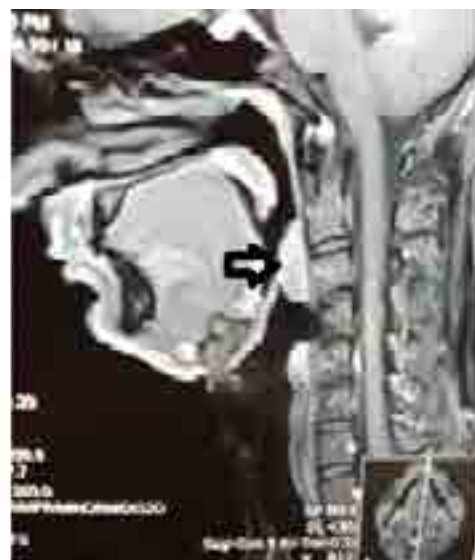


Fig. 2. Contrast enhanced MRI

on the right side of oropharynx (Figure 2). Excision of the mass was planned under general anesthesia.

Intraoperative findings revealed a 1.5 × 1 cm multilobulated mass attached to posterior tonsillar pillar on right side. The mass was completely excised using a coblator and the site of origin was also coblated (Figure 3). There was minimal intraoperative bleeding. The excised mass was sent for histopathological examination. Histopathological examination revealed a lesion with an intricate mixture of blood vessels, irregular fibrous stroma with loose edematous and fibrous area along with multinucleated stromal cells (Figure 4a, 4b). The images highlight the characteristic histopathological features of angiofibroma at 4X (fig 4a), 10X (fig 4b) magnifications. Immunohistochemistry panel applied:

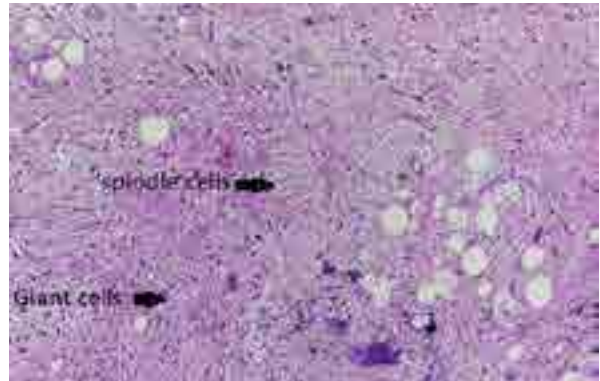
- S-100: Non specific positive. Negative in tumor cells.
  - Myo D1: Negative
  - Desmin: Negative
  - Ki-67: 1-2% Positive (Low proliferative index)
- Above features found suggestive of angiofibroma.



**Fig. 3.** Excised mass intended for histopathological examination



**Fig. 4a.** Histopathological examination of excised mass (4X)(H&E stain)



**Fig. 4b.** Histopathological examination of excised mass (10X)(H&E stain)

The patient was discharged on the third post-operative day and has remained disease free on his regular follow up for more than one year (Figure 5).



**Fig. 5.** Postoperative oropharynx

## Discussion

Angiofibroma is the most common benign vascular tumor in the nasopharynx, making up 0.5% of all head and neck tumors.<sup>4</sup> They are typically seen in adolescent males and are thus known as 'juvenile nasopharyngeal angiofibroma'. Celik et al. in 2005 proposed that angiofibromas presenting with at least one of the following criteria such as origin or location other than nasopharynx, presenting complaints other than nasal obstruction or epistaxis, age younger than seven or older than 25, female sex, atypical histopathology and multifocality were considered as atypical. Our case meets criteria via three parameters: age, location and presenting complaints, thus labeled as 'atypical angiofibroma'.<sup>2,5-8</sup>

Extra nasopharyngeal Angiofibroma (ENA) is a separate and rare entity. As compared to JNA, ENA occurs at older age with mean age of 22 years and the male to female ratio was 2.75:1. That is, there is a higher incidence of extra-nasopharyngeal angiofibroma in females compared to JNA. Windfur and Vent published a litera-

**Table 1.** Atypical extranasopharyngeal angiofibroma arising in oropharynx: literature review

S. No.	Author/ Year	Age/ Sex	Subsite	Presenting Complaints	Symptoms duration	Management	IHC	Reference
1.	Beeden (1971)	1/M	Posterior pharyngeal wall	Oral and nasal bleed	5 months	Radiotherapy further f/b transoral excision	Not Mentioned	<sup>9</sup>
2.	Kim (1972)	22/M	Not Defined	Dyspnea, Dysphagia, Altered Speech	4 Months	Transoral Resection	Not Mentioned	<sup>12</sup>
3.	Ali (1982)	28/F	Left Tonsil	Foreign Body Sensation, Occasional Oral Bleed	24 Months	Transoral Resection	Not Mentioned	<sup>13</sup>
4.	Chung (1995)	21/M	Soft Palate on Rt side	Accidental Finding	-	Transoral Resection	Not Mentioned	<sup>14</sup>
5.	Cejas Mendez (2000)	35/M	Right Tonsil	Dysphagia, Foreign Body Sensation	Few months	Right Tonsillectomy	Not Mentioned	<sup>15</sup>
6.	Celik (2005)	15/M	Right Tonsil	Dysphagia	1 Year	Transoral Rt Tonsillectomy	Not Mentioned	<sup>2</sup>
7.	Eftekharian (2008)	19/M	Right Posterior Tonsillar Pillar	Foreign Body Sensation in Throat	6-7 Years	Transoral Resection	Vimentin (Strong) Actin (Occasional)	<sup>11</sup>
8.	Mendoza Ramirez (2012)	60/M	Right Tonsil	Dysphagia	8 Months	Right Tonsillectomy	CD 34 CD 31 D2-40	<sup>16</sup>
9.	Szymanska (2013)	49/M	Right Tonsil	Dysphagia	1 Month	Right Tonsillectomy	Not Mentioned	<sup>17</sup>
10.	Nitin (2018)	3/M	Posterior Part of Soft Palate and Uvula	Dysphagia, Mouth Breathing, Intermittent Noisy Breathing	15 Days	Transoral Resection	Not Mentioned	<sup>10</sup>

ture review of 174 cases of extra nasopharyngeal angiofibroma from 170 publications published till 2015.<sup>3</sup> The most common site being nasal septum followed by maxillary sinus.<sup>3</sup> Out of the 174 cases, 39 (22.4%) cases had their origin outside nose and paranasal sinuses.

Many theories for pathogenesis have been put forward for origin of JNA but all have been debated. Origin from conjoined pharyngobasilar and buccopharyngeal fascia was suggested by Burner (1942). This theory might hold true for etiopathogenesis of oropharyngeal angiofibroma.

Till now only 10 cases of oropharyngeal angiofibromas have been described in the literature. The first case was reported by Beeden AD in 1971.<sup>9</sup> Out of the 174 cases reviewed by Windfur and Vent till 2015, only 9 cases had their origin in oropharynx. Later on in January 2018 another case of oropharyngeal angiofibroma originating from soft palate was reported in a 3 year old child by Nitin et al.<sup>10</sup> The ENAs can originate from any mucosal structure within the head and neck region, including the oral and nasal cavities. According to the literature, ENAs are most frequently localized in the maxillary sinus.<sup>3,7-9</sup>

Out of the 10 reported cases, five cases had their origin from tonsil, two from the soft palate, one from posterior pharyngeal wall, one from the posterior tonsillar pillar while in one case subsite of origin was not defined.

Our case had its origin from posterior tonsillar pillar on right side, very similar to the case reported by Eftekharian A.<sup>11</sup> Literature review of atypical extranasopharyngeal angiofibroma arising in oropharynx is presented in Table 1.

The most common presenting symptom for JNA is nasal obstruction and epistaxis. However, for oropharyngeal angiofibroma it is dysphagia. Other symptoms are foreign body sensation in throat, dyspnea, and change in voice if it extends in the laryngopharynx. On examination these generally appear as a vascular mass, however in our case it was a pale polypoidal mass not bleeding on touch.

Contrast enhanced CT/MRI is the investigation of choice for angiofibroma which reveals the extension and vascularity of mass along with its relation to surrounding structures.<sup>18</sup> Contrast enhanced CT Scan helps us to differentiate between JNA and ENA as JNA produces strong and homogenous enhancement as compared to heterogenous or even no enhancement by ENA due to its poor vascularity. Complete surgical excision of the mass is the treatment modality of choice which prevents further recurrence. The surgical specimen should be sent for histopathological analysis.

The definitive diagnosis of angiofibroma is made by histopathologic analysis of the surgical specimen. Ma-

lignant transformation has been reported in literature in JNA cases with radiotherapy treatment being the main cause.<sup>19</sup> However, in ENA cases no malignant transformation has been reported till date as the literature available is very sparse. Close follow up is warranted.

### Conclusion

Oropharyngeal angiofibroma being an atypical angiofibroma in terms of site and presenting complaints presents a diagnostic challenge. Though rare but should always be kept as a differential diagnosis in any oropharyngeal mass irrespective of its vascularity, typical age or sex of the patient.

### References

- Liu ZF, Wang DH, Sun XC, Wang JJ, Hu L, Dal PD. The site of origin and expansive routes of juvenile nasopharyngeal angiofibroma (JNA). *Int J Pediatr Otorhinolaryngol*. 2011;75 (9):1088-92.
- Celik B, Erisen L, Saraydaroglu O, Coskun H. Atypical angiofibromas: a report of four cases. *Int J Pediatric Otorhinolaryngol*. 2005;69:415-421.
- Windfuhr JP, Vent J. Extra nasopharyngeal angiofibroma revisited. *Clinical Otolaryngology*. 2018;43:199-222.
- Nomura K, Shimomura A, Awataguchi T, Murakami K, Kobayashi T. A case of angiofibroma originating from the inferior nasal turbinate. *Auris Nasus Larynx*. 2006; 33(2):191-193.
- Perko M, Uehlinger E, Hjorting-Hansen E. Nasopharyngeal angiofibroma of the maxilla: report of case. *J Oral Surg*. 1969;27:645-648.
- McDaniel RK, Glen D. Juvenile nasopharyngeal angiofibroma with lateral extension into the cheek: report of case. *J Oral Maxillofac Surg*. 1995;53:473-476.
- Guo G, Paulino AFG. Lipomatous variant of nasopharyngeal angiofibroma: a case report. *Arch Otolaryngol Head Neck Surg*. 2002;128:448-450.
- Rha KS, Byun SN, Kim TH, Kim YM. Bilateral juvenile nasopharyngeal angiofibroma. *Otolaryngol Head Neck Surg*. 2003;128:891-893.
- Beeden AG, Alexander FW. An unusual pharyngeal tumor. *J Laryngol Otol*. 1971; 85: 733-735.
- Gupta N, Dass A, Saini V, Anil Pol S, Mittal L. Extra nasopharyngeal Angiofibroma: A Diagnostic Dilemma. *Philipp J Otolaryngol Head Neck Surg*. 2018;33(1):39-42.
- Eftekharian A, Samiei F, Rakhshan M. Angiofibroma with oropharyngeal origin. *Pak J Med Sci*. 2008;24:319-320.
- Kim JK. A case of angiofibroma in the oropharynx. *Korean J Otolaryngol*. 1972;15:69-72.
- Ali S, Jones WI. Extranasopharyngeal angiofibromas. (Sex incidence and age distribution). *J Laryngol Otol*. 1982;96:559-565.
- Chung YS, Kim JK, Kweon HW, Lee WS, Lee HH. Angiofibroma: an unusual presentation in soft palate. *Korean J Otolaryngol*. 1995;38:779-785.
- Cejas Mendez L, Espejo Castro E, Artazkoz del Toro JJ, Gil Curbelo JA, Saavedra de la Torre JA, de Miguel Hernandez B. Tonsillar angiofibroma. *An Otorrinolaringol Ibero Am*. 2000;27:605-611.
- Mendoza-Ramirez S, Martinez-Hernandez J, Vicuna-Honoratob I, Mendoza-Ramirez M, Murguia-Pereza M. Angiofibroma atipico dela amigdala palatina. *Rev Esp Patol*. 2012;45:125-127.
- Szymanska A, Szymanski M, Morshed K, Czekajska-Chehab E, Szczerbo-Trojanowska M. Extra nasopharyngeal angiofibroma: clinical and radiological presentation. *Eur Arch Otorhinolaryngol*. 2013;270:655-660.
- Mehmet A, Orhan S, Supni M. Juvenile nasopharyngeal angiofibroma: Radiological Evaluation & preoperative Embolization. *KBB Forum*. 2006;5(1):58-61.
- Makek MS, Andrews JC, Fisch U. Malignant transformation of a nasopharyngeal angiofibroma. *Laryngoscope*. 1989;99:1088-1092.