Maxillary extranasopharyngeal angiofibroma: A case report and a review of similar cases

Case Report

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ABSTRACT

Extranasopharyngeal angiofibromas (ENA) are a rare subtype of angiofibromas that exhibit different demography, clinical presentation, differentials and management. We present a case of an extranasopharyngeal angiofibroma that has occurred in the maxillary sinus, as well as review published literature on similar cases.

A 14-year-old male presents with right-sided facial swelling, facial numbness and epistaxis secondary to a vascular lesion occupying the right nasal cavity. Biopsies revealed an angiofibroma. Imaging showed erosion of the anterior maxillary wall, thinning of the anterior wall of the pterygopalatine fossa whilst sparing of the nasopharynx, pterygoid plates and vidian canals. The tumour was excised uneventfully via a right hemi-facial degloving surgery.

Maxillary ENAs presents most often with facial swelling, followed by nasal obstruction and epistaxis. The anterior and medial maxillary sinus walls were more commonly affected. In our case, the posterior and inferior walls were unaffected. These tumours were treated primarily by surgery, and generally will require open surgical approaches. The main differentials of ENAs are angiomatous nasal polyps (ANP) and angiosarcoma.

Key Words: Angiofibroma, atypical, epistaxis, extranasopharyngeal, maxilla.

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CASE REPORT

We present a case of a 14-year-old male who presented with right facial swelling for two months. He additionally reports nasal obstruction and epistaxis for the preceding four months. There was right maxillary protrusion with right eye hypertelorism, with right maxillary nerve hypoaesthesia (CN V2). There was no diplopia or visual impairment

Rigid nasoendoscopy revealed a raspberry-like mutilobulated vascular lesion occupying the right nasal cavity (Fig. 1).

Computerised tomography scans showed a large tumour within the right maxillary sinus (Fig. 2, 3) with heterogenous contrast uptake. The tumour displayed bony erosion of the anterior wall of the maxillary sinus and bony defect in the lateral wall, close to the zygomatic recess. The mass eroded the medial maxillary sinus wall and caused bowing of the nasal septum to the opposite side. The typical Holman-Miller sign with bowing of the posterior wall into the maxillary sinus was absent. The pterygoid body and the pterygoid plates were intact. Periantral fat was still visible and the infratemporal fossa contents were not involved in the pathology. The anterior wall of the pterygopalatine fossa appears thinned out or possibly eroded. The vidian canal appeared intact on imaging.

Magnetic resonance imaging (MRI) demonstrates tumour comprising of blood vessels as seen by flow voids within it. The tumour is seen exerting its mass effect onto the right orbit and septum, as well as expansion of all maxillary sinus walls (Fig. 4). The sphenoid sinus was filled with retained secretions and blood products. Both imaging modalities confirmed absence of lymphadenopathy.

Due to the aggressive nature of this lesion, a differential of malignancy was of concern. The patient was subject to an urgent biopsy. Intraoperatively, there was minimal bleeding, easily arrested with packing and cautery.

Histopathological examination showed a tumour covered with benign respiratory epithelium that had focal metaplasia. It comprised of irregularly-shaped and variably-sized blood vessels lined with a single cell layer of plump endothelium that took up CD-34 staining. The underlying stroma was loose and hypocellular, with some extravasated fibrin, with some areas of haemorrhage. There were no malignant cells. A diagnosis of angiofibroma was made with a differential diagnosis of angiomatous polyp.

This tumour was resected via a hemifacial degloving approach (Fig 5-6). The tumour presented itself just deep

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to the facial soft tissues. There was a large jagged anterior maxillary wall defect from which the tumour was removed successfully, and this was likely the origin of the tumour. There was no communication of the tumour with the pterygopalatine fossa or attachment to other walls of the maxillary sinus

Macroscopically, it was a soft fleshy and vascular tumour arranged in lobules. No areas of necrosis or ulceration were noted. (Fig. 7).

Final histology of the tumour showed proliferation of connective tissue stroma interspersed with a thick vascular network, some with smooth muscle cuffing. There were large areas of haemorrhage and fibrin, with stromal haemorrhage. The blood vessels are in irregular sizes and shapes, lined by a single lining endothelial cell layer. Some of the stromal cells have enlarged and plump nuclei. No areas of necrosis seen. The cells demonstrated minimal atypia, with no cellular infiltration seen (Fig. 8). A final diagnosis of an extranasopharyngeal angiofibroma arising from the maxillary sinus was made.

The follow-up of this case over the next six months showed no residual tumour and no recurrence. His right maxillary swelling is still present, likely needing more time to remodel. The hypertelorism resolved.

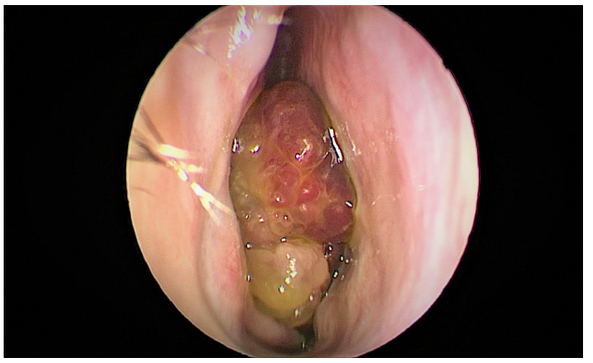

Fig. 1: Endoscopic view of a vascular-looking multilobulated mass in the right nasal cavity

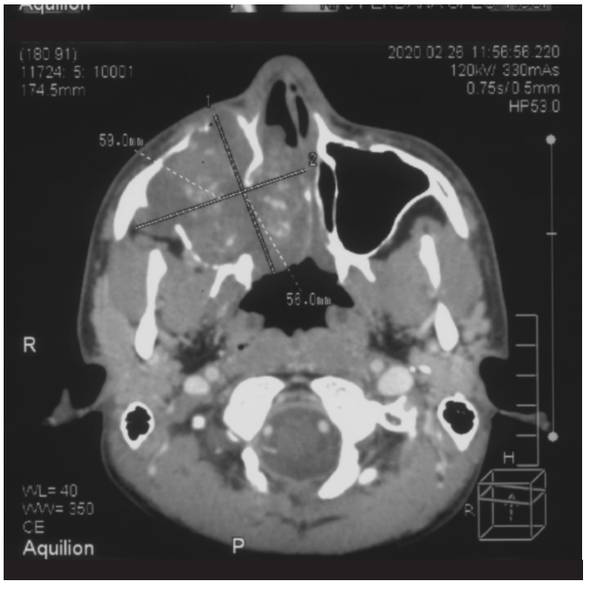

Fig. 2: An axial view of a contrast-enhance computed tomography scan of the paranasal sinus in soft tissue window shows right maxillary tumour with uneven contrast enhancement

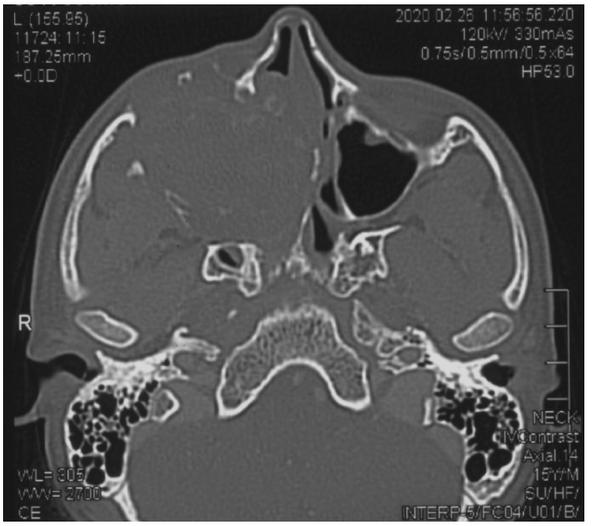

Fig. 3: An axial view of a contrast-enhanced computed tomography scan of the paranasal sinus in body window shows tumour erosion of the anterior, medial and lateral maxillary sinus wall. Holman-Miller sign was absent. The anterior wall of the pterygopalatine fossa on the right is thinned out or eroded.

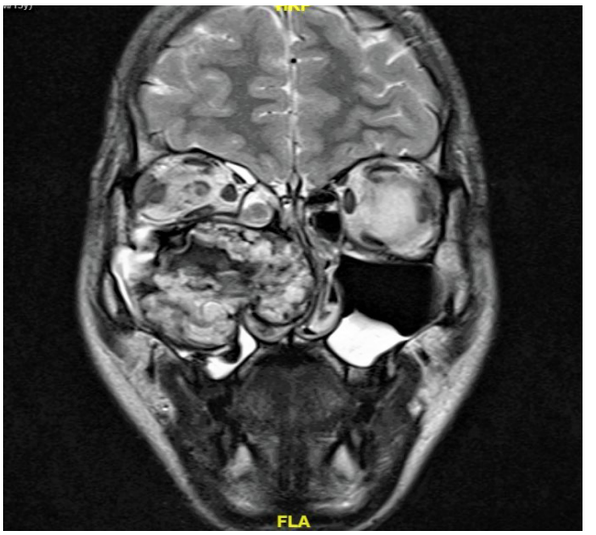

Fig. 4: A T2-weighted coronal magnetic resonance imaging showed superior and lateral displacement of the right orbital contents

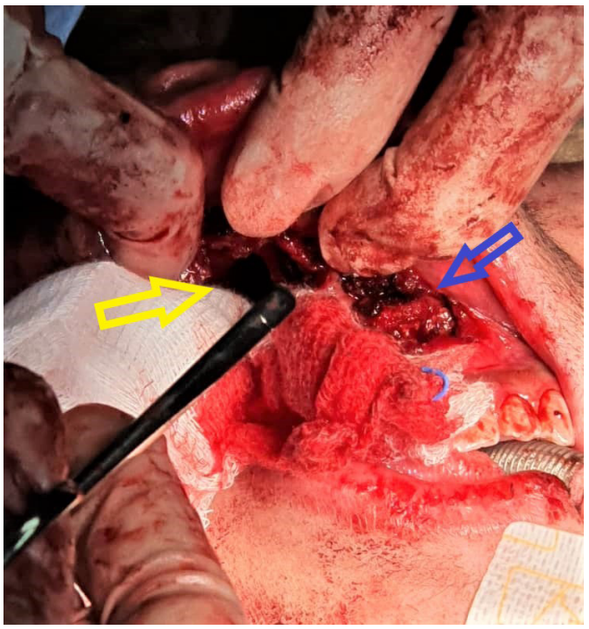

Fig. 5: Right hemi-facial degloving approach was used to excise the tumour. Blue arrow: Right pyriform aperture. Yellow arrow: Eroded anterior maxillary sinus wall.

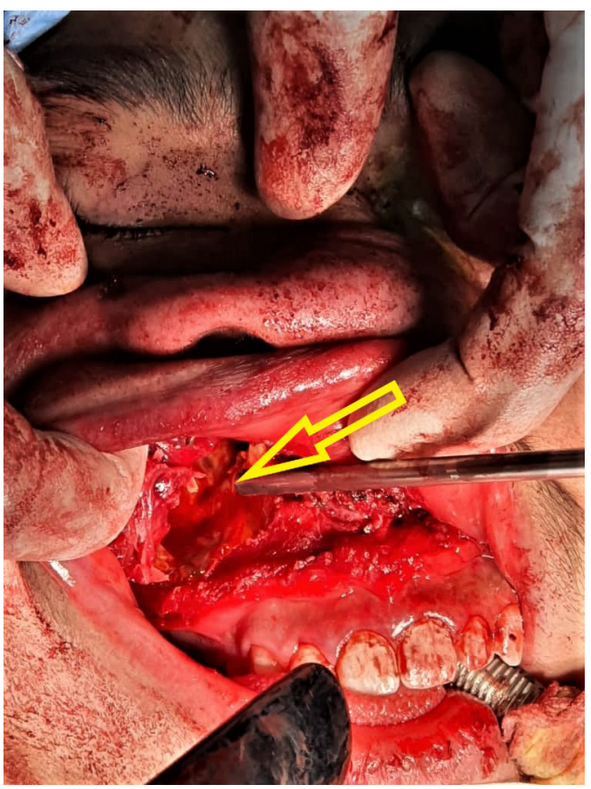

Fig. 6: Bony defect in the anterior maxillary sinus wall is seen deep to the soft tissue of the cheek. Yellow arrow: Bony defect in the anterior maxillary wall

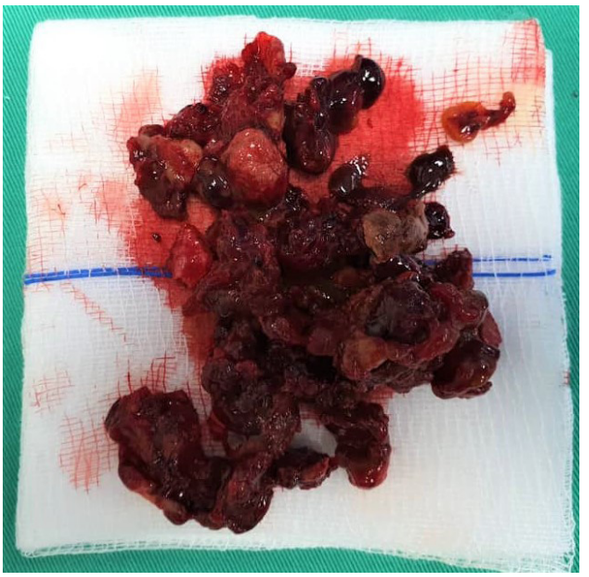

Fig. 7: Macroscopic view of the excised tumour.

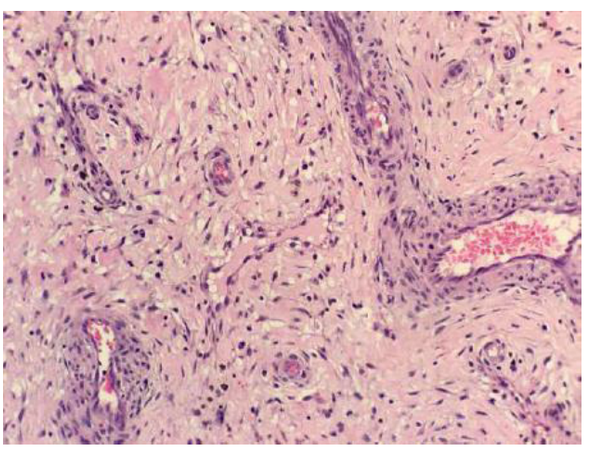

Fig. 8: H&E staining under 20x magnification

Table 1: Maxillary extranasopharyngeal angiofibroma – cases in literature ENA: extranasopharyngeal angiofibroma, JAF: Juvenile angiofibroma, AF: Angiofibroma

Author	Date	Title	Age (yrs)	Gender	Symptoms	Sites	Embolisation	Other Adjuncts	Surgical Approach	Bleeding	Recurrence
Alajmo [1]	1961	II fibroangioma prinitivo del le nasali e dei seni paranasali	6	Male	obstruction	Maxilla	no	RT	Lat Rhinotomy		9 years, fibrosarcoma
Alajmo [1]	1961	II fibroangioma prinitivo del le nasali e dei seni paranasali	58	Male	facial swelling	Maxilla / ethmoid	no	no	Resection	no	no
Bhagat [7]	2011	ENA in an Adult: A Rare Presentation	28	Male	facial swelling, bleeding post caldwell-luc	anterior, lateral, posterior	no	no	Weber-Fergusson	yes	no
Capodiferro [8]	2005	JAF: Report of a case with primary intra-oral presentation	21	Female	palatal swelling, loose teeth	floor	no	no	transmaxillary esection	no	no
Chakrabarti [1]	1973	Extranasopharyngeal JAF – a case report	17	Male	obstruction, epistaxis, facial swelling	maxilla, inf orbital fissure, check	no	no	transmaxillary esection	no	no
Hora [9]	1962	Paranasal JAF	13	Male	obstruction, epistaxis	medial, posterior, lateral	no	no	combined approach	yes	no
Irby[1]	1961	AF of cheek and infratemporal fossa	21	Male	facial swelling	cheek, parapharyngeal space, infratemporal fossa	no	no	transoral	yes	no
Ismi [10]	2015	ENA of maxillary sinus seen during surgery for epistaxis	22	Male	epistaxis	maxilla	no	no	Endoscopic transnasal	no	no
Juul [11]	1982	AF of the maxillary antrum	27	Female	headache, obstruction, rhinorrhoea	lateral, posterior	no	no	Caldwell-Luc	no	no
Kitano [12]	1992	JAF of the maxillary sinus. A case report	13	Male	palatal swelling, loose teeth	maxilla	no	RT	Partial maxillectomy	yes	no
Krutchkoff [1]	1977	JNA. An unusual presentation as an oral mass	12	Male	oral bleeding, loose teeth	maxilla	no	no	transoral resection	yes	no
Malvic [13]	2012	JA of the maxillary sinus	14	Male	obstruction, rhinorrhoea	lateral	no	no	endoscopic transnasal	yes	no
Maniglia [14]	1969	Maxillary sinus AF treated with cryosurgery	15	Male	obstruction, trivial epistaxis	posterior, medial	no	Freon cryoprobe	Weber-Fergusson	yes	no

Manjalay [15]	1992	A case of congenital AF	0	Male	facial swelling	anterior, medial and anterior	no	CO_2 laser	mid-facial degloving	no	no
Munson [16]	1941	AF of the left maxillary sinus	15	Male	epistaxis, facial swelli	medial, posterior	no	ECA ligation (bilateral), radium rods	Caldwell-Luc	yes	no
Ogura [17]	1962	Very rare JAF in the right maxillary sinus	16	Male	cheek swelling, pain, epistaxis	anterior	no	no	Weber-Fergus son	yes	no
Panesar [1]	2004	JAF of the maxillary sinus	1	Male	pain, facial swelling	medial	no	no	mid-facial degloving	no	no
Pathak [18]	1970	Extranasophary ngeal JAF - a case report	18	Male	obstruction, epistaxis	anterior, medial	no	ECA & AEA ligation	Caldwell-Luc	yes	no
Perko[1]	1965	Nasopharyngeal AF of the maxilla: report of case	33	Female	loose teeth	maxilla	no	RT (for recurren ce)	transmax illary resection, maxillec tomy	yes	yes
Ramanja ney ulu[1]	1974	JAF of antral origin	17	Male	obstruction, facial swelling	maxilla	no	no	lateral rhinotomy	yes	no
Ryc[1]	1973	JAF of maxillary sinus	17	Male	epistaxis	maxilla	no	no	Caldwell-Luc	yes	no
Schick [19]	1997	AF with atypical localizations in early childhood	9	Male	facial swelling, epistaxis	medial	no	no	transnasal endoscopic	no	no
Tsuru [1]	1937	Uber die Blutgeschwulst in der Nasenh€ohle und Nebenh€ohle	33	Female	obstruction, epistaxis, facial swelling	maxilla	no	no	transnasal resection	no	no
Veeranjan ey ulu [20]	2015	ENA of maxillar y sinus: a rare entity	27	Male	obstruction, facial swelling	anterior	no	no	lateral rhinotomy	no	no
Windfuhr [21]	2004	ENA of nasal cavity and the paranasal sinuses	13	Female	facial swelling	maxilla	no	Chemoth erapy (misdiag nosed as PNET)	lateral rhinotomy	no	no
Wan	2020	Maxillary extranasopharyn geal angiofibro ma – A case report and review of similar cases	14	Male	facial swelling, epistaxis, obstruction	anterior	no	no	mid-facial deglovin	no	no

Table 2: Clinical characteristics of maxillary extranasopharyngeal angiofibromas

Gender	
Male	21
Female	5
	18.4615385
Age (yrs)	11.5697218
S.D.	
Symptoms	
Obstruction	11
epistaxis	11
facial/cheek swelling	14
pain	2
oral / palate swelling	2
dental symptoms	4
oral bleeding	1
Sites	
not specified	9
anterior	5
posterior	4
lateral	3
medial	5
inferior / floor	1
orbital floor	0
Approach	
Open	23
Endoscopic	3

DISCUSSION

In this case, physical examination and imaging showed an aggressive tumour, eroding anteriorly from the maxillary sinus, distorting the facial architecture. These features coupled with the absence of the history of profuse epistaxis or post biopsy bleeding suggested an angiosarcoma. Histopathology of angiosarcomas show high-grade lesions; infiltrative atypical endothelial cells with profound nuclear pleomorphism.[4] This was absent in our specimens.

The other differential of an angiomatous nasal polyp (ANP) was also entertained. Histology of these benign lesions show no atypia or mitoses, but a great quantity of blood vessels, intravascular thrombosis and massive necrosis. Its dilated capillaries also show no elastic or muscular layer. The architecture of the specimens taken from biopsy and excision favour angiofibroma, compared to ANP, although there are many similarities. ANPs are thought to have arisen from antrochoanal polyps that have undergone haemorrhagic congestion and necrosis due to vascular constriction at the maxillary ostium. ANPs have a chronic disease history, and are more likely in older patients, those who have diabetes, and those who are on aspirin.[5,6]

Nasopharyngeal angiofibromas (NA) are benign vascular tumours that occur almost exclusively in adolescent males. These tumours are thought to have arisen from the sphenopalatine foramen and pterygopalatine fossa, pterygoid wedge or vidian canal, presenting into the nasopharynx once large enough to present through the sphenopalatine foramen.[3]

A rare entity of nasopharyngeal angiofibromas, as in our case are termed extranasopharyngeal angiofibromas (ENA) as they occur away from the nasopharynx. These tumours demonstrate histologic congruence with the conventional JNA tumours, but differ in age and gender distribution.[2]

Windfuhr *et al*.[1] compiled a comprehensive list of 174 ENA cases from a search of literature in Medline and Google Scholar. ENAs have been demonstrated across a wider range of ages, from congenital to middle age, as well as a more balanced gender ratio. These tumours have been described as occurring most commonly along the nasal septum, inferior turbinate followed by maxillary sinus.

We analysed the 26 cases of reported maxillary ENAs, including the above described.[1, 7-21]

The mean age of patients with maxillary ENAs is 18.4 years (S.D. ± 11.6 years), ranging from congenital to 58 years of age. The M:F ratio was 4.2:1, (21 male patients vs 5 female patients). Where described, the subsites most commonly involved were the anterior and medial walls (n=5), posterior (n=4) and lateral (n=3) walls.

The most common presentation for these patients were facial or cheek swelling (n= 14). Both the symptoms of epistaxis and obstruction were present in 11 out of 26 patients.

Of the 26 patients reported, 23 of them were treated via open approaches, and only 3 were found to be suitable for endoscopic excision. Roughly half (12 of 26) had bleeding as an intraoperative complication.

One case treated with preoperatively with radiotherapy recurred 9 years later as a fibrosarcoma, which was treated with radioactive radium seeds. Another recurrence was treated with radiotherapy alone.

2 cases had carotid artery ligations, while here was use of one report of surgery assisted with CO_2 laser and one with Freon cryotherapy probe. One case of maxillary ENA was misdiagnosed as a primary neuroendocrine tumour and treated with chemotherapy before the definitive diagnosis.

CONCLUSION

Maxillary extranasopharyngeal angiofibromas are rare type of angiofibroma. Its presents most often as with facial swelling, followed by nasal obstruction and epistaxis. The anterior and medial maxillary sinus walls were more commonly affected. In our case, the posterior and inferior walls were unaffected. These tumours were treated primarily by surgery, and generally will require open surgical approaches. The main differentials of ENAs are angiomatous nasal polyps (ANP) and angiosarcoma.

CONFLICT OF INTEREST

There are no conflicts of interest.

REFERENCES

1. Windfuhr JP, Vent J. Extranasopharyngeal angiofibroma revisited. Clin Otolaryngol 2017;1-24
2. Szymańska A, Szymański M, Morshed K *et al*. Extransopharyngeal angiofibroma: Clinical and radiological presentation. Eur Arch Otorhinolaryngol 2013;270:655-660
3. Li W, Ni Y, Lu *et al*. Current perspectives on the origin theory of juvenile nasopharyngeal angiofibroma. Discovery Medicine published June 2019;www.discoverymedicine.com/Wanpeng-Li/2019/06/perspectives-on-origin-theory-of-juvenile-nasopharyngeal-angiofibroma/ (accessed 29 August 2020)
4. Nelson BL, Thompson LDR. Sinonasal tract angiosarcoma: A clinicopathologic and immunophenotypic study of 10 cases with a review of the literature. Head Neck Pathol 2007;1:1-12
5. Assiri KS, Al-Ahmari MS, Alshahrani MS, *et al*. Clinical and pathological features of angiomatous nasal polyps: A report of four cases and review of literature. Cureus 2020;12(4):e7642. DOI:10.7759/cureus.7642
6. Tam YY, Wu CC, Lee TJ *et al*. The clinicopathological features of sinonasal angiomatous polyps. Int J Gen Med. 2016;9:207-212
7. Bhagat S, Verma RK, Panda NK. Extranasopharyngeal angiofibroma in an adult: A rare presentation. Indian J Otolaryngol Head Neck Surg 2011;63(Suppl 1):S25-26
8. Capodiferrro S, Favia G, Lacaita MG *et al*. Juvenile angiofibroma: Report of a case with primary intra-oral presentation. Oral Oncology Extra 2005;42:1-6
9. Hora JF, Brown AK. Paranasal Juvenile Angiofibroma. Arch Otolaryngol 1962;76:457-459
10. Ismi O, Arpaci RB, Yesilova M, *et al*. Extranasopharyngeal angiofibroma of maxillary sinus seen during surgery for epistaxis. ENTcase 2015;1(1);47-51
11. Juul A, Larsen K, Illum P. Angiofibroma of the maxillary antrum. Arch Otorhinolaryngol 1982;236:135-138
12. Kitano M, Landini G, Mimura T. Juvenile angiofibroma of the maxillary sinus. Int J Oral Maxilofac Surg 1992:21;230-232
13. Malvic G, Manestar D, Krstulja M, *et al*. Juvenile angiofibroma of the maxillary sinus. Coll Antropol 2012;36(Suppl 2);197-199
14. Maniglia AJ, Mazzarella LA, Minkowitz S, *et al*. Maxillary sinus angiofibroma treated with cryosurgery. Arch Otolaryng 1969;89:527-532
15. Manjalay G, Hoare TJ, Pearman K, *et al*. A case of congenital angiofibroma. Int J Pediatr Otorhinolaryngol 1992;24:275-278
16. Munson FT. Angiofibroma of the left maxillary sinus. Ann Otol Rhinol Laryngol 1941;50:561-569.
17. Ogura Y, Fushimi N, Ohue H, *et al*. A very rare case of juvenile angiofibroma in the right maxillary sinus. Nippon Jibiinkoka Gakkai Kaiho 1965;68:1234-1240
18. Pathak PN. Extranasopharyngeal juvenile angiofibroma–a case report. J Laryngol Otol 1970;84:449-451
19. Schick B, Kind M, Schwarzkopf G, *et al*. Angiofibroma with atypical localizations in early childhood. HNO 1997;45:1022-1028.
20. Veeranjaneyulu P, Yadlapalli AK, Chintapalli A. Extranasopharyngeal angiofibroma of maxillary sinus: A rare entity. J Pharm Biomed Sci 2015;05(09);713-718
21. Windfuhr JP, Remmert S. Extranasopharyngeal angiofibroma of the nasal cavity and paranasal sinuses. Laryngo-Rhino-Oto 2004;83:308-316