Uncommon presentations of angiofibroma in sinonasal tract: a two-case report

Apresentações incomuns de angiofibromas no trato nasossinusal: relato de dois casos

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ABSTRACT
Juvenile nasopharyngeal angiofibroma (JNA) are rare, histologically benign, but locally aggressive, vascular tumors of the nasopharynx that affects predominantly male adolescents, its origins from other sites is denominated extranasopharyngeal angiofibroma (ENPA), which is an even rarer entity afflicting more females and young adults, representing a diagnostic and therapeutic challenge to rhinologists. In this report we present 2 cases of similar clinical presentation, with unilateral nasal obstruction and previous episodes of nasal bleeding. One patient is a 14-year-old male with a soft-tissue mass occupying nasopharynx, middle meatus, and right nasal cavity. The second patient is a 12-year-old male with a soft-tissue lesion rising from right sphenoid sinus and obliterating right nasal cavity, obstructing ipsilateral maxillary sinus, ethmoidal cells, and frontal sinus. Both patients underwent complete endoscopic excision of the lesions, the first one recurred on sphenoid sinus and the second showed no recurrence after 2 years of post-operative follow-up. The unusual sites and presentations make these rare cases and thus worth of reporting.

Keywords: juvenile nasopharyngeal angiofibroma, JNA, extranasopharyngeal angiofibroma, ENPA, endoscopic surgery.

RESUMO
O angiofibroma nasofaríngeo juvenil (JNA) é raro, histologicamente benigno, mas localmente agressivo, os tumores vasculares da nasofaringe que afectam predominantemente adolescentes do sexo masculino, as suas origens são denominadas angiofibroma extranasofaríngeo (ENPA), que é uma entidade ainda mais rara que aflige mais mulheres e adultos jovens, representando um desafio diagnóstico e terapêutico para os rinologistas. Neste relatório, apresentamos 2 casos de apresentação clínica semelhante, com obstrução nasal unilateral e episódios anteriores de hemorragia nasal. Um paciente é um homem de 14 anos com uma massa de tecido mole que ocupa a nasofaringe, o meato médio, e a cavidade nasal direita. O segundo paciente é um homem de 12 anos de idade com uma lesão de tecido mole que se eleva do seio esfenoidal direito e oblitera a cavidade nasal direita, obstruindo o seio maxilar ipsilateral, células etmoidais, e o seio frontal. Ambos os pacientes foram submetidos a excisão endoscópica completa das lesões, a primeira recidiva no seio esfenoidal e a segunda não mostrou recidiva após 2 anos de seguimento pós-operatório. Os sítios e apresentações pouco usuais tornam estes casos raros e, portanto, dignos de serem relatados.

Palavras-chave: angiofibroma nasofaríngeo juvenil, JNA, angiofibroma extranasofaríngeo, ENPA, cirurgia endoscópica.

1 INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a rare, benign, locally aggressive, vascular tumor affecting predominantly male adolescents which usually originates from the posterior lateral wall of the nasopharynx. Its occurrence outside the nasopharynx is denominated extranasopharyngeal angiofibroma (ENPA) whose incidence is even lower, with few sporadic cases reported in the literature[1,2]. The gold standard treatment method is the surgical excision of the tumor[3].

The most common site of ENPA reported in literature is the maxillary sinus[4], although angiofibromas can occur anywhere in the mucosa of the upper respiratory tract. The clinical presentation of ENPA can be extremely variable however it often has epistaxis as primary symptoms[5]. The etiology of the ENPA is still unclear. Unlike JNA, its occurrence is more often in female young adults[6], with few cases reported in children[7].

Nasal polyps can be considered differential diagnosis for JNA and ENPA. Sphenochoanal polyp are rare benign tumors which originates from sphenoid sinus and expands through the choana into the nasopharynx. Its occurrence is more common in children and adolescents[8,9].

Thus, the aim of this study is to report a case of JNA expanding to sphenoid sinus and a case of ENPA originating from right sphenoid sinus, both are differential diagnosis of nasal polyps.

2 CASE REPORT

Case 1

A fourteen-year-old male with clinical history of snoring, mouth breathing, sneezing, nasal itching, nasal obstruction, and impaired sleep quality. In addition to such complaints, there was also a previous episode of bilateral moderate epistaxis, requiring assistance in the emergency department, but without need for blood transfusion.

On nasal endoscopy, enlarged inferior nasal turbinates, moderate septum deviation in areas II and III of Cottle of left nasal fossa (LNF) and an irregular mass occupying the posterior region, originating from contralateral nasal fossa were observed. In the right nasal fossa (RNF), a smooth, whitish-colored mucous mass was found between middle turbinate and superior septum, occupying the middle and posterior portion of right middle meatus, originating from nasal roof and extending to nasal floor.

Computed tomography (CT) scan of the nose and paranasal sinuses showed slight mucous thickening in left maxillary sinus and right ethmoidal cells, partial filling of right sphenoid sinus and a soft-tissue density material occupying nasopharynx, middle meatus and posterior half of RNF. Contrast-enhanced magnetic resonance imaging (MRI) of the nose and paranasal sinuses showed a solid expansive lesion with a polypoid aspect centered in the posterior portion of middle and inferior right nasal meatus, extending to right sphenoid sinus, nasopharynx and oropharynx. Its dimensions were 5,7 x 3.3 x 3.7 cm, with heterogeneous content, showing high and intermediate signal in the T1 and T2 weighted sequences, respectively (Figure 1). There was also

no evidence that the tumor originated from pterygopalatine fossa. After these findings, the initial diagnostic hypotheses were sphenochoanal polyp and ENPA.

Two months after the MRI, the patient underwent a total excision of the nasopharyngeal mass, with significant intraoperative nasal bleeding, however blood transfusion was not required. The tumor was extracted on from the sphenoid sinus with right sphenoidotomy. In immediate postoperative period, the patient presented a small amount of drainage of clear and bloody nasal discharge, showing improvement in previous symptoms and thus progressing satisfactorily.

Histopathological examination demonstrated a highly vascularized neoplasm, with collagenized hypocellular areas and star-shaped cells without atypia. Such findings were consistent with nasopharyngeal angiofibroma (Figure 2).

After 1 year of follow-up, no recurrence of ENPA was detected. Also, no nasal complaints nor sleep disorders were further reported. After 6 months, nasal endoscopy revealed patency of right sphenoid sinus ostium with irregularly shaped mucosa at its edges. After 2 years of post-operative follow-up, new CT scan of nose and paranasal sinuses showed a 30mm soft tissue density lesion in the sphenoid sinus, with obliteration of right sphenoethmoidal recess (Figure 3) and enlargement of the right pterygopalatine fossa.

The final diagnosis proposed for the case was JNA, with sphenoid sinus involvement.

Figure 1 – Nasopharyngeal angiofibroma. Preoperative computed tomography scans in bone window ((1A-1B) axial, (1C): coronal) showing a well-defined soft-tissue density material occupying nasopharynx, middle meatus, and posterior half of right nasal cavity. MRI scans ((1D and 1E) T1 weighted axial; (1F) T1 weighted coronal; (1G and 1F) T2 weighted axial; (1I) T2 weighted coronal) showing a solid expansive lesion centered in the posterior portion of middle and inferior right nasal meatus, extending to right sphenoid cells, nasopharynx, and oropharynx with heterogeneous content, showing high and intermediate signal in the weighted sequences in T1 and T2, respectively.

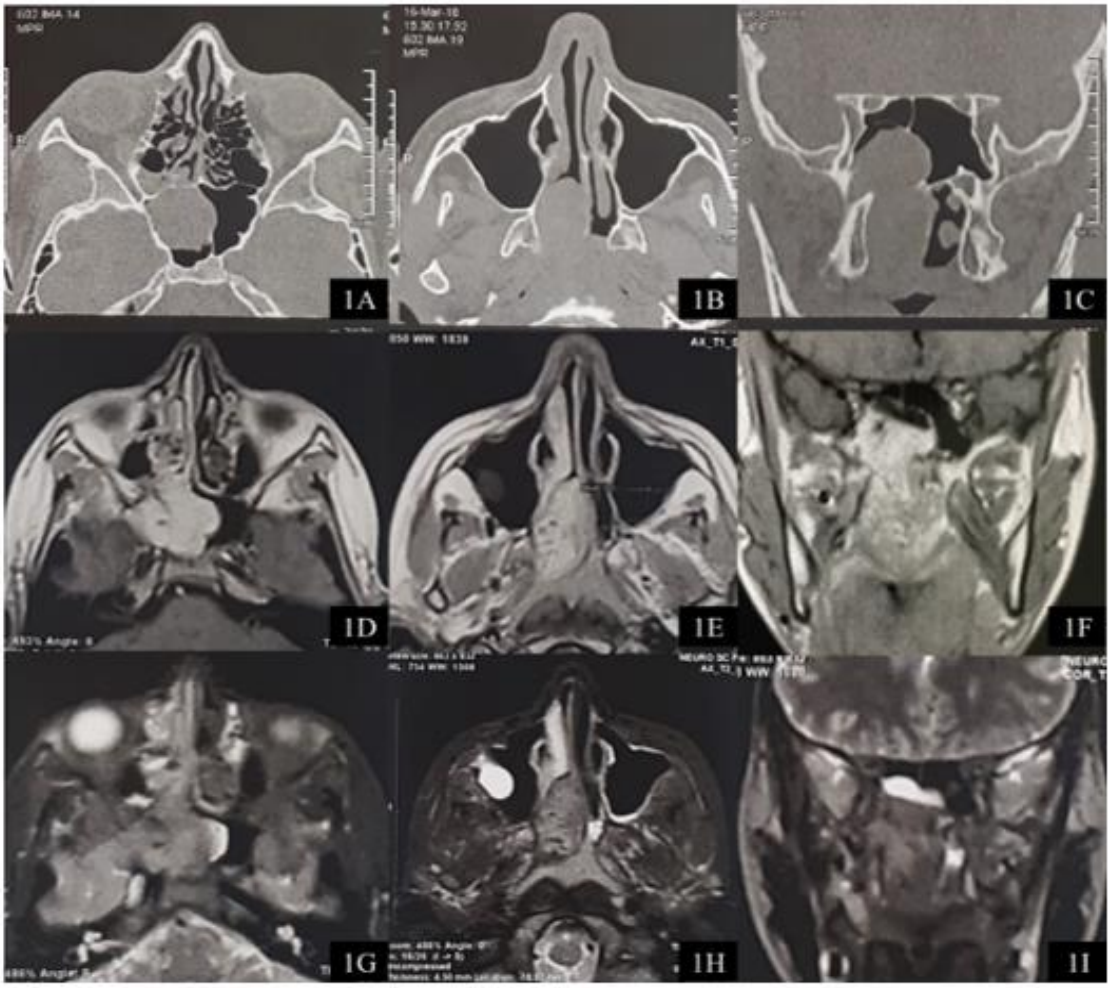


Figure 2 – Nasopharyngeal angiofibroma. (2A) Pre-operative contrast-enhanced, T1-weighted magnetic resonance imaging on sagittal section showing a capitating mass occupying sphenoid sinus, nasopharynx, middle meatus, and nasal cavity. Pathological features: (2B) macroscopic aspect of the lesion soon after endoscopic excision; (2C) H&E stain section showing highly vascularized lesion with star-shaped cells without atypia; (2D) Trichrome stain section showing collagenized hypocellular stroma.

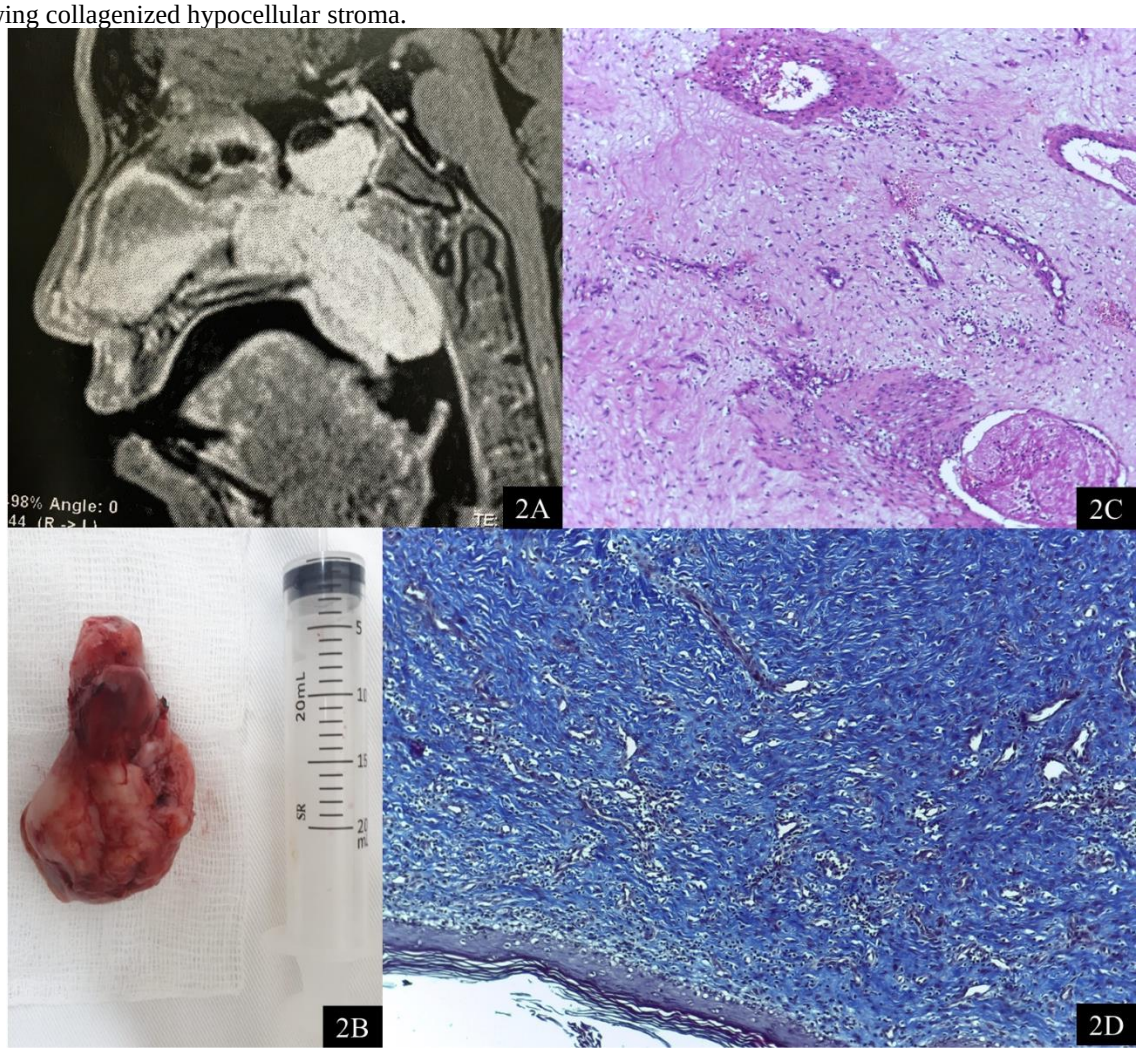
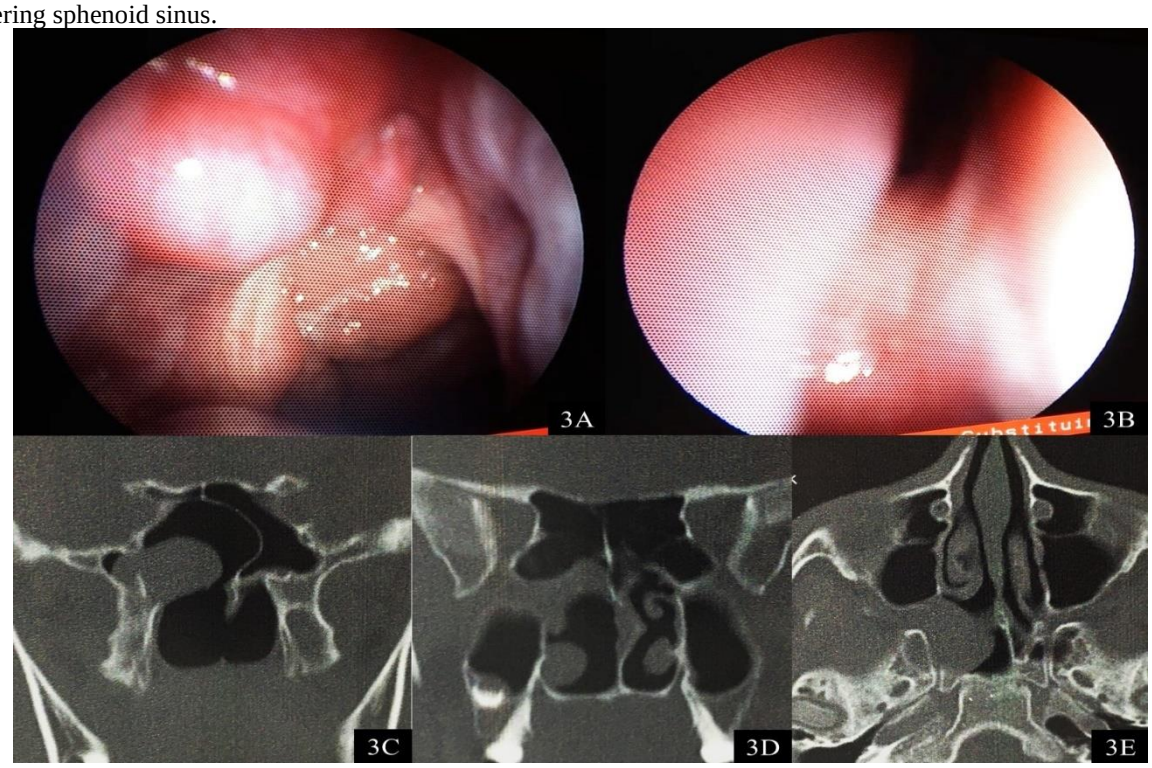


Figure 3 – Nasopharyngeal angiofibroma. (3A-3B) Nasal endoscopy showing non-friable exophytic lesion with hyperemic mucous cover, posterior to right middle turbinate, emerging from sphenoid rostrum on the right and obstruction of superior nasal meatus without visualization of superior nasal concha. Post-operative CT scan of the paranasal sinuses in bone window: (3C) coronal section showing soft tissue density lesion entering the right sphenoid sinus; (3D) coronal section with the same lesion also affecting right maxillary sinus and invading inferior orbital fissure; (3E) axial section showing enlargement of pterygopalatine fossa and sphenopalatine foramen by the lesion, entering sphenoid sinus.



Case 2

A twelve-year-old male patient referenced to our hospital with history of persistent nasal obstruction of the RNF for 12 months, associated with massive ipsilateral nasal bleeding, requiring hospitalization with need for blood transfusion. The patient also referred pharyngeal discomfort, dysphagia, and ipsilateral hearing loss as associated symptoms. Rhinoscopy showed a fibrous, violaceous, and erythematous lesion, surrounded by hematic crusts, occupying RNF. Oroscopy demonstrated a lesion of fibrous aspect, projecting from soft palate into the oropharynx. Nasal endoscopy exam showed a friable lesion occupying the entire extension of common meatus with externalization from RNF. CT scan of nose and paranasal sinuses showed expansive process arising from right sphenoid sinus, obliterating nasal septum and RNF, protruding to the cavum through right choana, obstructing right maxillary sinus, ethmoidal cells, and frontal sinus, also obliterating the ipsilateral ostiomeatal complex and sphenoethmoidal recess. No evidence of abnormal findings on pterygopalatine fossa (Figure 4).

Concluding preoperative step, the primary diagnostic hypothesis was sphenocoanal polyp, with inverted papilloma as differential diagnosis.

The lesion was submitted to endoscopic excision, with massive bleeding and need for stabilization in intensive care unit with transfusion of 01 unit of packed red blood cells. The patient was discharged after 04 days and went through postoperative period without complaints or further bleeding. Histopathological exam showed a well-defined lesion in some areas by a thin squamous epithelium with its interior consisting predominantly of intertwined arterioles and capillaries without epithelial atypia and a typical mature fibroblastic tissue, thus suggesting an angiofibroma (Figure 5).

After one year of post-operative follow-up, new CT showed no signs of primary or recurrent lesions at sinonasal tract. The final diagnosis proposed for the case was ENPA originating from right sphenoid sinus.

Figure 4 – A case of extranasopharyngeal angiofibroma. Pre-operative CT scan of nose and paranasal sinuses: (4A and 4B) coronal section showing expansive soft tissue density mass arising from right sphenoid sinus occupying maxillary sinus spaces and enlarging ipsilateral ostiomeatal complex; (4C - 4D) axial and (4E) sagittal sections showing the lesion obliterating right nasal septum and right nasal cavity, and protruding to cavum through right choana. Note thickening of right maxillary sinus walls, erosion of its medial wall, and preservation of anatomic limits of pterygopalatine fossa.

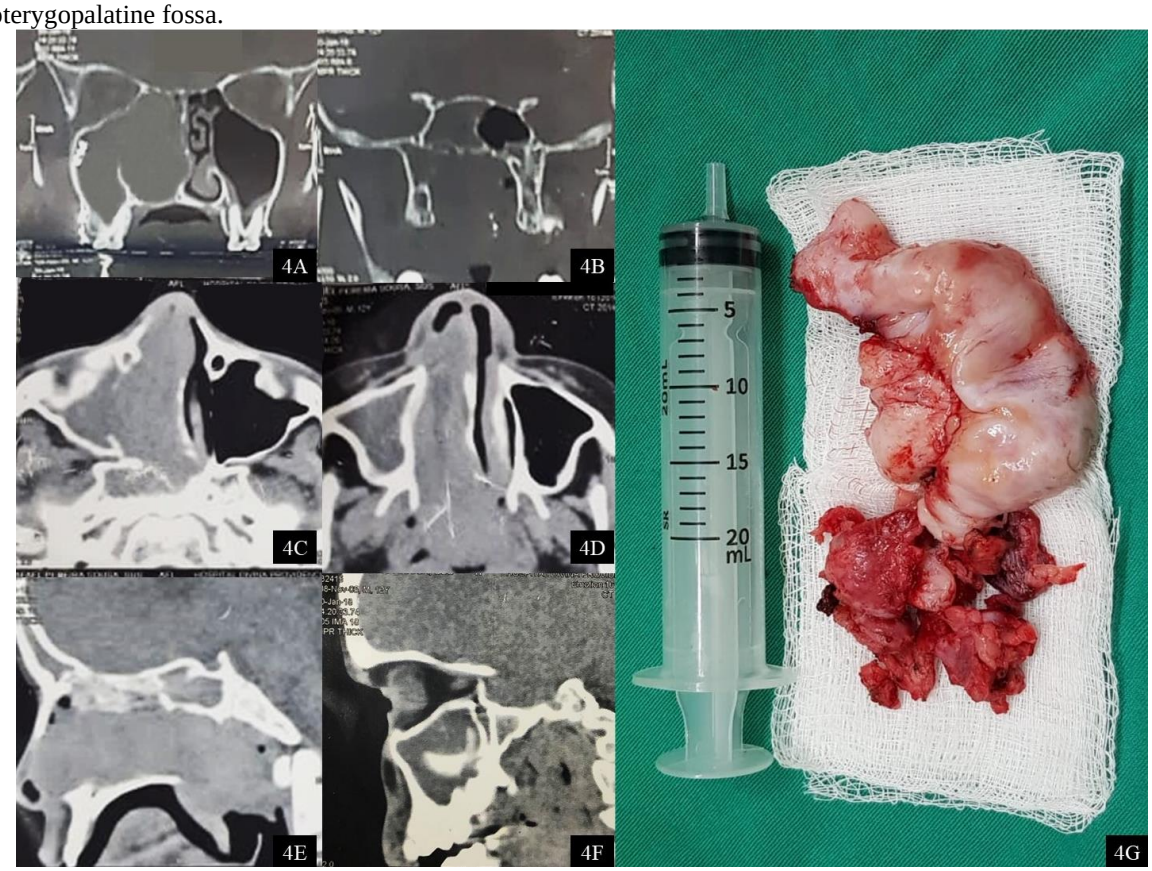
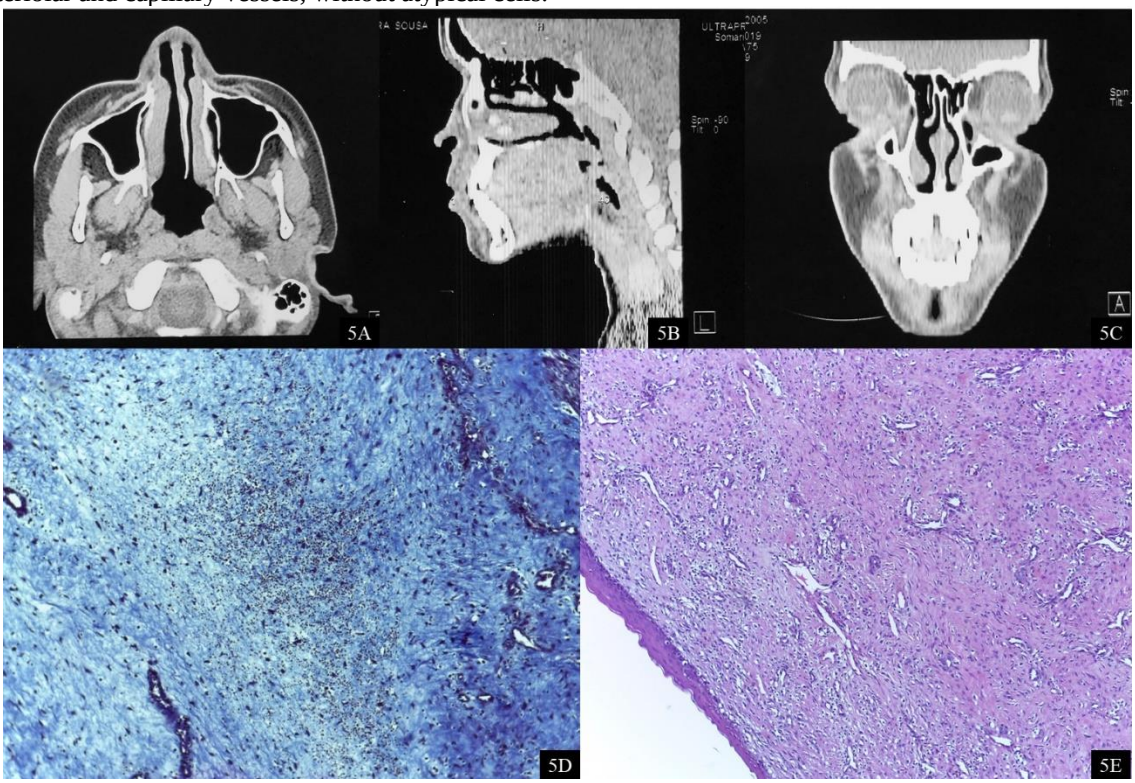


Figure 5 - A case of extranasopharyngeal angiofibroma. Pre-operative CT scan of nose and paranasal sinuses: (5A) axial (5B) sagittal and (5C) coronal sections showing no tumors at sinonasal topography and normal anatomical aspects of the pterygopalatine fossa. Pathological features: (5D) Trichrome stain section showing fibrous stroma; (5E) H&E stain section showing non-keratinized squamous epithelium delimiting the lesion, which consists predominantly of arteriolar and capillary vessels, without atypical cells.



3 DISCUSSION

JNA are rare benign, but locally aggressive, high vascularized tumors, which can lead to spontaneous bleeding or massive hemorrhage. The cases happen almost exclusively in male adolescents, probably due to androngen dependence, and account for less than 1% of all head and neck neoplasms. Its most common symptoms are nasal obstruction, recurrent epistaxis, facial swelling and hyposmia. Our cases showed these symptoms. It can affect adjacent soft tissue, cartilage and bone and may generate complications due to invasion of orbit and cranial base[10–12].

Clinic presentation of the patient in case 1 was highly suggestive of JNA, in addition, his age and gender agree with the epidemiology described in the literature. However, when JNA originates from outside the nasopharynx it is denominated extranasopharyngeal angiofibroma (ENPA) whose presentation is more common in young adults and females and can originate from various sites, although its most often one is the maxillary sinus[2,6]. These tumors are less vascularized and less aggressive than JNA[13]. The image findings of both cases turned a more complicated diagnosis conclusion, capable to confuse even experienced surgeons. Surgical

excision was encouraged during preoperative period due to absence of invasion to orbit or pterygopalatine fossa, and also no neurocranial complications.

ENPA can produce a variety of symptoms depending on its location on the sinonasal tract. Patient from case 2 reported persistent unilateral nasal obstruction and oropharyngeal discomfort due to lesion expansion to oropharynx. The massive epistaxis episode appeared due to its expansion to nasal cavity. In view of its imaging and intraoperative findings, the diagnosis of ENPA of sphenoid sinus was proposed, which are extremely rare, with very few cases reported on literature[14].

Current literature proposes that surgical excision of the mass is the treatment of choice for JNA and ENPA with low post-operative recurrence rates[15]. However, surgical approach to the lesion must depend the surgeon experience as well as extent of the lesion and clinical condition of the patient.

In both cases we performed endoscopic resection, because it is a lesser invasive procedure with lower morbidity than open approaches and also enables the radical removal[15,16].

Our cases bring to medical literature the difference between a JNA and an ENPA cases, with crucial surveillance in late postoperative control.

4 CONCLUSION

Angiofibroma should always be considered as differential diagnosis in cases of tumors of the nasal cavity and paranasal sinuses. In some cases, there will be unusual behaviors of the tumors, representing a diagnostic and therapeutic challenge to rhinologists. In case of unusual sites of origin of these neoplasms, ENPA must also be considered due to possible severe bleeding during the intraoperative period with need for blood transfusion.

Since ENPA is a less vascularized tumor, with different epidemiology from JNA, that originates from a variety of sites, it should be considered as a separated clinical entity.

DECLARATIONS

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REFERENCES

1. Sarpa, J. R. & Novelly, N. J. Extranasopharyngeal Angiofibroma. *Otolaryngol. Neck Surg.* **1**01, 693–697 (1989).

2. Huang, R. Y., Damrose, E. J., Blackwell, K. E., Cohen, A. N. & Calcaterra, T. C. Extranasopharyngeal angiofibroma. *Int. J. Pediatr. Otorhinolaryngol.* 56, 59–64 (2000).

3. Yenigun, A., Aksoy, F., Vural, O. & Ozturan, O. Angiofibroma Localized in the Sphenoid Sinus. *Case Rep. Otolaryngol.* 2017, 4287293 (2017).

4. Goud, S., Swapna, T., Bindu3, H. & Shylaja, S. R. G. Extra Nasopharyngeal Angiofibroma Simulating a Mucocele: A New Location for the Rare Entity. *J. Clin. DIAGNOSTIC Res.* 11, ZD28 (2017).

5. Windfuhr, J. P. & Vent, J. Extranasopharyngeal angiofibroma revisited. *Clin. Otolaryngol.* 43, 199–222 (2018).

6. Windfuhr, J. P. & Remmert, S. Extranasopharyngeal angiofibroma: Etiology, incidence and management. *Acta Oto-Laryngologica* vol. 124 880–889 (2004).

7. Lerra, S., Nazir, T., Khan, N., Qadri, M. S. & Dar, N. H. A case of extranasopharyngeal angiofibroma of the ethmoid sinus: A distinct clinical entity at an unusual site. *Ear, Nose Throat J.* 91, (2012).

8. Kesarwani, A. Sphenochoanal Polyp: A Rare Entity with Review of Literature. *J. Otolaryngol. Res.* 8, (2017).

9. Al Okbi, H., Jain, R. & Al-Abri, R. Sphenochoanal polyp - Surgical review. *Int. J. Pediatr. Otorhinolaryngol. Extra* 5, 99–101 (2010).

10. S, A., C, N., S, P. & SS, D. Juvenile Nasopharyngeal Angiofibroma: A Case Report. *J. Clin. DIAGNOSTIC Res.* 11, (2017).

11. Valkov, A., Duhlenski, B. & Tsvetanova, K. Modern Approach to Juvenile Nasopharyngeal Angiofibroma: A Literature Review. *Int. J. Sci. Res.* 5, 2319–7064 (2013).

12. Liu, Z. *et al.* Hormonal receptors and vascular endothelial growth factor in juvenile nasopharyngeal angiofibroma: Immunohistochemical and tissue microarray analysis. *Acta Otolaryngol.* 135, 51–57 (2015).

13. Szymańska, A., Szymański, M., Morshed, K., Czekajska-Chehab, E. & Szczerbo-Trojanowska, M. Extranasopharyngeal angiofibroma: Clinical and radiological presentation. *Eur. Arch. Oto-Rhino-Laryngology* 270, 655–660 (2013).

14. Karthikeyan, P. Extra nasopharyngeal angiofibroma of the sphenoid sinus. 6, 8–13 (2014).

15. Singhal, S. K., Gupta, N., Verma, H., Dass, A. & Kaur, A. Extranasopharyngeal angiofibroma: Report of two cases. *Egypt. J. Ear, Nose, Throat Allied Sci.* 15, 73–76 (2014).

16. Lund, V. J. *et al.* European position paper on endoscopic management of tumours of the nose, paranasal sinuses and skull base. *Rhinol. Suppl.* 22, 1–143 (2010).