

## CLINICAL CASE

# Unusual case of nasopharyngeal angiofibroma in adult male patient

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## ABSTRACT

The juvenile nasopharyngeal angiofibroma is a benign slow-growing tumor with vascular component which affects males in their early puberty and adolescence, and in very few cases men over 25 years of age. The authors describe a case of a nasopharyngeal angiofibroma in a 56 year-old male patient, confirmed by histological and immunohistochemical examination. The CT scan and the selective angiography made before surgery revealed the characteristics of a hypervascular mass occupying the nasopharynx and the posterior part of the nasal cavity. The tumor was successfully removed by lateronasal rhinotomy. The follow-up shows no signs of clinical, endoscopic and imagistic recurrence three years after surgery.

**KEYWORDS:** nasopharyngeal angiofibroma, adult patient, lateronasal rhinotomy, selective angiography

## INTRODUCTION

The juvenile nasopharyngeal angiofibroma is a benign, slow-growing tumor with vascular component, which affects mostly males between 14 and 25 years of age. In very few cases it has been reported in men over 25 years old or females<sup>1</sup>. The nasopharyngeal angiofibroma is a rare tumor representing about 0.5% of all head and neck tumors<sup>2</sup>. Although it is a benign tumor, it has a high morbidity and mortality due to its local invasiveness<sup>3</sup>. Having a slow-growing rate, the patient usually complains about progressive and persistent nasal obstruction, often accompanied by recurrent epistaxis. Diagnosis is based upon patient's history, physical examination and radiological findings. The complete surgical excision represents the gold standard treatment for this kind of tumors.

## CASE REPORT

A 56 year-old male patient presented with progressive and persistent nasal obstruction for the past 12 months, accompanied by intermittent mucous rhinorrhea and headache. There was no episode of nasal bleeding or any other pathology in patient's medical history.

The anterior rhinoscopy and nasal endoscopy revealed the presence of a white-pink, non-ulcerated tumor, apparently descending from the ceiling of the nasal cavity and occupying the posterior half of the right nasal fossa and one third of the left nasal fossa (causing

the obstruction of choanal orifices), the nasopharynx and descending till the margins of the soft palate.

Contrast-enhanced cranio-facial computed tomography (CT) showed an intensely enhancing mass occupying the nasopharynx, both nasal cavities and descending to the margins of the soft palate. The tumor caused the osteolysis of the posterior part of the nasal septum (Figure 1). There were no signs of extension to the sphenoid sinus or invasion of the pterygomaxillary or infratemporal fossa.

Before surgery, a carotid angiography was performed revealing a hypervascular mass with the main blood supply from the anterior internal maxillary artery, branch of the right external carotid artery (Figure 2). Unfortunately, the selective embolization of the main arterial branches during angiography was not possible. We decided that the first step of the surgical procedure must be external right carotid artery ligation. This procedure was performed in order to minimize hemorrhagic complications. Afterwards, the patient underwent open surgery by lateronasal rhinotomy. This classical approach was used because of the tumor's dimensions (11/5/7 cm) which made intranasal endoscopic resection almost impossible. Using this surgery technique, the tumor was completely removed, in one piece (Figure 3), without bleeding or other complications during surgery or in the following days.

The histological examination of the tumor revealed a highly vascularized, mature, fibrous tissue characteristic for angiofibroma. Also, the immunohistochemistry (IHC) tests confirmed the histological diagnosis. The CD31 and CD 34 antigens were identified in

blood vessels tissue. The CD 34 antigen was also found in the tumor's stromal tissue (Figure 4).

The patient was discharged 9 days after surgery. The follow-up was made at one month, three months and six months after surgery and then once a year. The patient, still under surveillance, shows no signs of clinical, endoscopic and imagistic recurrence.

## DISCUSSIONS

Although extremely rare, the nasopharyngeal angiofibroma can occur in elderly adults. Lukomski

et al. presented in their study only 36 cases during a period of 50 years<sup>4</sup> and this statistic confirms the low prevalence of this disease.

The pathogenesis of the juvenile nasopharyngeal angiofibroma is still unknown. Because of its high frequency in teenage males, Andrade explained this selectivity by intranuclear accumulation of androgen receptors and by a high number of growth factors (VEGF - endothelial growth factor, TGF $\beta$  - transforming growth factor beta)<sup>5</sup>. Moreover, there are some very recent theories which state that the nasopharyngeal angiofibroma could be a vascular malformation due to incomplete regression of the first branchial artery<sup>6</sup>.

This tumor usually originates from the region of sphenopalatine foramen, enlarges to fill the postnasal space and frequently extends to the sphenoid sinus, pterygo-maxillary and infratemporal fossae, orbit or even to the middle cranial fossa<sup>7</sup>. The radiologic examinations remain the most important diagnostic tools, because they allow the correct evaluation of tumoral extension and help the surgeon to choose the most correct surgical approach, in order to remove the tumor<sup>6,8,9</sup>.

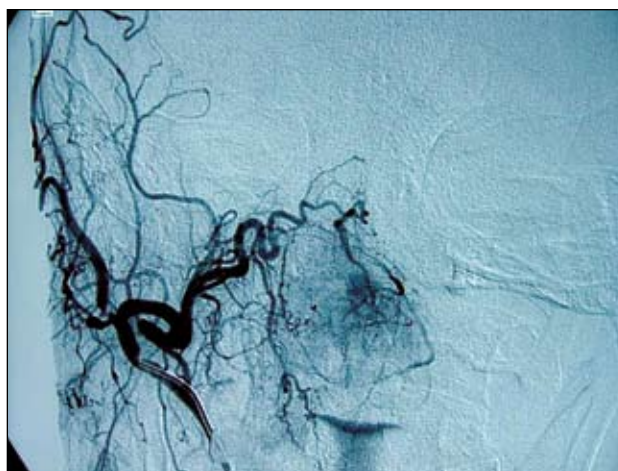
In order to be able to predict the outcome of the tumor, it is important to know its extent. For that purpose, a staging system must be used. There are many staging criteria developed when evaluating a juvenile nasopharyngeal angiofibroma, like Fisch, Radkowski or Andrews. The Radkowski criteria, based on clinical and radiological criteria, seems to be the most used in modern literature<sup>3</sup> and by us also. According to the latter, our case was included in stage IB.

Regardless of the stage or chosen surgical approach, the patient must take a selective angiography on internal and external carotid arteries, in order to evaluate the main arterial branches which ensure the tumor's blood supply. In some cases, the angiography can be continued with the embolization of the main arterial branches<sup>9,10,11</sup>. Unfortunately, the embolization could not be performed in our case.

Concerning the surgical approach and considering that most of the patients are still in their growing period, there has been a marked shift towards less invasive surgery. The endoscopic approach is reported having extremely good



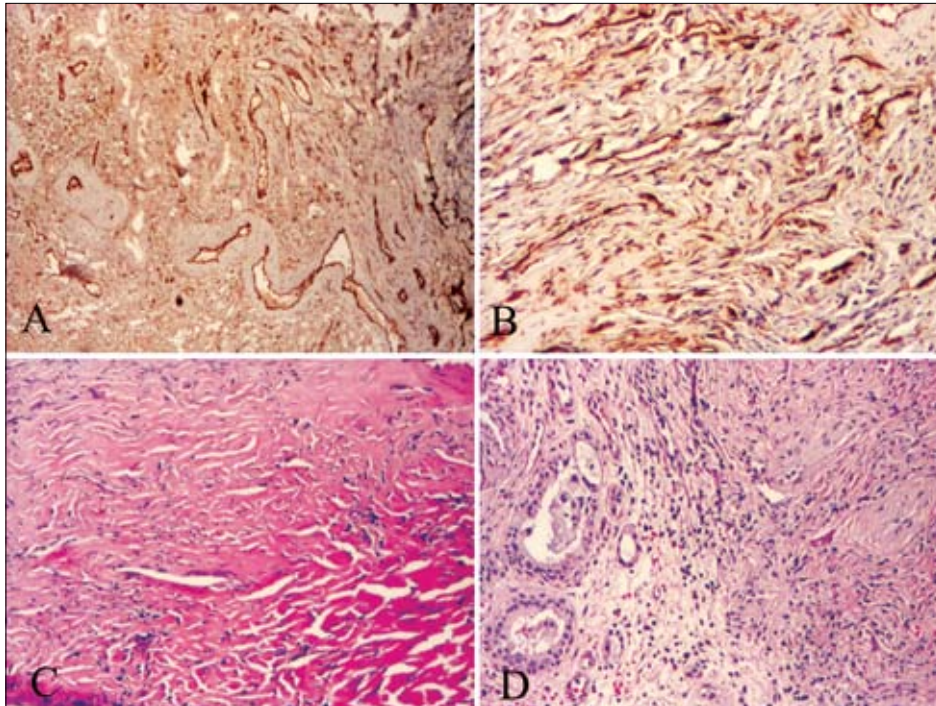
**Figure 1** CT scan revealing the tumor's extension in the nasal cavity and osteolysis of the posterior part of the nasal septum (the CT scan was performed while the patient had nasal packing, the arrows revealing the delimitation between nasal packing and tumor)



**Figure 2** Selective angiography on right external carotid artery → a hypervascular mass with the main blood supply from the anterior internal maxillary artery, branch of the right external carotid artery



**Figure 3** Macroscopic view of the surgical specimen



**Figure 4** Histological findings: (A) Immunostaining for CD 31 antigen found positive in blood vessels 4X; (B) CD 34 staining positive in blood vessels and stromal tissue 10X; (C) Vascular gaps and areas of diffuse sclerohyalinization HE 4X; (D) Marginal mucosa with glandular epithelium and angiofibromatous proliferation HE 4X

results and low recurrence rates, even if the tumor has minimal intracranial invasion. In certain selected cases, with very small tumors, Gamma knife has also been used and the reported results have been extremely encouraging<sup>12</sup>.

External approach, like lateral rhinotomy or midfacial degloving, is used because it ensures a good exposure of the tumor and its extensions, but, especially in children, it may cause undesirably facial growth retardation<sup>8,9</sup>. If the tumor hasn't been completely removed, it has a significant tendency to recur, new surgical intervention being sometimes necessary<sup>8,9,10,13,14</sup>.

## CONCLUSIONS

Nasopharyngeal angiofibroma is a benign but locally aggressive tumor and appears extremely rare in adults. Imaging studies are the most important tools in positive and differential diagnosis. Interventional radiology can help in the attempt to cure this tumor. It is partially involved in the process by providing the means of minimizing bleeding through selective embolization and it can also serve as curing treatment by the use of Gamma knife surgery. However, the surgery remains, up to date, the gold standard treatment for nasopharyngeal angiofibroma. It can be performed by endoscopic or external approach, considering individual anatomy, pathology, blood supply, location and size of the tumor. The surgery allows complete tumor resection and prevents recurrences or other complications.

## REFERENCES

- Gallard A.L., Anastacio V.M., Piatto V.B., Maniglia J.V., Molina F.D., A seven-year experience with juvenile nasopharyngeal angiofibroma. *Braz J Otorhinolaryngol*, 2010;76(2):245-50.
- Paris J., Guelfucci B., Moulin G., Zanaret H., Triglia M., Diagnosis and treatment of juvenile nasopharyngeal angiofibroma. *Eur Arch Otorhinolaryngol*, 2001;258:120-4.
- Zimmermann E., Selomke I., Gavazzoni F.B., Pereira R.G., Machado S., Tanamati T.K., Endoscopic surgery of nasopharyngeal angiofibroma. *Intl Arch Otorhinolaryngol*, 2010;14(2):206-211.
- Lukomski M., Danilewicz M., Pajor A., Juvenile angiofibroma in adults. *Otolaryngol Pol*, 2008;62(1):20-6.
- Andrade N.A., Andrade J.S.C., Silva P.D.M. et al., Angiofibroma Nasofaríngeo: Revisão dos aspectos genéticos e moleculares. *Arq Int ORI*, 2008;12(3):442-449.
- Eloy Ph., Watelet J.B., Hatert A.S. et al., Endonasal endoscopic resection of juvenile nasopharyngeal angiofibroma. *Rhinology*, 2007;45:24-30.
- Liang J, Zixiang Y, Ping L. (2000) The nature of juvenile nasopharyngeal angiofibroma. *Otolaryngol Head Neck Surg* 123:475-81.
- Önerci M., Ögretmenoglu O., Yücel T., Juvenile nasopharyngeal angiofibroma: a revised staging system. *Rhinology*, 2006;44:39-45.
- Karci B., Öner K., Günhan Ö. et al., Nasomaxillary osteotomy in lesions of the central compartment of the middle cranial base. *Rhinology*, 2001;39:160-165.
- Sennes L.U., Butugan O., Sanchez T.G. et al., Juvenile nasopharyngeal angiofibroma: the routes of invasion. *Rhinology*, 2003;41:235-240.
- Lim I.R., Pang Y.T., Soh K., Juvenile Angiofibroma: Case Report and the Role of Endoscopic Resection. *Singapore Med J*, 2002;43(4):208-210.
- Park C.K., Kim D.G., Paek S.H. et al., Recurrent Juvenile Nasopharyngeal Angiofibroma Treated with Gamma Knife Surgery. *J Korean Med Sci*, 2006;21:773-7.
- Panesar J., Vadgama B., Rogers G. et al., Juvenile angiofibroma of the maxillary sinus. *Rhinology*, 2004;42:171-174.
- Schick B., Rahman A.E., Tahan E. et al., Experiences with endonasal surgery in angiofibroma. *Rhinology*, 1999;37:80-85.