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Aggressive Tumor and Challenging Location: A Case Report of Nasopharyngeal Fibroma

Y. Bouktib[1*], M. Raboua[1], A. Elhajjami[1], B. Boutakiout[1], M. Idrissi Ouali[1], N. Cherif Idrissi Ganouni[1]

[1]Radiology Department, ER-RAZI Hospital, CHU Mohammed VI University Cadi Ayad Marrakech, Morocco

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***Corresponding author:** Y. Bouktib
Radiology Department of Ibn Tofail Hospital, CHU Mohammed VI, Marrakech, Faculty of Medicine and Pharmacy, Cadi Ayyad University, Marrakech, Morocco

Abstract **Case Report**

Nasopharyngeal angiofibroma refers to a locally aggressive benign vascular tumor composed of vasogenic and myofibroblastic elements, representing between 0.05% and 0.5% of all head and neck tumors. Here, we report a case of a 21-year-old patient with no particular personal or familial pathological history, who presented with recurrent epistaxis and anterior rhinorrhea and headaches for 1 year. Nasofibroscopy revealed a mass filling the entire left nasal cavity, streaked with blood. A face MRI revealed a highly vascularized and lytic nasopharyngeal tumor infiltrating the deep left lateral facial spaces with the beginning of endocranial infiltration, supplied by the homolateral maxillary artery, initially suggestive of a nasopharyngeal fibroma. Preoperative arteriography showed a voluminous vascular blush at the arterial time to the nasopharyngeal projection with branches arising from the left external and internal carotid arteries and multiple arterial shunts (between the left internal and maxillary carotid arteries), thus contradicting embolization. Early diagnosis is important as it is associated with a high risk of morbidity, but advances in imaging and surgical treatment methods have modified the sites associated with high morbidity risk.

Keywords: Nasopharyngeal angiofibroma, magnetic resonance imaging, Angiography.

INTRODUCTION

Nasopharyngeal fibroma is a rare, richly vascularized benign tumor affecting almost exclusively adolescent males. Its severity is linked to its hemorrhagic potential and its local aggressiveness, representing between 0.05% and 0.5% of all head and neck tumors. His management is essentially surgical. It has largely benefited from the development of endonasal surgery and interventional arteriography. We report a case of a young patient who presented with recurrent epistaxis and anterior rhinorrhea and headaches for 1 year revealing a nasopharyngeal Fibroma

CLINICAL CASE

A 21-year-old patient with no significant personal or family medical history has been experiencing recurrent nosebleeds, anterior rhinorrhea, and headaches for the past year. The patient has been in good general health and has not experienced any hearing loss, visual impairment, or other signs. Examination revealed swelling of the right jugular vein. Nasal fibroscopy revealed a mass filling the entire left nasal cavity, which appeared to be streaked with blood.

CT scan revealed a large, poorly defined tissue mass in the left maxillary sinus, which showed intense enhancement after contrast injection. The tumor had invaded the papyraceous lamina, resulting in bone lysis of the maxillary sinus walls, and extended into the left nasal cavity, sphenoidal sinus, and left temporal fossa. The mass enlarged the left superior orbital fissure without intra-orbital involvement.

MRI showed a highly vascularized and lytic nasoendopharyngeal tumor mass with early endocranial infiltration (Figure 1). It had polylobed contours and showed intense and heterogeneous enhancement after contrast injection. The mass extended from the left maxillary sinus to the left sphenoid sinus and through the left foramen round to the endocranium with meningeal contrast opposite the homolateral temporal lobe and invades inferiorly to the homalateral infratemporal fossa. It enlarges the superior orbital fissure without evidence of intra-orbital extension or

invasion of sella. The process was mainly supplied by the superior maxillary artery without involvement of the cavernous sinus or left internal carotid artery. These findings suggest a nasopharyngeal fibroma.

Preoperative arteriography was performed, showing a large vascular blush in the arterial phase with projection to the nasopharynx, with the presence of branches from the left external and internal carotid arteries and multiple arterial shunts (between the left internal carotid and maxillary arteries), contradicting embolization.

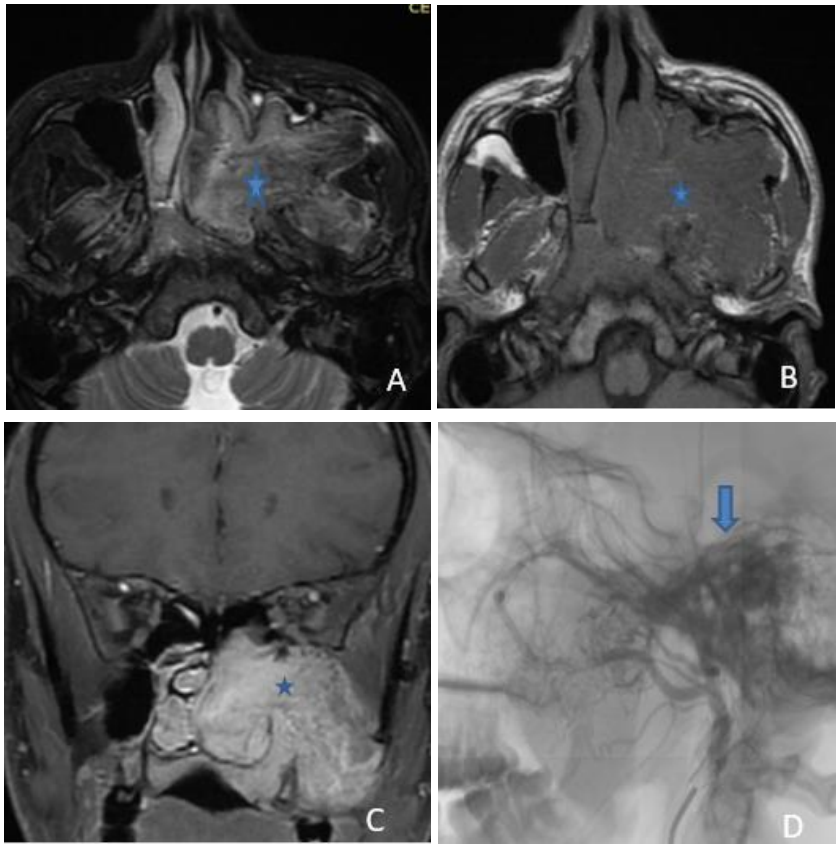

Figure 1: Axial T2 (A) and T2 (B) weighted magnetic resonance and coronal contrast enhanced T1 Fat Sat. Highly vascularized and lytic nasoendopharyngeal tumor (Asterix) with polylobed contours and intense and heterogeneous enhancement after contrast injection (C). Arteriography (D) showing a large vascular blush in the arterial phase (arrow) with projection to the nasopharynx

DISCUSSION

Nasopharyngeal fibroma is a rare vascular tumor that is histologically benign but locally aggressive. It almost exclusively develops in male adolescents between the ages of 14 and 18 [1, 2]. The youngest reported patient in the literature was 7 years old [3]. NP fibroma can also occur in female patients, such as a 71-year-old woman, or in adults, such as a 79-year-old patient [4].

The etiopathogenesis of NP fibroma is not yet well understood. Several theories have been proposed but are controversial [5]. The discovery of the condition in 5% of female cases, as well as in a pregnant woman (Péloquin *et al*., 1997) [3], has ruled out the long-held hypothesis of hormone dependence [1, 2, 6]. It is now accepted that NP fibroma or angiofibroma of the pharynx implants itself on the periphery of the spheno-palatine foramen, more specifically on its posterosuperior border [7]. However, 47 extrapharyngeal localizations have been reported, with maxillary sinus involvement being the most common [8, 9].

From its point of implantation, NP fibroma extends into the underlying mucosa, erodes bone structures, and infiltrates surrounding tissues through several cracks and foramina. Its modalities of extension are very polymorphic and escape any systematization. This local aggressiveness can affect functional and vital prognoses [7].

Thus, the origin, pathogenesis, and natural history of nasopharyngeal fibroma remain hypothetical. The positive diagnosis of this entity is clinical in the majority of cases. Epistaxis is usually the telltale sign, as we found in our patient. Rhinologic warning signs may initiate the clinical picture (nasal obstruction, rhinorrhea, etc.) [7, 3]. Nasal endoscopy can visualise

the purple-red vascular tumour, which bleeds on contact and more or less fills the nasal cavity and adjacent structures, as in our case [8]. The right jugal swelling observed in our patient's examination is indicative of tumor progression. Signs of skull base invasion should be sought. The absence of associated cervical lymphadenopathy should draw attention and suggest the diagnosis of NP fibroma, ruling out differential diagnoses. Biopsy may be considered in certain circumstances, such as in a woman or an elderly patient [4]. This biopsy must be taken with caution due to the risk of hemorrhage.

In our case, imaging of the facial mass established the diagnosis. NP fibroma is the only hypervascularized tumor, strongly enhanced by contrast injection, that develops around the spheno-palatine foramen. This imaging also allows for a locoregional extension assessment: CT scan allows for a study of bone structures, while MRI explores soft tissues. In our case, the MRI showed an infratemporal fossa and endocranial invasion with meningeal contrast opposite the homolateral temporal lobe without involvement of the cavernous sinus, optic nerve or sella.

Angiography visualizes the feeding pediclesn, shows the tumor blush and can guide potential embolization [2].

Elective angiography can identify the feeding vessels and allow for preoperative embolization to control the blood flow to the tumor. This procedure can show the size and location of the lesion as well as the size and location of the feeding vessels [10]. Vascularization typically arises from the external carotid branch of the maxillary artery, with background vascularization coming from blood vessels in the ascending pharyngeal artery and internal carotid artery [11]. In our case, these findings were also observed.

After radiological assessment, the tumor is classified according to its stage of extension. The Radkowski classification is the most recent (1996) and widely used (table 1). It is better suited for preoperative assessment and therapeutic strategy for these tumors. This corresponds in our case to a classification of IVa. Thus the combination of radiotherapy and chemotherapy was proposed in our case.

Table 1: Fisch classification of JNA based on its extension in CT and MRI

Stage I	The tumor is limited to the sphenopalatine foramen, nasopharynx and nasal cavity without bone destruction
Stage II	The tumor invades the nasal sinuses or the pterygomaxillary fossa with bone destruction
Stage IIIa	The tumor invades the infratemporal fossa or orbit without intracranial involvement
Stage IIIb	The tumor invades the infratemporal fossa or orbit with intracranial and extradural involvement
Stage IVa	The tumor shows intracranial, extradural and/or intradural invasion, without invasion of optic nerve, sella, or cavernous sinus
Stage IVb	The tumor in stage IVa with invasion of optic nerve, sella and/or cavernous sinus

JNA: Juvenile nasopharyngeal angiofibroma; CT: Computed tomography; MRI: Magnetic resonance imaging

CONCLUSION

Nasopharyngeal fibroma is a rare and highly vascular tumor that arises in the nasopharynx. It can be locally invasive and has a high risk of morbidity if left untreated. Early diagnosis through imaging studies, such as CT scan, MRI, and preoperative arteriography, is important for prompt management of the condition. Treatment options include surgical excision, radiation therapy, or a combination of both.

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