Juvenile angiofibroma: A case report

Tiago Lourenço Coelho • Hugo Figueiredo • Ana Beatriz Ramada • Davide Lourenço Marques • Ricardo Caiado • Jorge Migueis • Luís Filipe Silva

ABSTRACT

Juvenile Angiofibroma is a rare pathology, with high associated morbidity.

A 14-year-old male was referred to the otolaryngology consultation due to left nasal obstruction, snoring and epistaxis with 6 months of evolution. The anterior rhinoscopy showed the presence of an expansive formation occupying the entire left nasal cavity. In the examination of the oropharynx, a lobulated mass could be observed, bulging the left soft palate. Contrast-enhanced Computed Tomography of the Peri-Nasal Sinus confirmed the previous findings. Angioembolization of the left external carotid artery was performed with subsequent surgical excision. Histopathology confirmed that it was a juvenile nasopharyngeal angiofibroma (JNA).

There were no complications after surgery. At 6 months of follow-up the patient was asymptomatic and without evidence of recurrence.

The clinical suspicion of this pathology is essential, allowing a timely diagnosis, with a consequent reduction in associated comorbidities.

Keywords: juvenile angiofibroma; epistaxis.

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INTRODUCTION

Juvenile nasopharyngeal angiofibromas (JNAs) are rare benign tumors that almost exclusively affect adolescent boys.[1-4]

Anamnesis highlights episodes of unilateral recurrent epistaxis and ipsilateral nasal obstruction.[2]

These tumors account for < 0.5% of all head and neck tumors.[1-4]

JNAs usually originate in the posterior nasal cavity, near the lateral base of the sphenoid and the superior margins of the sphenopalatine foramen.[2] Angiofibromas can also occur in extranasopharyngeal sites, namely in the nasal septum and inferior turbinate.[5,6]

These tumors are locally invasive and characterized by a major vascular component, with preoperative angiography being an important resource.[7] Invasion of the skull base occurs in 10–20% of patients.[7,8]

JNAs may occur as a component of the familial adenomatous polyposis (FAP) syndrome, with abnormal nuclear localization of beta-catenin in tumor stroma cells.[9]

Several growth factors and receptors appear to have implications for the development of JNA, including vascular endothelial growth factor (VEGF), VEGF receptor[2], transforming growth factor beta 1, and insulin-like growth factor[2,9]

Open or endoscopic surgical excision is the recommended treatment.[2,11,12] Radiotherapy is reserved for invasive and/or recurrent tumors.[10-12] The recurrence rate varies between 6% and 24%.[10]

We describe an adolescent boy who was diagnosed with JNA.

CASE DESCRIPTION

A 14-year-old boy attended the otorhinolaryngology consult with a clinical presentation of left recurrent epistaxis and snoring, associated with pain and ipsilateral facial edema that had persisted for six months. He also reported nausea and vomiting during the night, leading to nocturnal awakening a few months prior. He felt tired after mild exertion and experienced daytime sleepiness.

Although his personal medical history was irrelevant, he had a family (maternal) history of FAP.

An objective otorhinolaryngological examination revealed otitis media and bilateral effusion. Tone audiometry revealed bilateral conductive deafness and a bilateral type B tympanogram (Fig. 1).

Anterior rhinoscopy showed thick anterior rhinorrhea and an expansive formation occupying the left nasal fossa; the

FIGURE 1
Tone audiogram (A) and tympanogram (B)

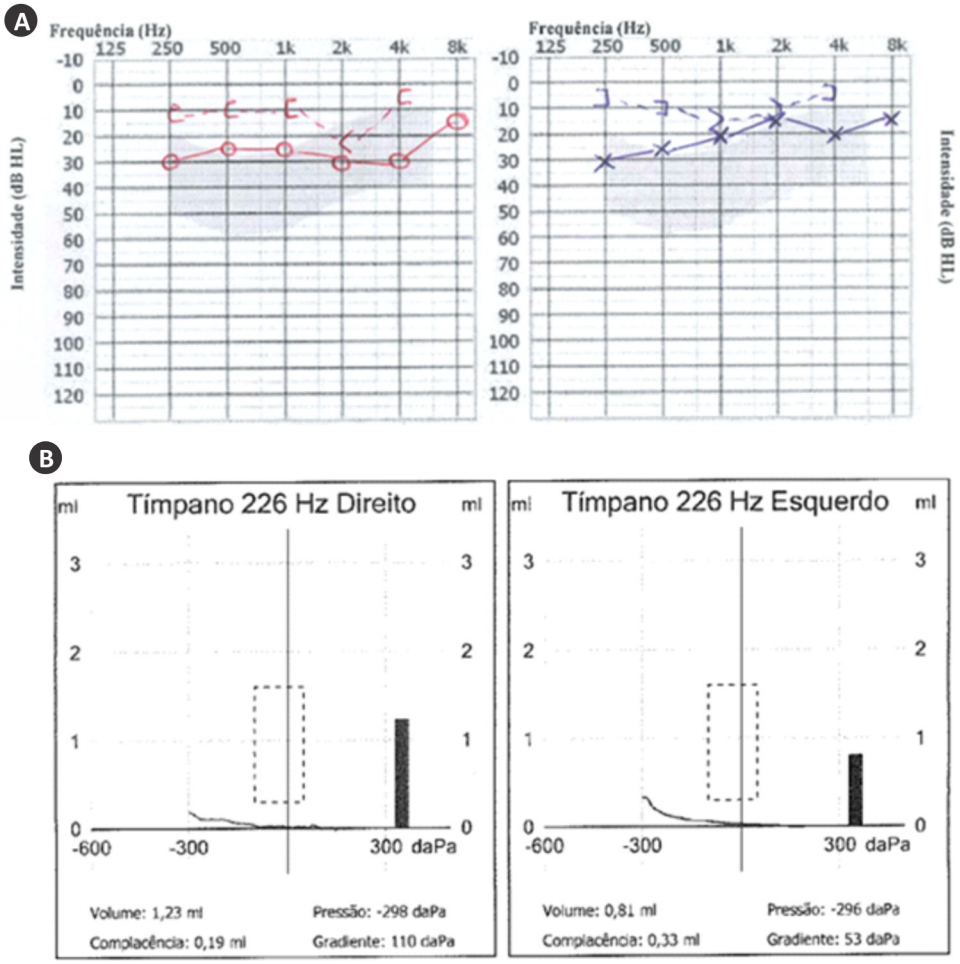
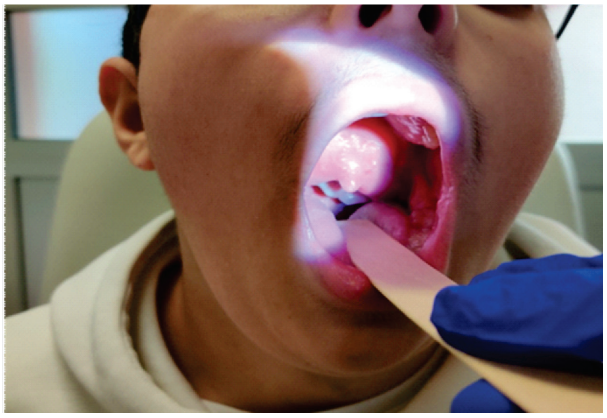


FIGURE 2
Expansive formation occupying the rhinopharynx extending to the oropharynx and bulging left soft palate



right nasal fossa was unaltered. Nasal fiber endoscopy confirmed these findings. Inspection of the oropharynx showed a bulging soft palate, especially of the left, with a lobulated mass in the rhinopharynx with oropharyngeal involvement (Fig.2).

Computed tomography of the nose and perinasal sinuses with contrast enhancement confirmed a large expansive tissue lesion that filled the posterior half of the left nasal fossa, sphenopalatine foramen, and pterygopalatine fossa, with significant extension to the oropharynx, as well as on the left side (Fig. 3).

Magnetic resonance imaging (MRI) of the nose and perinasal sinuses with gadolinium enhancement allowed better characterization of the margins of the tumor mass (largest diameters: 85 × 46 and 55 × 38 mm in the sagittal and axial planes, respectively). Endocranial or intraorbital extension of the mass was excluded (Fig. 4).

The diagnostic hypothesis was stage IIC JNA according to the Radkowski classification based on the clinical and imaging findings (Table 1).

Preoperative tumor angioembolization revealed a large tumor vascular component with supply via the branches of the left external carotid artery (ECA), especially the sphenopalatine artery, with contributions from the middle meningeal, facial, and ascending pharyngeal arteries (Fig. 5).

Open surgery proceeded 24 hours after embolization via a sublabial transmaxillary approach, with a Rouge-Denker incision. The medial wall was removed and the posterior

FIGURE 3
Computed tomography of the nose and perinasal sinuses with contrast enhancement. Axial plane (A), coronal plane (B), and sagittal plane (C).

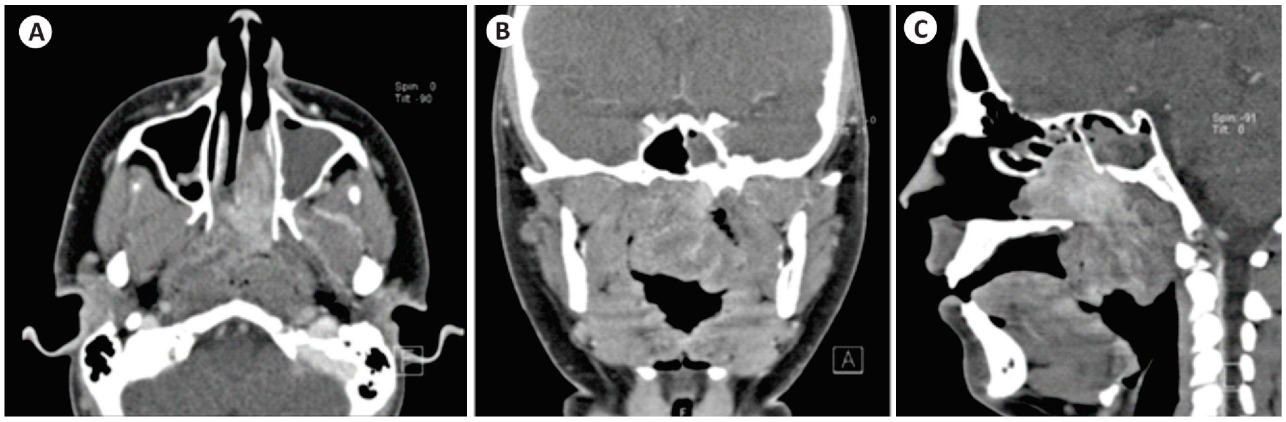


FIGURE 4
Magnetic resonance imaging of the nose and perinasal sinuses. T2 axial plane without gadolinium (A and B) and T1 sagittal plane with gadolinium (C)

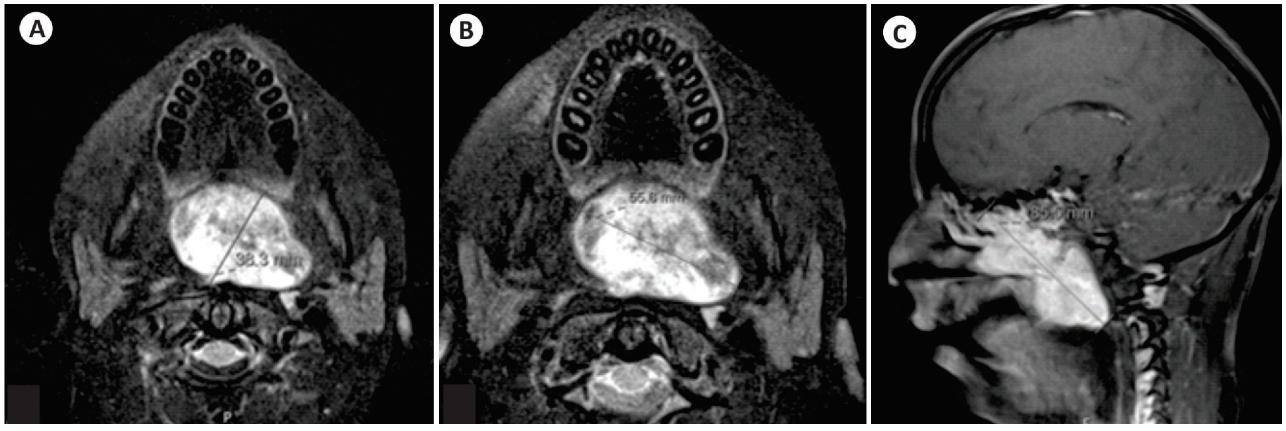


TABLE 1
Stages of juvenile angiofibromas according to Radkowski classification[13]

Stage	Description
IA	Tumor limited to nasal cavity and/or nasopharynx.
IB	Tumor that invades nasal cavity and/or nasopharynx and extends to at least one perinasal sinus.
IIA	Minimum invasion of pterygopalatine fossa.
IIB	Invasion of entire pterygopalatine fossa with or without erosion of the orbital wall
IIC	Extension to infratemporal fossa or posterior extension beyond plates of pterygoid process.
IIIA	Erosion of base of the skull – minimum intracranial extension.
IIIB	Massive intracranial extension with or without invasion of cavernous sinus.

FIGURE 5
Preoperative tumor angioembolization
A – Angiography with extensive blush and supply by branches of the left external carotid artery (ECA); B - After embolization with particles (300 µ), with obvious reduction in tumor capillary blush.

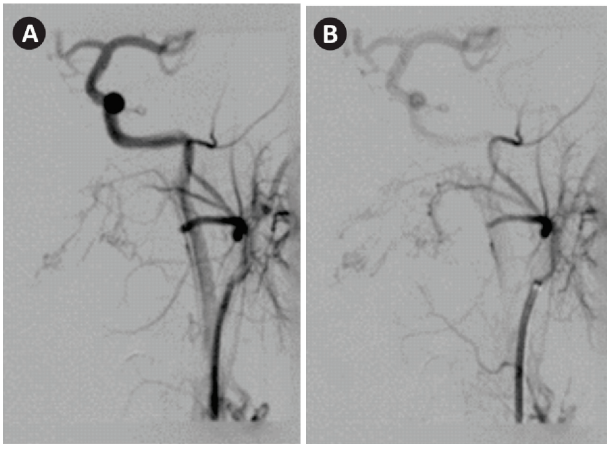


FIGURE 6
Sublabial transmaxillary route with Rouge-Denker incision (A) and tumor lesion extraction (B)

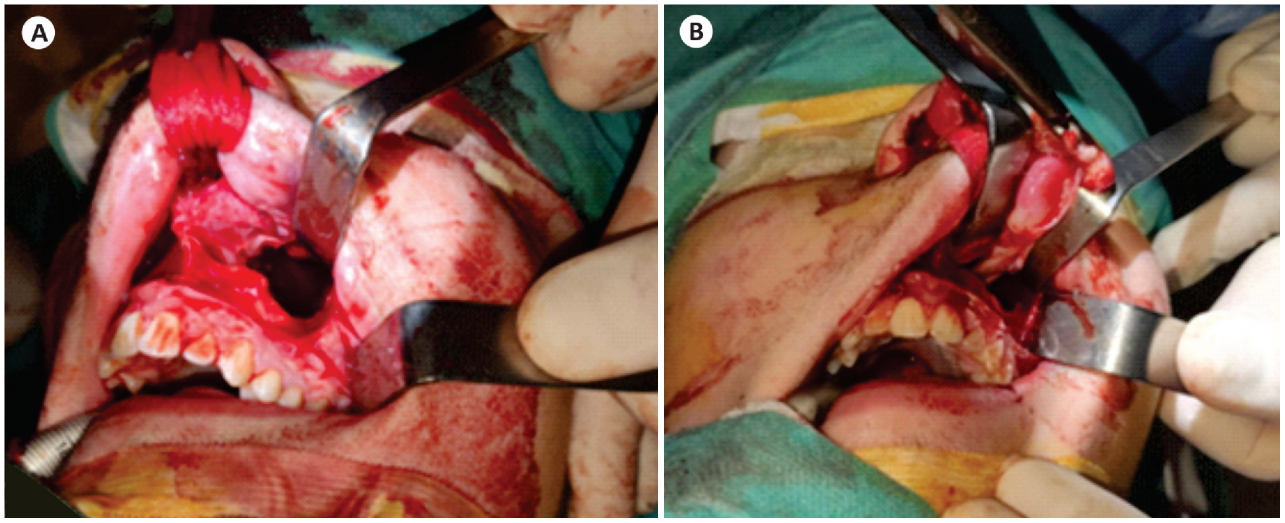
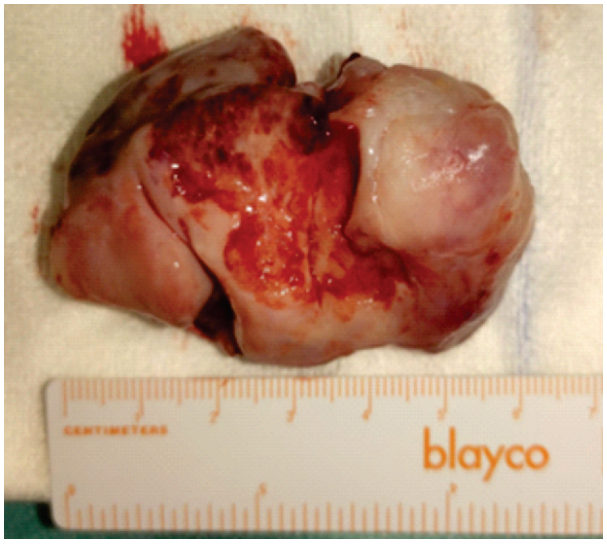


FIGURE 7
Surgical specimen, translucid pink 6.5 × 4.8 × 4 cm-mass with smooth lobulated surface, and fibroelastic consistency.



wall of the left maxillary sinus was opened, associated with turbinectomy of the ipsilateral inferior turbinate (Fig. 6).

The gross pathological findings of the surgical specimen showed a translucid, pink, 6.5 × 4.8 × 4-cm tumor with a smooth, lobulated surface and fibroelastic consistency (Fig. 7). The histological findings were compatible with JNA.

No postoperative complications developed, and the patient was discharged 5 days later. He remains under otorhinolaryngological follow-up and has remained asymptomatic at 6 months of follow-up, without evidence of relapse (Figs. 8 and 9).

He awaits a genetic counseling appointment because of the maternal family history of FAP.

FIGURE 8
Oropharynx on objective examination (A) and rhinopharynx on nasal fiber endoscopy (B), at six months postoperatively

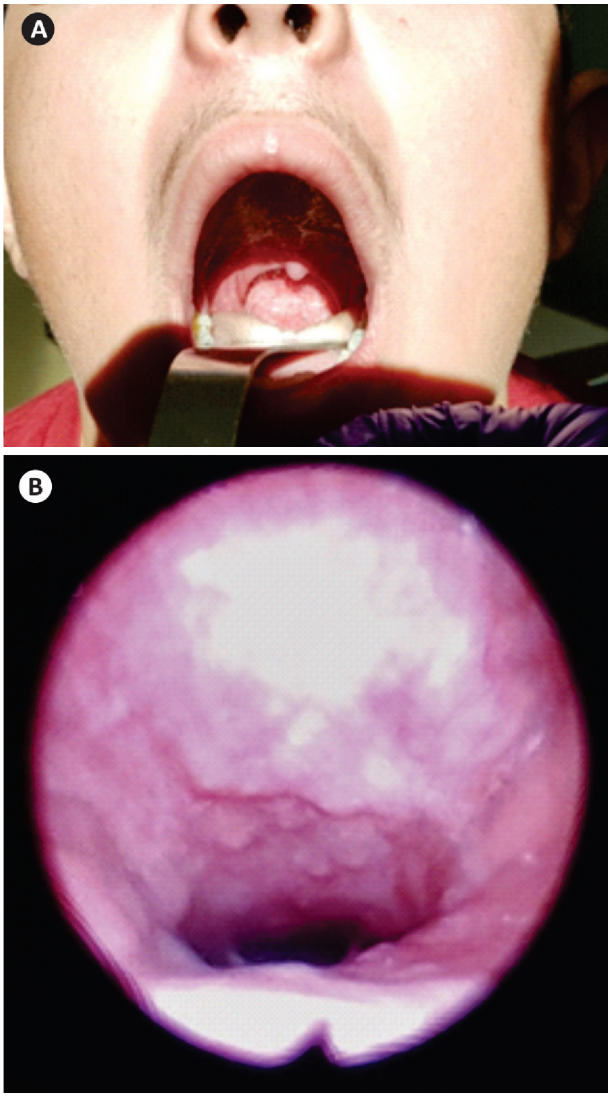
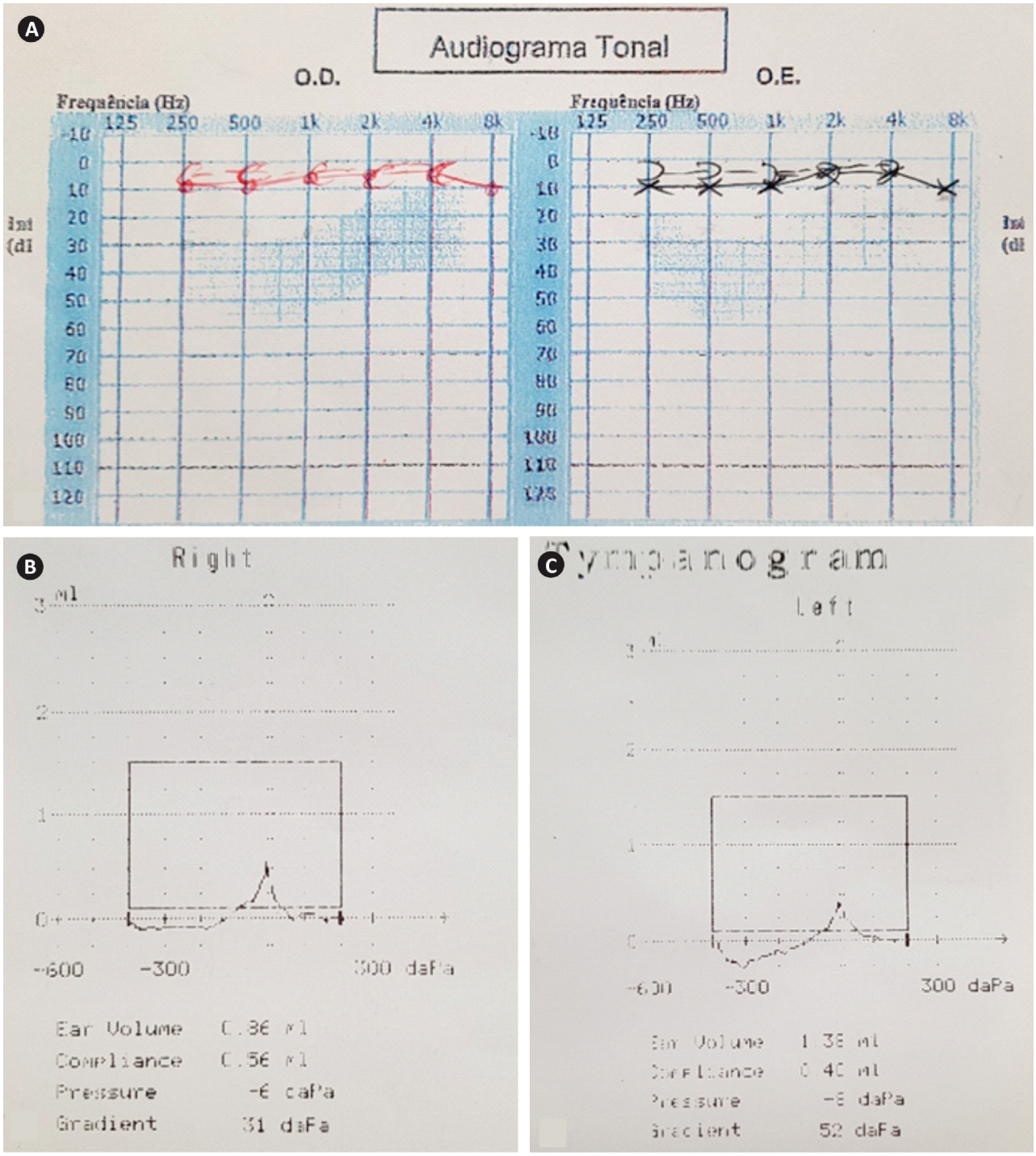


FIGURE 9
Tone audiogram (A) and tympanogram (B and C), at six months postoperatively



DISCUSSION

JNA is a rare benign tumor that almost exclusively affects adolescent boys 1-4 with an estimated incidence of 1:150.000.[2] The etiopathogenesis of JNA is not fully understood. It may arise due to vascular malformations, or from remnants of the first branchial arch artery.[14]

The pathogenesis of JNA is complex and involves androgenic hormonal factors, factors associated with angiogenesis, and the adenomatous polyposis coli/beta-catenin pathway.[15-17,20]

Because JNA occurs mostly during puberty, it is an androgen-dependent tumor. However, the results of a series of studies into the binding activity of sex hormones showed that the expression of androgen, estrogen, and progesterone receptors in angiofibroma in situ was not significant.[9,15,16]

Some evidence supports the notion that JNA is a predominantly vasoproliferative tumor.[14,21] The typical profile of vascular proliferation of this tumor suggests that some angiogenic growth factors are involved, including VEGF.[21]

Patients with FAP are diagnosed with JNA 25-fold more frequently than the general population.[15-17,20] The present patient had a family history of FAP and this disease will be excluded when the patient attends a medical genetics appointment.

The symptoms associated with this disease consist mainly of unilateral recurrent epistaxis and a sensation of ipsilateral nasal obstruction, as found in our patient. Other less frequent symptoms can arise, such as diplopia, reduced visual acuity, proptosis, facial edema, and facial hypoesthesia.[2] The latter symptoms can indicate an invasive lesion.

Large JNAs can obstruct the upper airway, leading to snoring and apnea, especially at night. This was one of our patient's complaints.

Because this is a lesion of the rhinopharynx, its close association with the opening of the Eustachian tube may lead to its obstruction, often causing dysfunction of the auditory tube.[2] This can be visualized by an objective examination. Otoscopy revealed otitis media with effusion in our patient, which explained the audiogram with conductive bilateral deafness and the type B tympanogram.

Otorhinolaryngological findings of JNA usually comprise a friable vascular mass that fills the entire affected nasal fossa and can extend to the oropharynx. Nasal fiber endoscopy can confirm these findings.

Obtaining a biopsy in clinical practice is not recommended given the risk of hemorrhage.[1,4,7]

Computed tomography (CT) of the NPS is the gold standard in diagnostic imaging. Enlargement of the pterygopalatine fossa is characteristic of JNA, with bone remodeling in the posterior wall of the maxillary sinus.[1,2,4,22]

Because of the expansive nature of these tumors, MRI SPN appears useful for characterizing the invasion of adjacent anatomical spaces, such as the orbits and masticator space, and of intracranial extension.[1,2,22]

Among several systems of JNA staging, the most popular is the Radkowski classification, which considers the tumor size and its relationship with the anatomical structures that it invades.[13]

Angiography is not essential for the diagnosis.[2,7] However, most surgeons consider preoperative embolization to be fundamental. Its main benefit is improved visualization during surgery and reduced blood loss. Vascular supply to JNAs is mainly provided by the internal maxillary and ascending pharyngeal arteries[2], which are branches of the external carotid artery (ECA). Although embolization of the ECA is safe, that of branches of the internal carotid artery carries a high risk of complications, including stroke, visual loss, facial paralysis, and carotid dissection.[2,7,8]

The treatment of these tumors is essentially surgical[2,11,12]

The surgical approach should be based on the patient's comorbidities, age, tumor extension and location, and the experience of the surgeon's.[2,22,23]

We applied a sublabial transmaxillary surgical approach with de Rouge-Denker incision to our patient due to extension to the masticator space (Radkowski stage IIC).

The application of radiotherapy has not gained a consensus. It is usually reserved for tumors with intracranial invasion and/or recurrence.[22,24,25]

A long-term follow-up of at least 5 years is recommended that includes annual nasal fiber endoscopy and MRI SPN.[2,25] Tumor relapse is primarily associated with incomplete resection and usually occurs between 6 and 36 months of follow-up.[22] However, tumors can stabilize over many years and even regress with age.[2,10-12]

CONCLUSION

JNA is a rare entity for which anamnesis and objective findings are strongly suggestive. However, its definitive diagnosis relies on histological findings.

CT, MRI, and angioembolization are important complementary means of presurgical diagnosis, which can establish anatomical relationships and help to determine the optimal approach. The gold standard of treatment is surgical excision.

Clinical suspicion in JNA is important as it allows a timely diagnosis and consequently reduces the associated comorbidities.

Conflict of Interest

The authors declare no conflict of interest regarding this article.

Data confidentiality

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

Human and animal protection

The authors declare that the followed procedures complied with regulations established by the Ethics and Clinical Research Committee and according to the Helsinki declaration of the World Medical Association.

Privacy policy, informed consent and approval by the ethics committee

The authors declare having written consent for the use of patients' photographs in this article.

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Availability of scientific data

There are no publicly available datasets related to this study.

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