Juvenile nasopharyngeal angiofibroma in a male of 16 years old. A case report

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Abstract

Juvenile nasopharyngeal angiofibroma (JNA) is a tumor occurs only in 0.05% of all head and neck tumors. We report a multisystemic approach combining study methods and collaboration with radiologist of a 16-years old male patient, diagnosed with a JNA measured 2x3x5 cm with retarded diagnosis who received endoscopic surgery management

Introduction

Juvenile nasopharyngeal angiofibroma (JNA) was first described in 1897 by Chelius as a polyp that took place during the adolescence and early adulthood stage. This type of tumor occurs only in 0.05% of all head and neck tumors, it is benign, and as the name implies, it is installed in the nasopharynx, more specifically, in the sphenoid process of the palatine bone with the horizontal wing of vomer and on the roof of the pterygoid process of the sphenoid bone. Histologically it is formed by a cylindrical and pavement epithelium with the characteristics of the epithelium found in the nasopharynx, below is a loose cellular tissue and the neoformation itself constituted by fibrous and highly vascularized tissue. JNA is almost exclusive to men, so it is believed that there is a certain hormonal relationship with the etiology that causes it, but still it hasn´t been described. Despite the histological benignity, its clinical behavior can be malignant due to the growth along natural holes and fissures related to its site of origin, causing bone and vascular destruction, accompanied by large hemorrhages. The vessels lack a muscular layer, this leads to excessive bleeding due to minimal manipulation or trauma of the tumor in question. (1-3)

Its symptomatology is somewhat unspecific and generalized, characterized by a progressive nasal obstructive syndrome, usually unilateral, which ends up obstructing the choana, all this is accompanied by night snoring, sleep apnea, headache, daytime oral breathing and mild epistaxis, Eustachian tube dysfunction may occur if there is a nasopharyngeal component by the tumor. Symptoms usually become present 6 months to 1 year prior to diagnosis (1,2)

On physical examination with anterior rhinoscopy, it usually reveals a reddish lobed mass located in the back of the nasal cavity and the cavum; Preoperative diagnosis is based on clinical and imaging characteristics; Incisional biopsy can lead to massive bleeding and is not recommended routinely. (2,3)

Unilateral carotid angiography is necessary to evaluate the vascular supply of the angiofibroma and allow embolization of the nutritional vessels before surgery, the main nutritional vessel is the internal maxillary artery. Occlusion of nutritional vessels reduces intraoperative bleeding, a major cause of morbidity and may reduce tumor size. With respect to the preferred method for embolization, there are 3 main methods for

embolization: trans-arterial, embolization with direct injection on the tumor and a combination of the two, there is no preferred technique on all three to perform an approach. (1,2)

Within the diagnostic approach, computed tomography of the skull and sinuses is useful to show the extent of bone erosion due to the tumor, while magnetic resonance imaging is useful to delimit the margins of the tumor, especially if there is any intracranial extension.

JNA can be staged with Fisch and Radkwoski classification.

Fisch classification:

- Stage I: Tumors limited to the nasal cavity and nasopharynx without bone destruction
- Stage II: Tumors that invade the pterygomaxillary fossa and the sinuses with bone destruction
- Stage III: Tumors that invade the intratemporal fossa, the orbit and / or the paraselar region, but that maintain a lateral location with respect to the cavernous sinus
- Stage IV: Tumors that invade the cavernous sinus, the area of the optic chiasma and / or the pituitary fossa.

Radkwoski Staging:

- Type IA Limited to nose or nasopharynx
- Type IB Extension into at least one paranasal sinus
- Type IIA Minimal extension into sphenopalatine foramen, Includes minimal part of medial pterygomaxillary fossa
- Type IIB Full occupation of pterygomaxillary fossa with Holman-Miller sign, Lateral or anterior displacement of maxillary artery branches, May have superior extension with orbital bone erosion
- Type IIC Extension through pterygomaxillary fossa into cheek, temporal fossa, or posterior to pterygoids
- Type IIIA Skull base erosion with minimal intracranial extension
- Type IIIB Skull base erosion with extensive intracranial extension +/- cavernous sinus

Regarding treatment, it was customary to perform a more extensive approach such as lateral rhinotomy and degloving of the facial middle third, open surgical methods are also used for locally advanced tumors, including those with intracranial involvement, optic nerve or the internal carotid artery. The currently recommended treatment is surgical excision, which may or may not be accompanied by preoperative embolizations.

Recently, thanks to the advances in the area of endoscopic surgery, this method has been used as a therapeutic option in appropriate cases, since in larger lesions, extrusion of the tumor is preferred.

Radiation has been used in tumors that are not resectable or that are already advanced. It is a second option, although it is not needed in a large percentage of cases. (2,3)

Recurrence in this type of tumor does not respond to cell metastasis or dedifferentiation, but rather to the persistence of residual tissue in the surgical site or in sites that, due to their location, are not resectable during the first surgical period. The main sites where residual tissue is found are: pterygoid canal, pterygoid process, pterygopalatine foramen, nasopharynx, pterygopalatine fossa, sphenoid sinus, and infratemporal fossa (to name a few). (4)

Case history / examination

16-year-old male patient with no important background history. His condition began 3 months ago, after a flu-like illness, where he perceives a predominantly right nasal obstruction, with greater intensity at night, accompanied by hyposmia, occasional bloody rhinorrhea; as well as retro nasal discharge, otic fullness, autophone and sometimes accompanied by mild epistaxis. Later, right hemicranial headache, facial heaviness, sensation of retro-ocular pressure is added to the movement; Hiponasal voice and nighttime snoring. He reports having a cough and tiredness for a long time already. He also describes having for a month, a feeling

of instability to the movement, accompanied by nausea or sometimes vomiting, and in the end a total nasal obstruction.

He goes to the general doctor and a gastroenterologist who only give symptomatic management. Afterwards he is referred to an otoneurologist who, rules out any neurological alteration that is causing the symptoms. While performing rhinoscopy, obstruction in the right nostril is found. The patient refers occasional mild epistaxis in the left nostril, without reporting it to be uncontrollable, as well as the flu 1 week ago, which was managed symptomatically. After 6 months he is referred to us for medical consultation in which the physical examination gives us the next findings:

Using a 0mm 4mm endoscope support: Left septal deviation. Turbinate hypertrophy. A pinkish-pearl right nasal tumor with a smooth, vascularized, ovoid surface that hangs from the back of the middle meatus and occupies the choana. Using a 70-degree endoscope retrogradely, nasopharyngeal occupation is seen incipiently. Right thick mucus is aspirated. Mouth, pharynx and otoscopy without alterations. Hematic biometry and blood chemistry within normal values

Differential diagnosis, investigations and treatment

The differential diagnosis should be made with other nasopharyngeal tumors, such as: Coanal Polyp, Adenoid Hyperplasia, Syphilis, Nasopharyngeal Tuberculosis, Chordoma, Craniopharyngioma, Paraganglioma, Teratoma, Lymphomas, Epidermoid Carcinoma, Adenoid Cystic Carcinoma, Sarcomas, among others.

Simple skull tomography with contrast was made with the following findings:

Tumor in the depth of the nasal passage on the right side, apparently hyper vascular that is insinuated in the sphenoparietal fossa, is related to a nasoangiofibroma as the first diagnostic possibility, other findings associated with inflammatory changes in the maxillary sinuses and hypertrophy in the mucosa of the lower cortices.

Hypervascular lesion of the choana and right nasopharynx with slight pterygomaxillary extension. As a finding, there is a 2cm arachnoid cyst. (Image 1)

Nuclear magnetic resonance imaging of the skull and facial mass: Tumor in the right nose and nasopharynx, without 2x3x5 cm extensions. Arachnoid cyst in the posterior fossa. Slight septal deviation. Concluding as probable diagnosis a right JNA (Stage I-II / IV) (Image 2)

At the day of hospital admission, a selective angiography of external carotid arteries was made, with a supraselective approach performing an embolization of branches of the right sphenopalatine artery. During the surgical procedure, the following information was reported as findings: The known tumor shows irrigation dependent on the sphenopalatine artery through three main branches: The vessels show delayed staining, whose appearance is irregular and tortuous. After embolization, the result is occlusion of 100% of the blood supply. (Image 3a and b).

Two days later, the otorhinolaryngology surgical procedure was performed. With prior embolization, made 48 hours before with gelfoam and stainless steel coils by Seldinger technique, endoscopic surgery is performed as follows: Right anterior and posterior ethmoidectomy, opening of the recess and maxillary antrostomy III, sphenoidectomy (Image 4a), cauterization of the lower corneal tail, cut of middle and upper turbinates; The medial part of the posterior wall of the right maxillary sinus is resected, identified, dissected and coagulated (Image 4b) and the sphenopalatine artery (already embolized) is cut, and a piece of the tumor is resected, extracted through the right nostril (Image 5). Evista, Gelfoam and Merocel are placed in the surgical bed. The patient evolves adequately and is discharged to his home two days after surgery.

Outcome and follow-up

During the postoperatively revision consultation in the office, a week after the procedure, tamponade removal and healing were performed, without complications. The patient is now a year free of complications and with an excellent otcome.

Discussion

JNA is a pathology with a low incidence, however, this should not be a factor for us to forget its possibility as a differential diagnosis in the presence of symptoms such as epistaxis, progressive nasal obstruction, sleep apnea, among others. In the case of our patient, a correct managment was delayed for more than 6 months until an adequate diagnosis could be made after being evaluated by multiple specialists. (1,2)

Despite its benign nature, JNA is a rapidly growing and aggressive tumor, leading to bone destruction as well as invasion of adjacent structures. It has a well-defined capsule that allows for correct manipulation and dissection during surgery. (7,8,9)

The resection selected for our patient according to his Radikowski stage (I-II /IV) was endoscopic surgical correction. This procedure is complicated but when performed correctly is an effective surgery, managing to limit recurrence with follow-up to 24 months, attributing it to the advantageous multi-angle view of the anatomical structures. Enabling effective, atraumatic dissection using bipolar coagulation with gentle manipulation of the tumor. Among the decisive factors that were carried out correctly in our patient was a correct perforation of the bone invaded by the tumor, endonasal surgery combined with a preoperative embolization of the arterial supply of the tumor, thus achieving serious blood loss avoidance. (7,8,9,10)

In tumors that fall within the Radkowski IA-IIB classification, endoscopic surgery remains an appropriate option. In the event that the tumor extends laterally in the fossa infratemporalis or deep into the skull base, endoscopic surgery is no longer recommended (10)

Authorship list

Author contributions:

Author 1: Head of the project, leader of the literature revision

Author 2: Head surgeon of the endoscopic procedure

Author 3: Head neuroradiologist that made the supra-selective embolization.

Author 4: Assistant neuroradiologist that made the supra-selective embolization.

Author 5: Assistant surgeon of the endoscopic procedure

Author 6: Assistant of the literature revision

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Tables and images

Image 1- Axial cut of CT findings

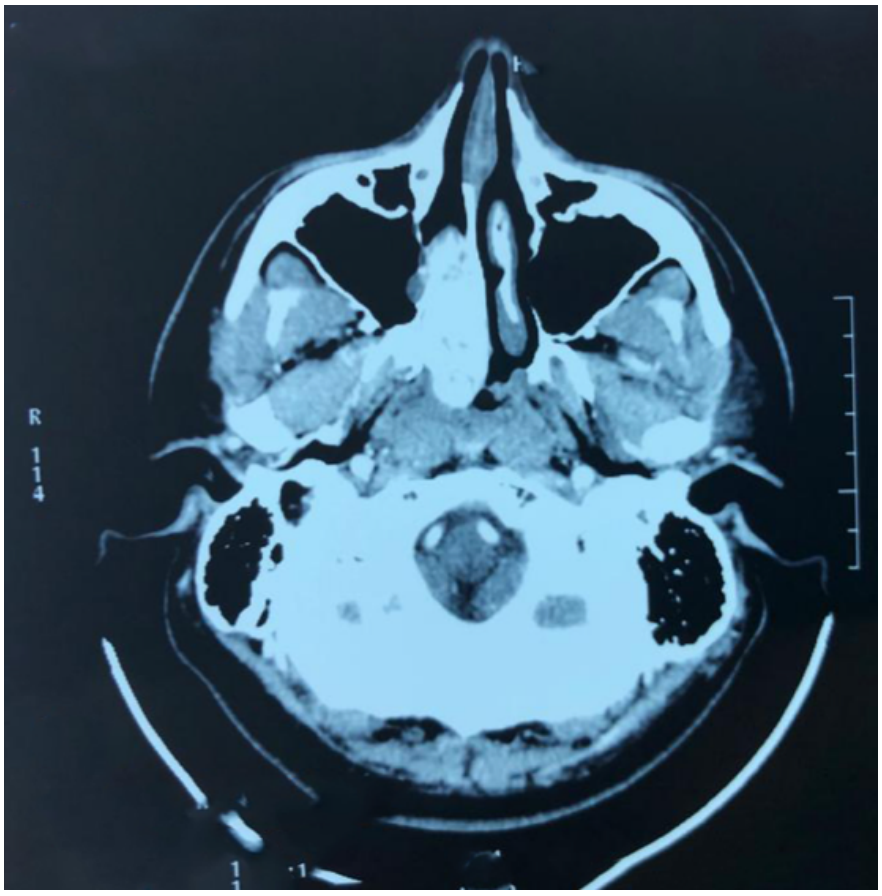
Image 2- Axial cut of Magnetic Resonance findings

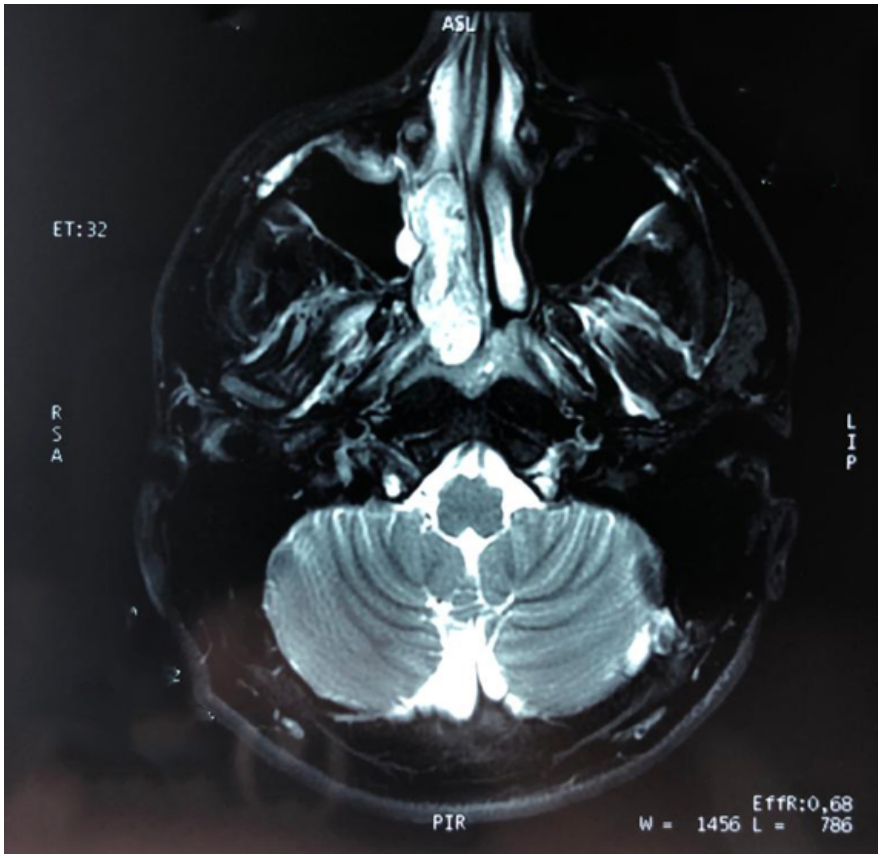
Image 3a and b- Carotid angiography – a) Pre and b) post embolization of supra-selective approach of the right sphenopalatine artery

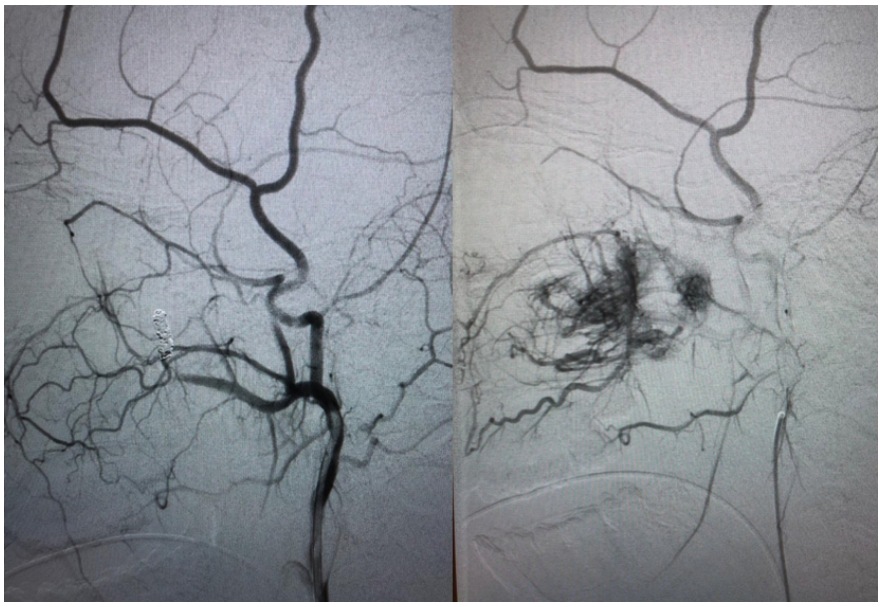
Image 4a and b - Endoscopic resection – a) Maxillary and sphenoid sinusotomy. b) View and wide opening of maxillary, ethmoid and sphenoid sinuses together with tumor pedicle

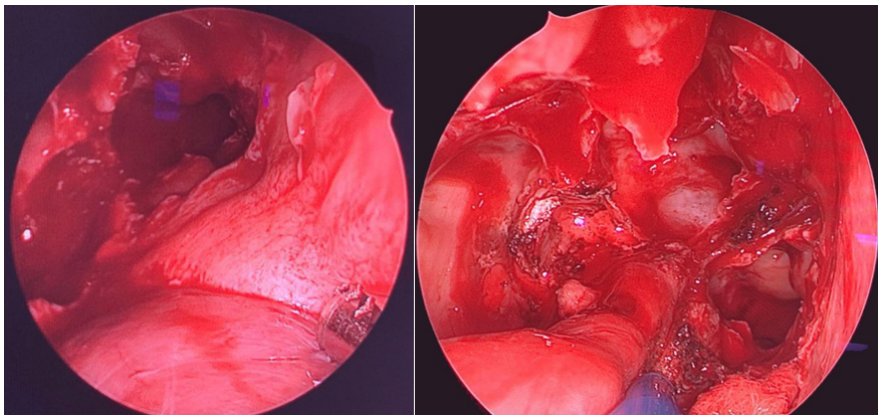
Image 5- Extraction of the NJA

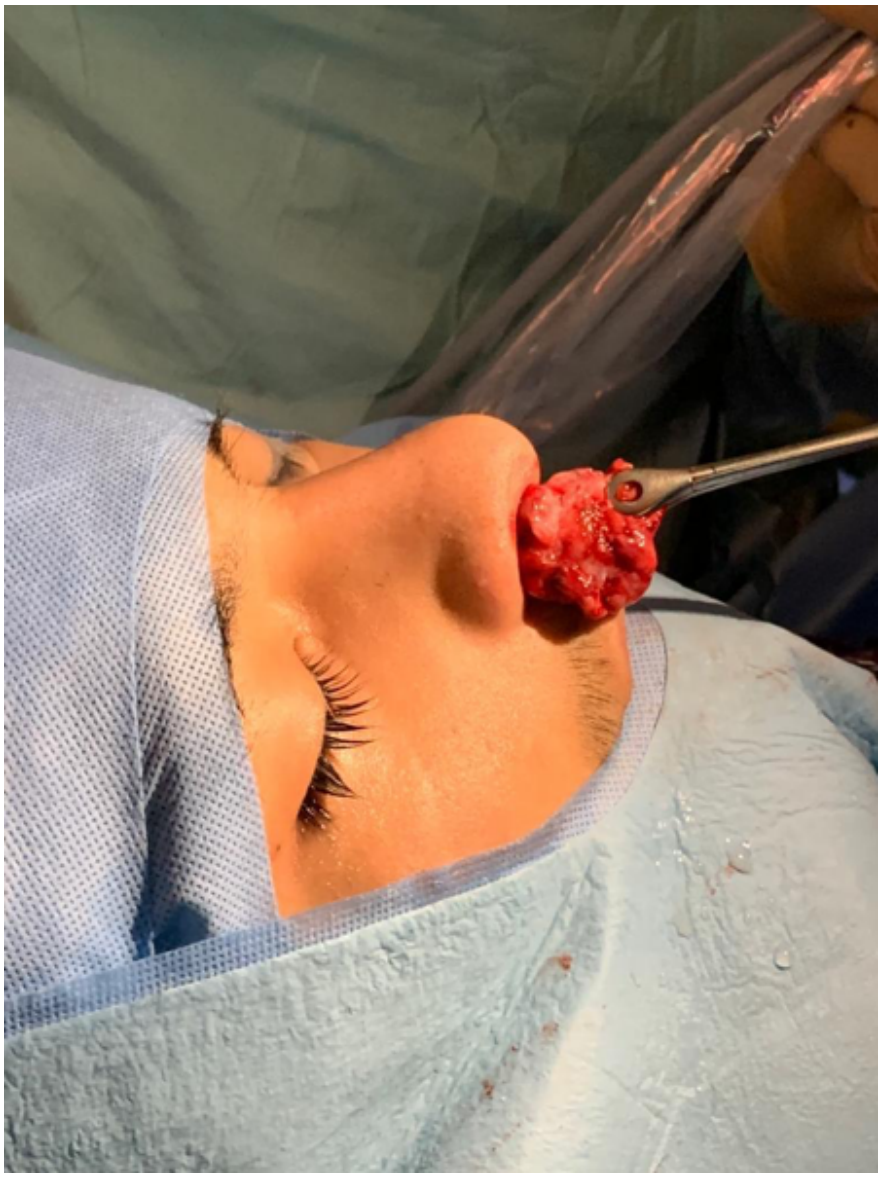
Video 1 - Moment when the pedicle is sectioned and cauterized











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Video 1.mov available at https://authorea.com/users/353829/articles/477551-juvenile-nasopharyngeal-angiofibroma-in-a-male-of-16-years-old-a-case-report