



Endoscopic Approach of Juvenile Nasopharyngeal Angiofibroma - A Sustained Surgical Challenge

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Introduction

Juvenile Angiofibroma (JNA) is a benign vascular tumor arising usually from the posterior part of the nasal cavity (pterygoid region), which appears at young male teenagers. The tumor has a destructive growth in the surrounding areas; therefore the management of JNA needs to be initiated in early stages of tumor development.

The ethiopathology of JNA is controversial and not yet completely explained. There are several theories to be considered. Monoclonal factors like exogenous testosterone can induce tumor hypertrophy [1]. There is also a genetic theory of JNA appearance which is still to be proved by studies on large series of angiofibromas [2]. There are authors who try to demonstrate the role of viral infections (Herpes simplex virus, Epstein Barr virus, HPV) in the appearance and growing of JNA [3]. The vascular theory, which reveals the over expression of receptors for vascular endothelial growth factor in presence of JNA, can explain the development of this tumor [4].

Preoperative Protocol

First step in this pathology is to have an accurate diagnosis. In every patient of our series we counted on clinical history, endoscopic exam and CT imaging. Unilateral nasal obstruction with moderate growth, associated with repetitive unilateral epistaxis, facial pressure, headache and conductive hearing loss on the same side, which appeared at an adolescent man in the last 6 months, determined us to take into account a JNA.

The endoscopic view shows a reddish lobulated mass, with a firm structure which seems to have the originating point close to sphenopalatine foramen (Figure 1).

CT scan is the major imagistic investigation which gives complex information about the tumoral mass and its extensions and is a leading point in establishing the tumoral stage (Radkowski staging system) and the surgical management of the tumor [5].

There are two very important signs on CT scan which are characteristic for JNA. The destruction of the pterygoid plates and the enlargement of the sphenoid foramen on the tumor site are images of choice for the diagnosis of angiofibroma (Figure 2).

After establishing an endoscopic and imagistic diagnosis of JNA, the next step in the management of this pathology is the bilateral control angiography [6]. Embolization of feeding vessels in angiofibroma leads to:

- Decrease the vascular supply of the tumor (usually from the internal maxillary artery)
- Exact visualisation of the vessels which feed the tumor
- Decrease the tumor volume which allows an easier surgical procedure, especially in the approach of the tumoral extensions in the surrounding areas (sinuses, orbital cavity, pterygomaxillary fossa, etc)

We perform angiography with embolization 24 hrs to 48 hrs before surgery, using Polyvinyl Alcohol (PVA) as embolic substance (Figure 3).

Dealing with a unilateral bleeding mass appeared at an adolescent male we do not perform a biopsy from the tumor because of the high risk of bleeding. In our experience, the involvement of choice for all steps of JNA is the surgical approach. Depending on the staging system [7], we perform an endoscopic approach (in stage I and II Radkowski) and usually an open approach in Radkowski stage III.

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Figure 1: Left JNA-endoscopic view.



Figure 2: CT scan for a right JNA.

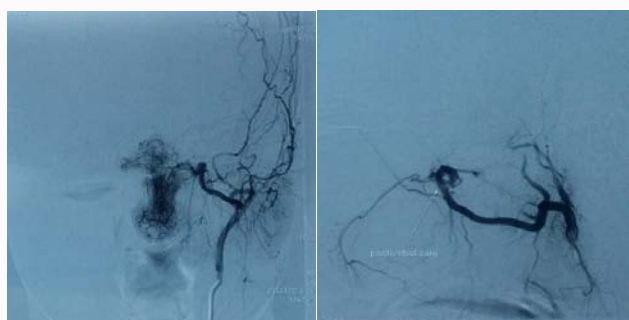


Figure 3: Angiography, pre and post-embolization.

Endoscopic Surgical Tricks

We tried to summarize some surgical tricks in endoscopic approach of JNA. So while dealing with a tumor arising from the sphenopalatine foramen we perform a meticulous local hemostasis with bipolar cautery, radiofrequency or coblation of the exact site of origin of the tumor. One of the most frequent sources of intraoperative bleeding is the vidian artery which can arise from internal carotid artery and is difficult to embolize. In case of extension of the tumor in the maxillary sinus we perform a middle maxillectomy and if there is a pterygopalatine fossa involvement we open the posterior wall of the maxillary sinus in order to find the margin of the tumor in contact with the fat of the region (Figure 4).

The surgical approach for the extension of the tumor in the ethmoid sinus consists of complete ethmoidectomy, usually with middle and superior turbinate resection (in order to get enough space for complete resection of the tumor) and the opening of the frontal sinus.

The sphenoidal extension of the angiofibroma can be safely

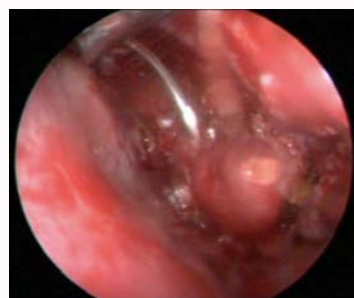


Figure 4: Tumor in right pterygopalatine fossa (arrow-posterior wall of the maxillary sinus).



Figure 5: Transtumoral stitch for posterior displacement.

removed by opening the anterior and inferior walls of the sphenoid sinus.

Sometimes the complete resection of the tumoral extensions can be very challenging for the surgeon. This is why we are using a surgical stitch which passes transtumoral and is turned into the rhinopharynx and then exteriorized through the mouth. By pulling the stitch, the tumor is displaced into the nasal cavity from its extensions (very useful for pterygopalatine localization of the angiofibroma) succeeding by this surgical maneuver.

The posterior mobilization of the tumor, with low risk of bleeding. By this technique we can visualize the arising part of the angiofibroma, safely detach the tumor from the surrounding tissue and perform a complete removal through the mouth for large fibromas (Figure 5).

Another issue we faced in our series of JNA was the way of resection: piece meal versus en block. It is indeed more secure and safe to have to tumor resection in one piece, and if the volume is too big we can take out the angiofibroma through the mouth. Having in mind that this tumor usually induces a displacement of the surrounding structures and not a true invasion, the option of piece meal resection is acceptable, but the surgical endoscopic technique is mandatory to take into account the position of all vessels feeding the JNA.

Conclusions

It is well-known that there is no recurrence of JNA after surgery, but only incomplete resection [8]. In the endoscopic approach of JNA our goal is complete removal of the tumor without any leftovers. In order to avoid any residual tumor, we recommend:

- Early diagnosis of the tumor
- Complete embolization of the feeding vessels
- Precise endoscopic surgical technique
- Dry surgical field, with no source of intraoperative bleeding

- Meticulous endoscopic control of the tumoral extensions site of the end of the surgery
- Rigorous follow-up (endoscopic exam, CT imaging)

In practice, our experience has shown that even if all the steps above are followed, JNA is still a challenging pathology and the surgeon has to be able at any time to assure complete hemostasis during surgery (endoscopic or if needed open approach).

Endoscopic endonasal surgery of JNA is only limited by the extensions of the tumor in surrounding anatomical regions and the experience of the surgical team.

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