The Rhinologist

The Rhinologist 2.2018

ISSN 2531-9299

Italian Academy of Rhinology

Table of Contents

Editor: PSA&CF srl Via Procaccini, 24 21100 Varese

Registered in Varese Court at n. 5/2016

www.therhinologist.com

Editor in Varese
Four monthly Magazine

Varese, 1-7-2018

ISSN 2531-9299

ISSN 2531-9299

Certificato Qualità ISO 9001:2008
ICIM
Cert.N. 5232/0

ENDOSCOPIC ENDONASAL JUVENILE NASOPHARINGEAL ANGIOFIBROMAS RESECTION: NECESSARY SURGICAL STEPS

Mariella Scarano MD, Iacopo Dallan MD, Lodovica Cristofani-Mencacci MD, Veronica Seccia MD, PhD

Affiliations:
1st Otorhinolaringology Unit, Azienda Ospedaliero-Universitaria Pisana, Via Paradisa 2, 56124 Pisa, Italy

Running Title: Endoscopic endonasal resection of limited an extended vascular nasopharyngeal tumour

Conflict of Interest: The authors declare that there is no conflict of interest regarding the publication of this article.

Corresponding Author:
Mariella Scarano, M.D., 1st Otorhinolaringology Unit, Azienda Ospedaliero-Universitaria Pisana, Via Paradisa 2, 56124 Pisa, Italy, Email: mariella.scarano88@gmail.com Tel and Fax: 050997545
1: 1st Otorhinolaringology Unit, Azienda Ospedaliero-Universitaria Pisana, Via Paradisa 2, 56124 Pisa, Italy

ABSTRACT

Juvenile nasopharyngeal angiofibroma (JNA) is an uncommon, highly vascular neoplasm originating in the nasopharynx that is generally seen in prepubertal and adolescent males. Although it is histologically benign, it has a high destructive potential and recurrence. Its symptoms are generally scarce and unspecific and are represented by nasal obstruction and epistaxis, but when present in a persistent manner, they are highly suspicious.
Routine preoperative imaging (CT, MRI with contrast media enhancement; angiography) is used to confirm the diagnosis and define tumour's extension and staging, and imaging also assists in the treatment planning.
The mainstay treatment is resection combined with preoperative embolization; for early stage JNA, endoscopic endonasal resection is considered the treatment of choice.
This paper provides a detailed description of the surgical endoscopic steps of this procedure. Tips and tricks learned from our experience are provided to perform a safe and minimally disabling surgery for the patient while obtaining the oncologic goals of surgery.
A review of the literature with particular emphasis on histological features, differential diagnosis, and treatment of choice is included.

Keywords: juvenile nasopharyngeal angiofibroma, endoscopic resection, surgery

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a rare, benign, locally aggressive lesion composed of vascular and fibrous elements in varying proportion. It represents between 0,05% and 5% of all head and neck tumours and typically affects adolescent males. Its incidence appears to be higher in the Middle East and India than in Europe (1).

The exact nature of JNA is still in dispute. Although the histological appearance of the tumour is benign, some theories suggest that it could possibly be a vascular malformation (or hamartoma) related to incomplete regression of the first branchial arch artery (2-3).

The clinical presentation of a JNA is characterized by monolateral persistent nasal obstruction coupled with rhinorrhoea and sometimes epistaxis. Nowadays, the typical JNA is diagnosed by computed tomography (CT) scanning or magnetic resonance imaging (MRI) with contrast media and has high levels of diagnostic safety, making pre-operative biopsy unnecessary (4).

The gold-standard therapy for JNA is radical surgery; other treatment methods (e.g., chemotherapy, radiotherapy, hormone therapy) have been emphasized in the past, but they are currently only used as occasional complementary treatments (5).

Endoscopic approaches have become the treatment of choice, especially for tumours limited by size and location criteria.

A number of case reports and series of endoscopically resected JNAs have been published that demonstrate that smaller lesions can be resected with minimal morbidity and with persistence and recurrence rates that are at least as good as those achieved with open approaches (6-7).

Therefore, we report a paradigmatic case of a JNA that was treated with exclusive endoscopic endonasal surgery. The 15-year-old male patient was treated in our unit (1st Otorhinolaryngology Unit, Azienda Ospedaliera Universitaria Pisana, Pisa, Italy) in February 2015. This paper focuses on the surgical technique that can be adopted to enable limited-extent tumours to be removed endoscopically and the anatomic features that are likely to define the limits of tumour extension that permit a successful endoscopic resection.

CASE REPORT

At our institution, endoscopic JNA resection was performed on a 15-year-old patient presented with bilateral nasal obstruction and recurrent epistaxis and rhinophonia.

The CT scan and the MRI with contrast enhancement revealed a 6,5 cm mass obstructing the rhinopharynx, extending to right pterygopalatine fossa (PPF) without evidence of intracranial extension [stage IIA according to Radkowski *et al.* classification (8)] **(Fig 1A,2A).**

Preoperative angiography showed a highly vascular tumour with feeding vessels from the right maxillary artery. The patient underwent a selective arterial embolization followed by an endoscopic-endonasal resection.

Surgical endoscopic technique for tumour removal

1- Exposure of the sinonasal corridor. The initial step of the procedure makes space in order to make the sinonasal corridor visible. Therefore, generally, the lower portion of the right middle turbinate is removed, together with an uncinectomy and a large middle meatal antrostomy, to expose the posterior wall of the maxillary sinus. The bulla ethmoidalis and posterior ethmoid cells are removed to expose the anterior wall of the sphenoid. The natural ostium of the sphenoid can be identified using the tail of the superior turbinate as a surgical landmark. The same surgical steps are performed in the contralateral side **(Fig.3A)**. In our case, the tumour extended through the floor of the sphenoid sinus, so the anterior wall of both sphenoid sinuses was drilled out in order to create a single sphenoidal cavity.

2- Identification of the surgical anatomic landmarks. Bilaterally, a subperiostal dissection on the lateral nasal wall is performed, starting approximately 1 cm anterior to the tail of the middle turbinate. Just behind the crista ethmoidalis of the palatine bone, the sphenopalatine artery (SPA) is identified. Then, the SPA is cauterized and cut to expose the sphenopalatine foramen (SPF) **(Fig. 3B)**.

3- Opening the surgical window to approach the PPF. On the right side (tumor side)**,** the surgical window is expanded inferiorly by drilling out the vertical plate of the palatine bone as well and by removing the posterior half of the inferior turbinate together with the posterior portion of the medial maxillary wall. The posterior wall of the maxillary sinus is removed in a medial-to-lateral direction, generally as far as the sagittal plane passing through the infraorbital nerve (ION). The tumour within the PPF and any infratemporal extension is exposed. Once the periosteal layer containing the PPF has been incised, the fibrofatty tissue surrounding the maxillary artery and its branches are exposed. Vascular clips are positioned on the medial aspect of maxillary artery to avoid bleeding into the surgical field. Visible collateral vessels are cauterized with a bipolar diathermy and cut **(Fig. 3C)**.
Then, the tumour is detached from its attachments on the septum, postnasal space, and lateral nasal wall, and the angiofibroma is mobilized.
Generally, the mass cannot be brought out through the nasal cavity because of its bulkiness; rather, it is pushed posteriorly toward the posterior nasopharyngeal wall, brought out through the oropharynx, and finally delivered through the mouth.
4- Drilling out the limiting bone due to the risk of recurrence. What seems to be important for preventing recurrences of JNA is drilling out the cancellous bone of the pterygoid roots and basisphenoid, particularly around the vidian canal, to remove any residual disease that may not be immediately evident (9)**(Fig. 3D)**.
The tumour specimen itself measured approximately 6,5 cm in diameter. The tumour was resected en bloc with minimal bleeding. There were no intraoperative complications. The definitive histological diagnosis was nasopharyngeal angiofibroma. A post-operative MRI shows a total resection with no remaining macroscopic disease **(Fig. 1B,2B).**

DISCUSSION

JNA is a rare, benign, highly vascular tumour that appears in proximity to the sphenopalatine foramen and occurs in young males. Recurrent, unprovoked, painless, profuse, unilateral epistaxis (60%), and unilateral nasal obstruction (80%) with rhinorrhea characterize the typical clinical presentation of JNAs. Nasal examination is recommended for every young man with these symptoms to

exclude JNA. In rare, underestimated cases, advanced tumours may present with facial swelling and visual or neurological disturbances (10).

From a histologic point of view, at a microscopic examination, the stroma of the tumour contains blood vessels of different sizes and shapes lined by endothelial cells but with little or no smooth muscle or elastic fibres. This structure, lacking muscles fibres, contributes to the capacity of JNAs to bleed excessively after minimal manipulation (11).

The tumour shows an expansive and destructive growth pattern. It usually arises near the sphenopalatine foramen and extends towards nearby structures. It has the peculiar tendency to grow in the submucosal plane into the nasopharynx, along the vidian canal into the basisphenoid, and laterally towards the pterygopalatine and infratemporal fossa. The tumour may expand vertically from the pterygopalatine fossa through the inferior orbital fissure into the orbit. From the infratemporal fossa, the tumour can grow superiorly through the base of the pterygoid process and reach the middle cranial fossa. The lateral and posterior walls of the sphenoid sinus can be eroded as well as the cavernous sinus and pituitary gland involved. Erosion of the anterior skull base is seldom observed (12).

Early diagnosis and treatment are required for a good prognosis; biopsy is almost unanimously considered contraindicated, since it carries a considerable and undue risk of haemorrhage (13).

According to Radkowski *et al.* (8), JNA is classified into three types. Type I includes lesions fundamentally localized to the nasal cavity and nasopharynx with involvement of at least one paranasal sinus. Type II is a JNA extending into the pterygopalatine fossa and infratemporal fossa with or without orbital bone erosion. Type III is a JNA that erodes the skull base with intracranial extension. It is clear that a careful preoperative radiologic evaluation is essential to determine the nature of the lesion, its staging, and an optimal surgical approach. Additionally, bilateral carotid angiography is required to assess the vascular supply of JNA, which generally derives from branches of the external carotid arteries and its maxillary and ascending pharyngeal branches (14), and allow pre-operative embolization of feeding vessels, generally 24 to 48 hours before surgery.

Though some authors questioned the usefulness of this procedure, since in their experience no significant difference in surgical bleeding was observed, there is increasing evidence that embolization is a safe and effective method to reduce intra-operative blood loss (15-16). Nevertheless, the lesion shrinkage achieved by embolization has been indicated as a contributory cause to incomplete excision of juvenile angiofibroma by McCombe et al. (17).

As previously stated, surgery is considered the mainstay of treatment for JNAs. In the past, surgical approaches were mainly trans-facial, including lateral rhinotomy, transpalatal, and transmaxillary routes as well as midface degloving approaches, depending on the size of the tumour as well as on the surgeon's experience. The use of endoscopic sinus surgery for the resection of JNAs is a relatively new phenomenon that has become the standard approach for tumours that are limited to the nasal cavity and nasopharynx (12).

During all surgical endoscopic steps, the main anatomic landmarks must be constantly kept under control to guarantee safe access to deep structures. Moreover, complications can be minimized by excellent exposure of the surgical field and meticulous dissection of neurovascular structures contained into PPF, improving tumour dissection and removal.

The step-by-step surgical procedure is tailored to the extension of the lesion and should be treated as follows: expose the sinonasal corridor, identify the surgical

anatomic landmarks, open the surgical window to approach the PPF that is primarily involved in the tumour, and drill out the limiting bone (18).
A team approach is essential. The treatment decision should be approved by a multidisciplinary group so as to provide the patient with maximum benefits from a group with broad expertise.

CONCLUSIONS

An exclusively endoscopic management of juvenile nasopharyngeal angiofibroma appears to be effective for small to medium tumours. It should be considered as a first-choice option for these cases due to its minimal bleeding, shorter duration, and efficacy.
Endoscopic endonasal surgery in JNA is only limited by the extension of the tumour in surrounding anatomical regions and the experience of the surgical team.

REFERENCES

1. Gullane PJ, Davidson J, O'Dwyer T et al. Juvenile Angiofibroma: a review of literature and a case series report. Laryngoscope 1992;102:928-933.
2. Zhang M1, Sun X, Yu H, at al. Biological distinctions between juvenile nasopharyngeal angiofibroma and vascular malformation: an immunohistochemical study. Acta Histochem. 2011 Oct;113(6):626-30.
3. Shick B, Plinkert PK, Prescher A et al. Aetiology of angiofibromas:relation on their specific vascular component. Laryngorhinootologie 2002;81:280-284.
4. Mairoldi R, Niccolai P. Imaging in Treatment Planning for Sinonasal Diseases. 2015.
5. López F, Triantafyllou A , Snyderman CH et al. Nasal juvenile angiofibroma: Current perspectives with emphasis on management. Head Neck. 2017 May;39(5):1033-1045. Epub 2017 Feb 15.
6. Pryor SG, Moore EJ, Kasperbauer JL. Endoscopic versus traditiona lapproaches for excision of juvenile nasopharyngeal angiofibroma. Laryngoscope 2005;115:1201-1207.
7. Nicolai P, Villaret AB, Farina D, et al. Endoscopic surgery for juvenile angiofibroma: a critical review of indications after 46 cases. Am J Rhinol Allergy 2010;24:e67–e72.
8. Radkowski D, McGill T, Healy et al. Angiofibroma: changes in staging and treatment. Arch Otolaryngol Head and Neck Surg 1996;122:122-129.
9. Howard DJ, Lloyd G,Lund V. Recurrence and its avoidance in juvenile angiofibroma. Laryngoscope 2001; 111:1509–1511.
10. Lopez F, Suarez V,Suarez C et al. Treatment of juvenile angiofibromas: 18-year experience of a single tertiary centre in Spain. Rhinology 2012;50:95–103.
11. Garca MF, Yuca SA, Yuca K. Juvenile nasopharingeal angiofibroma. Eur J Gen Med. 2010, 7(4):419-425.
12. Roger G, Tran Ba Huy P, Froehlich P, et al. Exclusively endoscopic removal of jouvanile nasopharyngeal angiofibroma: trands and limits. Arch Otolaryngol Head Neck Surg 2002;128:928–935.
13. Scholfield DW, Brundler MA, McDermott AL, et al. Adjunctive treatment in juvenile nasopharyngeal angiofibroma: how should we approach recurrence? J Pediatr Hematol Oncol 2016;38:235–239.
14. Mehan R, Rupa V, Lukka VK et al. Association between vascular supply, stage and tumour size of juvenile nasopharyngeal angiofibroma. Eur Arch Otorhinolaryngol 2016;273:4295– 4303).
15. Siniluoto TM, Luotonen JP, Tikkakoski TA et al. Value of pre-operative embolization in surgery for nasopharyngeal angiofibroma. J Laryngol Otol 1993;107:514–521.
16. Li JR, Qian J, Shan XZ, Wang L. Evaluation of the effectiveness of preoperative embolization in surgery for nasopharyngeal angiofibroma. Eur Arch Otorhinolaryngol 1998;255:430–432.
17. McCombe A, Lund VJ, Howard DJ. Recurrence in juvenile angiofibroma. Rhinology 1990 June;28(2):97-102.

18. Sprekelsen M B, Alobid I. Endoscopic approaches to the paranasal sinuses and skull base. s.l. : Thieme, 2017.
19. Coutinho–Camillo CM, Brentani MM, Nagai MA et al. Genetic alterations in in juvanile nasopharyngeal angiofibromas. Head Neck 2008 mar; 30(3):390-400.
20. De Vincentiis M, Gallo A, Minni A et al. Preoperative embolization in the treatment protocol for rhinopharyngeal angiofibroma: comparison of the effectiveness of various materials [in Italian]. Acta Otorhinolaryngol Ital 1997;17:225-232.
21. Allensworth JJ, Troob SH, Lanciault C, Andersen PE. High-grade malignant transformation of a radiation-natıve nasopharyngeal angiofibroma. Head Neck 2016;38(Suppl 1):E2425–E2427.
22. Beham A, Kainz J, Stammberger H et al. Immunohistochemical and electron microscopical characterization of stromal cells in nasopharyngeal angiofibromas. Eur Arch Otorhinolaryngol. 1997;254(4):196-9.

Acknowledgements
We thank Ms. Anna Giovinazzo for her help in organizing the scientific work.

Conflicts of Interests
We state that all Authors have contributed to, read, and approved this manuscript.
We declare that this manuscript has not been previously published, nor is it under consideration elsewhere and that none of the authors have any conflicts of interests, financial or otherwise.

Disclosure Statement
The authors have no commercial, proprietary, or financial interest in the products or companies described in this article.

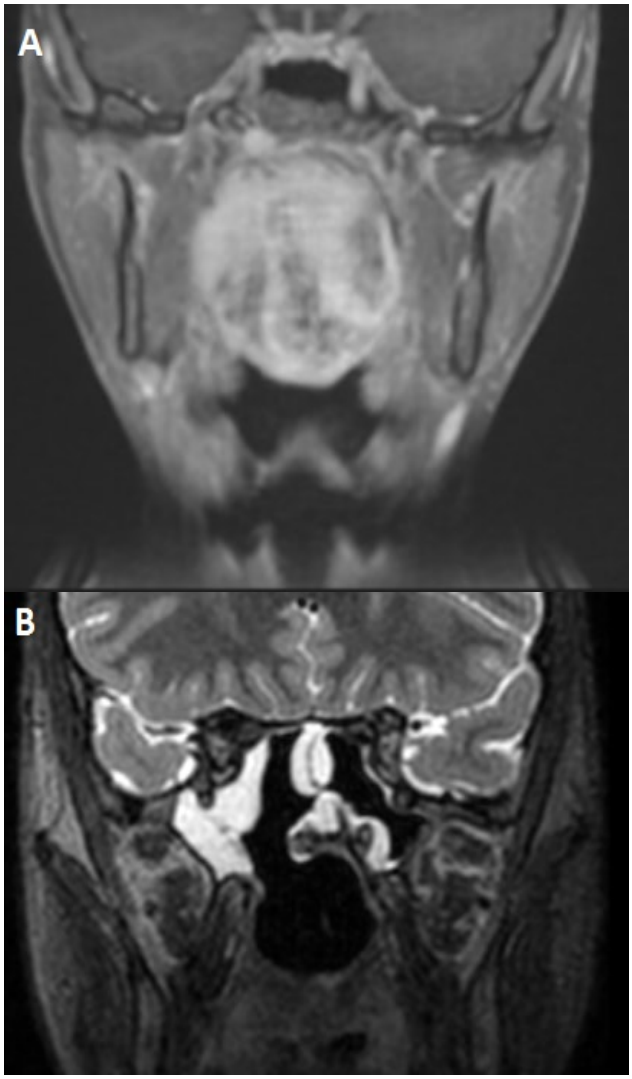
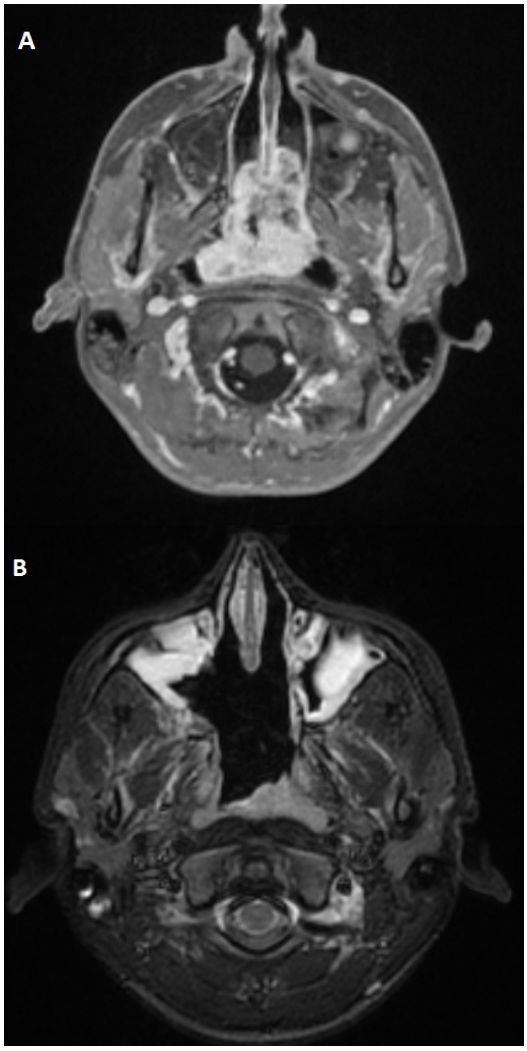


Figure 1. A) Preoperative postcontrast, T1 coronal MRI scan. **B)** Postoperative, T2 coronal MRI scan.



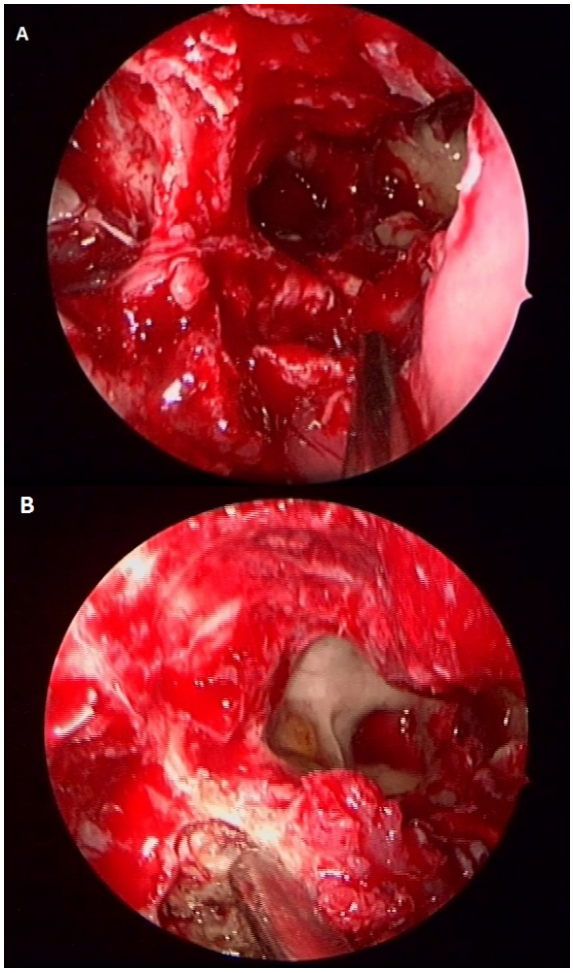


Figure 2. A) Preoperative postcontrast, T1 axial MRI scan. **B)** Postoperative, T2 axial MRI scan.

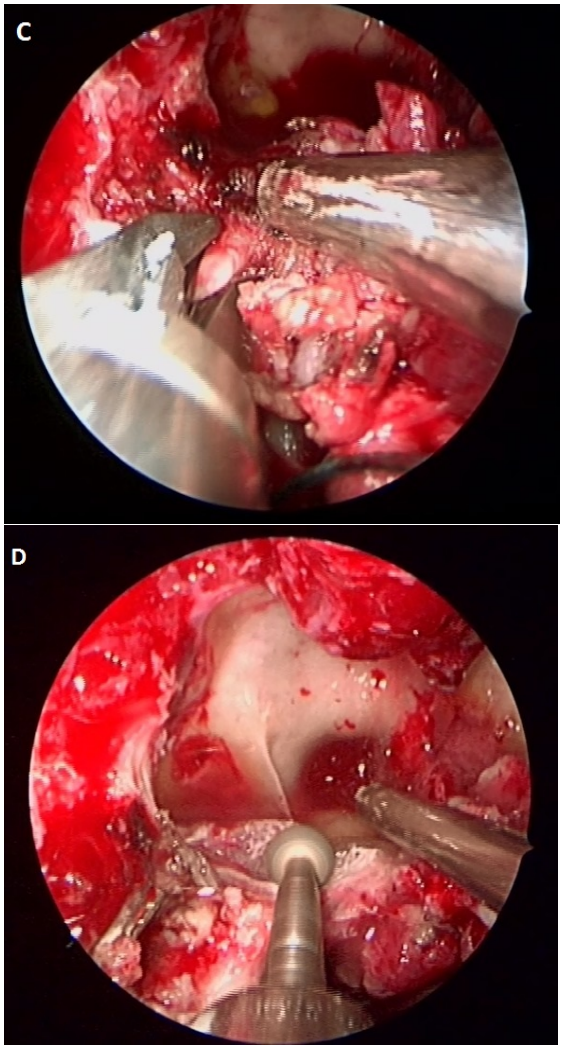


Fig 3. Surgical steps. A) Exposure of the sinonasal corridor. **B)** Identification of the surgical anatomic landmark. **C)** Opening the surgical window to approach the PPF. The right maxillary artery is exposed and clipped**. D)** Drilling out the limiting bone.