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(CASE REPORT)

Juvenile nasopharyngeal angiofibroma: A clinical case report and a literature review

John Esteban Verdugo Parra [1,*], Kevin Fabricio Orellana Barros [1], Hugo Xavier Guamán Roldan [2] and Janneth Alexandra Villavicencio Benavides [3]

[1] *Students of the Faculty of Dentistry at the University of Cuenca, Ecuador.*
[2] *Oral and maxillofacial surgeons from the National Autonomous University of Mexico, Mexico; Lecturer at the Faculty of Dentistry at the University of Cuenca, Ecuador.*
[3] *Specialist doctor in otorhinolaryngology at the Portoviejo Specialty Hospital, Ecuador.*

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Abstract

Juvenile nasopharyngeal angiofibroma (JNA) is a benign tumor with locally aggressive behavior, given the significant epistaxis it causes, frequent complications, and therapeutic complexity presented by this condition, diagnosis and treatment should be thorough; it occurs exclusively in adolescent males and accounts for 0.5%-0.05% of all head and neck tumors. This case corresponds to an 18-year-old patient with progressive respiratory difficulty over 2 years, nasal obstruction, and epistaxis. Clinical and imaging studies revealed a diagnosis of JNA at stage IIb according to Radkowski's classification. A Degloving approach was performed, and the tumor was completely removed. One year after resection, no tumor recurrence has been recorded.

Conclusion: Juvenile nasopharyngeal angiofibroma predominantly affects adolescent males and is primarily characterized by recurrent episodes of epistaxis. Surgical management remains the cornerstone of treatment, with various effective approaches available. The local recurrence rate is low, underscoring the effectiveness of current interventions in most cases.

Keywords: Angiofibroma; Epistaxis; Nasal Obstruction; Classification; Diagnosis; Approach; Degloving.

1. Introduction

Juvenile nasopharyngeal angiofibroma (JNA) is a benign, highly vascularized, locally aggressive tumor located in the nasopharynx, specifically affecting the lateral wall in prepubescent and pubescent boys. It occurs almost exclusively in adolescent males, accounting for 0.5%-0.05% of all head and neck tumors, with varying figures in some Asian and African countries. Presentation in females may require chromosomal studies or other differential diagnoses. The tumor invades the infratemporal fossa, ethmoid, orbital region, and sphenoid in 90% of cases, and the pterygopalatine fossa in 76%. Brain invasion incidence ranges from 4.3% to 11%, reaching up to 26% in some cases. Common manifestations include nasal obstruction and unilateral epistaxis in early stages, with other symptoms depending on the lesion's extent and compression of adjacent structures. Radiological imaging and endoscopy are essential for diagnosis and surgical planning, while biopsy is contraindicated due to the risk of severe hemorrhage. The complexity and interest in this pathology have led to various classifications, with Radkowski's being the most accurate. Several approaches for angiofibroma intervention exist, including endoscopic approaches like lateral rhinotomy and transpalatal, and open approaches like LeFort Type 1 osteotomy, Weber Ferguson extraoral approach, and intraoral Degloving approach. Each should be selected according to the pathology's extent and severity. This article presents a clinical case addressed by the intraoral Degloving technique and a literature review on the diagnosis, classification, and management of JNA.

* Corresponding author: John Esteban Verdugo Parra

Objective

The objective of this article is to present a clinical case of long-evolving juvenile nasopharyngeal angiofibroma treated with the Degloving approach and to review the literature on this pathology, including its history, pathophysiology, staging, classification, treatment, and recurrences.

2. Literature review

2.1. History

Described in the 5th century by Hippocrates in relation to nasal polyps, it was in 1847 that Chelius identified its association with puberty, and in 1940 Friederig coined the term angiofibroma.

2.2. Definition

Nasopharyngeal angiofibroma is a rare tumor, representing 0.5%-0.05% of head and neck tumors, with benign mesenchymal and vascular histological qualities. Although benign, it is often aggressive and destructive, invading surrounding tissues and sometimes bone. These characteristics are due to its rich vascularization and lack of encapsulation.

2.3. Etiology and Pathophysiology

Juvenile nasopharyngeal angiofibroma is almost universally present in the second decade of life, leading the World Health Organization to rename it juvenile angiofibroma. They are more common in males, suggesting androgen dependence. This explains the relationship with hormonal flow during male puberty. Theories propose an embryonic origin from the occipital cartilage plate, or an abnormal periosteal response in nasopharyngeal bones to an ectopic hamartomatous vascular nest. Its most common origin is the sphenopalatine foramen, spreading locally to the pterygopalatine fossa (affected in 89%), nasal cavity, and paranasal sinuses through areas of least resistance. It generally extends to the nasal cavity and nasopharynx through the sphenopalatine foramen, spreads to the maxillary (43%) and ethmoidal (35%) sinuses, and reaches the infratemporal fossa via the pterygomaxillary fissure. Through the inferior orbital fissure, it reaches the greater wing of the sphenoid or sphenoid sinus (affected in 61% through the nasopharyngeal roof), eventually reaching the cranial fossa (5-20%). Extrapahryngeal angiofibromas have been sporadically described in the literature, most commonly located in the maxillary sinus. Unlike nasopharyngeal ones, these lesions are detected early, have less vascularization, and occur in older patients.

2.4. Histology

Histologically, it has two components: a vascular component and a stroma rich in collagen fibers, fibroblasts, and mast cells, with edema, infiltration, and necrotic areas at the periphery. The vascular component is characterized by thin walls, lack of endothelium, and absence of elastic fibers. The deeper vessels are larger with multiple anastomoses, resembling a cavernous hemorrhagic hemangioma.

2.5. Tumor Classification and Staging

Several classification types have been described, but the most commonly used are Andrews' staging and Chandler and Radkowski's classification systems (Table 1).

Table 1 Classification of Juvenile Nasopharyngeal Angiofibroma by Topography according to Authors: Andrews, Radowski, Fish, and Chandler

Stage	ANDREWS (1989)	RADKOWSKI (1996)		FISH (1983)	CHANDLER (1984)
I	Limited to the nose and nasopharynx	A	Limited to the nose and/or nasopharynx	Tumor limited to the nasopharynx and/or nasal cavity without bone invasion.	Tumor localized in the nasopharynx.
		B	Extension into one or more sinuses		
II	Invasion of the pterygopalatine or maxillary fossa,	A	Minimal extension into the pterygopalatine fossa	Tumor invading the pterygomaxillary fossa or the maxillary,	Tumor extends to the nasal fossa and/or sphenoidal sinus.

	ethmoidal and sphenoidal sinuses, with bone destruction		B	Complete occupation of the pterygopalatine fossa with or without erosion of the orbital bones	ethmoidal, or sphenoidal sinus with bone destruction.		
			C	Posterior invasion of the sphenoid wings			
III	A	Invasion of the infratemporal fossa or orbital region without intracranial involvement	A	Erosion of the skull base with minimal intracranial extension	A	Tumor invading the infratemporal fossa or orbit without intracranial invasion.	Tumor extends to one or more of the following structures: maxillary sinus, ethmoidal sinus, pterygomaxillary fossa, infratemporal fossa, orbit, and/or cheek.
	B	Invasion of the infratemporal fossa or orbital region with extradural involvement	B	Erosion of the skull base with extensive intracranial extension with or without invasion of the cavernous sinus	B	Tumor with extradural intracranial invasion.	
IV	A	Intracranial intradural involvement without infiltration of the cavernous sinus, pituitary, or optic chiasm			A	Tumor with extradural and intradural intracranial invasion without invasion of the optic nerve, sella turcica, or cavernous sinus.	Tumor invades the cranial cavity 1.
	B	Intracranial intradural involvement with infiltration of the cavernous sinus, pituitary, or optic chiasm			B	Tumor with extradural and intradural intracranial invasion with invasion of the optic nerve, sella turcica, or cavernous sinus.	

2.6. Genetic and Macroscopic Analysis

Genetically, a deletion of chromosome 17 has been found, which includes the p53 suppressor gene and the Her-2/neu oncogene.

Macroscopically, we find a non-encapsulated mass, with a reddish-wine, pale, or whitish coloration, covered by nasopharyngeal mucosa.

2.7. Clinical Presentation

Initially, symptoms include unilateral nasal obstruction and unilateral nasal bleeding that is unprovoked (as soon as the tumor occupies the nasopharynx). As the tumor grows, the bleeding becomes more frequent and bilateral, along with obstructive sleep apnea and rhinophonia. At this stage, it is common for the patient to have conductive hearing loss due to Eustachian tube obstruction caused by tumor growth.

2.8. Diagnosis

An adequate diagnosis is based on a thorough clinical history, otorhinolaryngological physical examination, and imaging studies. Biopsy is contraindicated in this pathology due to the high risk of bleeding. The radiological examination of

choice is contrast-enhanced computed tomography of the nose and paranasal sinuses. Gadolinium-enhanced magnetic resonance imaging can also be used. Imaging diagnosis is based on three factors: site of origin, growth pattern, and vascularity after contrast. Angiography serves as a diagnostic test to identify feeding arteries and as a therapeutic tool for presurgical embolization. Differential diagnosis is based on entities that produce a mass effect at the nasal and nasopharyngeal level, such as antrochoanal polyps, Killian's polyp, hypertrophic adenoids, malignant tumors like sarcomas, which are accompanied by lymphadenopathy, infiltrative and erosive characteristics, and ulcerated and fetid masses.

2.9. Treatment

Initially, treatment was based on surgical excision, either by endoscopic or open surgical approach. Various endoscopic approaches exist (lateral rhinotomy, transpalatal); for open approaches, those that expose the mid-facial third are indicated, such as LeFort Type I osteotomy, Weber Ferguson extraoral approach, and intraoral Degloving approach. LeFort Type I osteotomy allows a "panoramic" view of the posterior maxilla, maxillary sinus, and nasal cavity. Weber Ferguson approach is used when trauma or pathology extends beyond the infraorbital rim or the malar region. Degloving approach is used when the procedure does not exceed these areas. The chosen modality depends on size and extent, as well as the resources and instruments available at the hospital and its personnel. Minimally invasive endoscopic approaches are currently used, although endoscopic approaches have advantages such as less bleeding and shorter hospital stays. However, they have limited value due to the number of patients.

2.10. Measures to Reduce Bleeding During Surgery

Preoperative embolization is an essential step in treatment, demonstrating reduced formation of collateral circulation and hence the probability of massive hemorrhage. The inverse Trendelenburg position of 15 to 30 degrees diverts blood from the surgical field due to gravity and facilitates surgical exposure.

2.11. Recurrences

Recurrence is the clinical and radiological presence after six weeks of treatment. The recurrence rate varies between 5% and 37%, especially when the tumor invades the bone structure at the skull base.

3. Clinical case presentation

The following describes a clinical case of juvenile nasopharyngeal angiofibroma which, due to its extent, was approached using the Degloving technique.

An 18-year-old male patient presented to otorhinolaryngology consultation with progressive respiratory difficulty of 2 years' evolution, nasal obstruction, and recurrent minor epistaxis not requiring treatment. Physical examination via rhinoscopy revealed a violaceous mass in the right nasal cavity and purulent discharge from the same, along with left septal deviation. Cardio-thoracic, abdominal, and extremities examination showed no abnormalities. Neurological examination revealed an alert patient with no alterations in higher functions, preserved superficial and deep sensitivity, and normal reflexes. Fibroscopy identified a solitary mass in the right nasal cavity, characterized by its hard consistency and violaceous color, also extending to the left nasal cavity. A simple and contrast-enhanced CT scan of the facial massif revealed an extensive area of soft tissue occupying the entire right maxillary sinus and nasal cavities, causing bone destruction, with dimensions of 45x68 mm and slightly heterogeneous. This was complemented with angiotomography, showing an infiltrating mass in the right nasopharynx, causing adjacent bone destruction and invasion of the anterior ethmoidal cells and sphenoid sinus, with probable origin at the sphenopalatine foramen, obstructing the nasopharynx. The common carotid arteries were of adequate caliber and trajectory. The internal, external, and vertebral carotid arteries were of normal trajectory and caliber, with the arterial vessel feeding the lesion, apparently the internal maxillary artery and, to a lesser extent, the facial artery, being difficult to visualize. These findings suggest a presumptive diagnosis of JNA.

According to the Radkowski Classification System, due to the extent of the tumor in this case, it corresponds to Stage IIB; Stage III according to Chandler, and Stage II according to Fish. Pre-surgical examinations and cardiological evaluation were requested. Surgery was planned for the ligation of the right external carotid artery, which was performed without complications the day before tumor extraction.

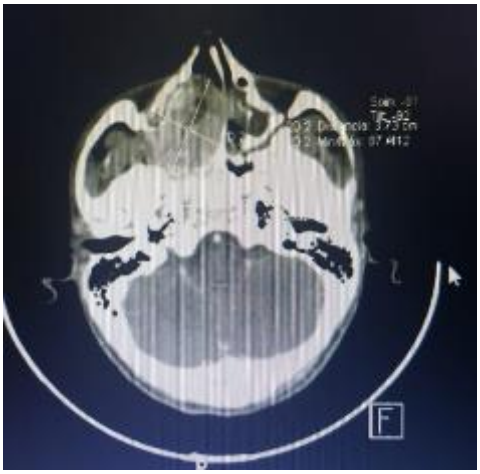

Figure 1 Axial slice of contrast-enhanced computed tomography showing an occupying mass in the left nasal cavity and maxillary sinus**.**

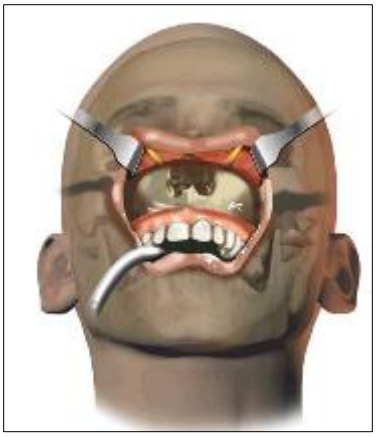

Figure 2 Anterior Craniofacial Resection/Midfacial Degloving [Internet]. 2024. Available from: https://entokey.com/anterior-craniofacial-resection-midfacial-degloving/. Accessed 2024 Jun 11.

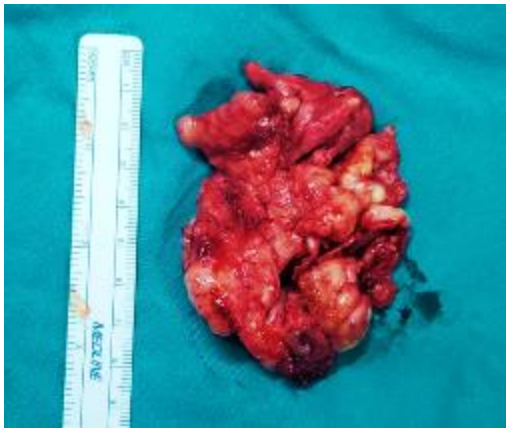

Figure 3 Anatomical specimen obtained.

A Degloving approach was performed. Due to the extent of the lesion, a wide exposure of the right mid-third of the face was achieved. An anterior, medial, and posterior maxillectomy was performed. The internal maxillary artery was located at the right pterygopalatine fossa, followed by a right inferior turbinectomy and resection of the anterior surface of the sphenoid, allowing for the extraction of the whitish tumor adherent to the nasopharyngeal mucosa, measuring approximately 8x9 cm. The histopathological study confirmed JNA as the definitive diagnosis.

After the extraction of the anatomical specimen, a sample was sent for histopathological examination. The histopathological findings were compatible with JNA as indicated by the laboratory report (Figure 3).

Figure 4 Histopathological examination of the lesion sent as a sample.

A new CT scan of the facial massif was requested one year after the surgery. The patient recovered without complications, and periodic check-ups confirmed the absence of any mass in the nasal cavities.

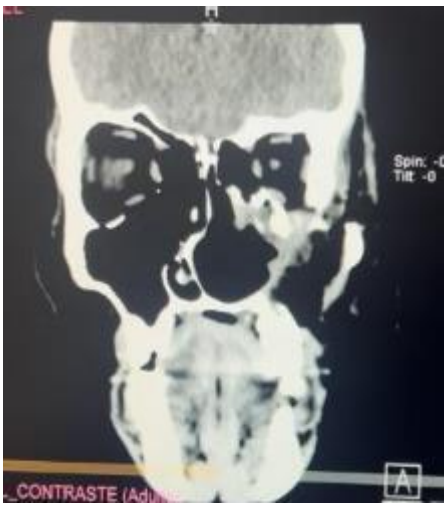

Figure 5 Coronal slice of contrast-enhanced computed tomography showing absence of lesion.

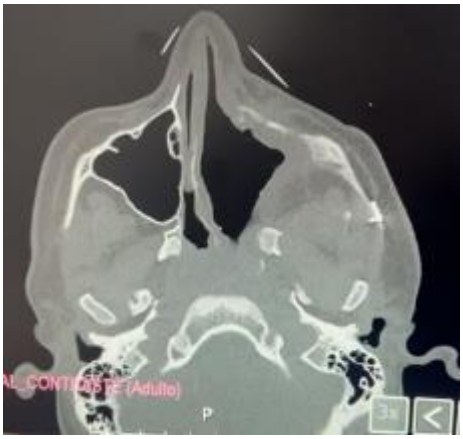

Figure 6 Axial slice of contrast-enhanced computed tomography showing absence of lesion.

4. Discussion

Juvenile nasopharyngeal angiofibroma being a highly vascularized tumor with a high likelihood of bleeding, should always be treated interdisciplinarily, typically involving both an otorhinolaryngologist and a maxillofacial surgeon. The early diagnosis of this type of tumor often presents a challenge. It is frequently diagnosed in stages II or III of Radkowski's classification, as observed in the presented case, which was classified as stage IIB due to the complete occupation of the pterygopalatine fossa without evidence of orbital bone erosion.

The use of embolization and the inverse Trendelenburg position significantly aid in surgical resection, as they can reduce both the time and difficulty of the surgery. Various surgical techniques for approaching this pathology have been described in the literature. The selection of each technique depends on the severity and extent of the lesion, but care must always be taken with the chosen surgical technique due to the recurrence rate of this pathology, especially if it extends to areas such as the skull base.

In any case where this pathology is present, it is always necessary and important to monitor for possible recurrence with imaging studies for several years after the surgical procedure.

5. Conclusion

juvenile nasopharyngeal angiofibroma despite being a benign tumor, can also be very invasive, and if not correctly diagnosed and treated, it can lead to serious intraoperative and postoperative complications. The choice of approach always depends on the extent and severity as determined by the classification of this pathology. Regardless of the chosen approach, postoperative care and monitoring must always be carried out to ensure the success of the procedure. Accurate clinical and imaging diagnosis in this and all pathologies will always lead to correct management and treatment by the involved professionals.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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