Annals of Clinical Case Reports

Case Report
Published: 16 Mar, 2024

# Concomitant Juvenile Nasopharyngeal Angiofibroma and Moyamoya Disease: A Rare Clinical Confluence

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## Abstract

Juvenile Nasopharyngeal Angiofibroma (JNA) is a rare and invasive tumor primarily affecting adolescent males. This case report presents a unique scenario of a 15-year-old adolescent diagnosed with JNA concurrently with Moyamoya disease, a cerebrovascular disorder characterized by abnormal brain vascular networks. The patient experienced persistent nasal obstruction, anosmia, and recurrent episodes of epistaxis for 2.5 years which progressively worsened. Clinical examination revealed a prominent pink/red mass in the nasopharynx, and imaging confirmed a sizable mass in the sphenopalatine foramen, causing luminal obliteration and extending into nearby structures. The staging of the JNA indicated stage IIIa according to the Andrews-Fisch system. Additionally, the patient was found to have advanced Moyamoya disease (Suzuki stage VI) through cerebral angiography, showing complete occlusion of both internal carotid arteries and proximal segments of the anterior and middle carotid arteries. Due to the high stroke risk associated with Moyamoya disease and the presence of collateral blood supply, embolization as a preoperative intervention was not feasible. Unfortunately, delayed diagnosis and limited resources made surgical resection for JNA and revascularization for Moyamoya disease impractical. This case highlights the diagnostic complexities and resource limitations encountered when managing the coexistence of JNA and Moyamoya disease, underscoring the importance of early recognition, interdisciplinary collaboration, and optimal resource allocation in challenging clinical scenarios. The rarity of this coexisting condition calls for increased awareness and understanding among clinicians to make timely and informed decisions, ultimately improving patient outcomes. Further research and multidisciplinary collaboration are needed to develop optimal management strategies for complex presentations like this, considering the impact of resource availability and regional limitations on treatment options and outcomes.

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**Received Date**: *26 Feb 2024*
**Accepted Date:** *12 Mar 2024*
**Published Date:** *16 Mar 2024*

***Citation:***
*Ovais H, Ahmed H, Ejaz AD, Malaika H, Maliha R, Preet I, et al. Concomitant Juvenile Nasopharyngeal Angiofibroma and Moyamoya Disease: A Rare Clinical Confluence. Ann Clin Case Rep. 2024; 9: 2594.*
***ISSN: 2474-1655.***



## Introduction

Juvenile nasopharyngeal angiofibroma is a rare condition predominantly affecting adolescent males. It is a highly vascular, benign and locally aggressive neoplasm. These non-encapsulated growths typically originate near sphenopalatine foramen in the nasopharynx [1]. The typical triad of symptoms that patients with JNA present with, include a mass in the nasopharynx, unilateral nasal obstruction, and recurrent epistaxis [2].

Moyamoya disease, meaning puff of smoke, is an idiopathic cerebrovascular disorder characterized by unilateral or bilateral stenosis of an intracranial segment of the Internal Carotid Artery (ICA) and the proximal parts of the anterior and middle cerebral arteries. Chronic ischemia leads to the formation of abnormal vascular networks at the base of the brain. It is predominantly seen in children and most of the cases are reported in Asia and non-Caucasian regions [3]. Patients with moyamoya disease usually present with TIAs, stroke, headaches, intracranial hemorrhage, or neuropathy.

## Case Presentation

A 15-year-old male presented to the outpatient department of a tertiary care hospital, complaining of nasal obstruction and anosmia for over 1 year and recurrent episodes of epistaxis for over 2.5 years from the left side of the nose. The patient reported that the bleeding had progressively increased both in terms of volume and frequency over time. He reported receiving two pints of blood due to severe episodes of bleeding. There was no history of fever, rash, nasal discharge, dyspnea, weight loss, stroke, hemorrhage, or focal neurological deficits. Clinical examination indicated a slight septal

deviation on the right side with profuse purulent nasal discharge that was not unpleasant smelling. A pink/red mass filled the nasopharynx, as seen by the posterior rhinoscopy. Laboratory investigations conducted on the patient revealed a hemoglobin level of 12.7 g/dl (14.0-17.4 normal range), hematocrit was 41.4% (42-58), and mean corpuscular hemoglobin was 24.2 pg (27.0-32.0). Contrast-Enhanced CT scan (CECT) of the head and neck revealed a mass in the posterior aspect of the nasal cavity, which measured approximately 5.7 cm × 5.0 cm × 5.0 cm (AP × TS × CC). The mass was heterogeneously enhanced and centered within the sphenopalatine foramen, causing its expansion. Posteriorly, it extended into the nasopharynx, causing complete luminal obliteration. According to the Andrews-Fisch system, the patient has been diagnosed with stage IIIa nasopharyngeal angiofibroma [4]. The patient underwent cerebral angiography to perform preoperative embolization to prevent excessive blood loss during the surgical resection of the nasopharyngeal angiofibroma. The angiogram revealed complete occlusion of the bilateral Internal Carotid Artery (ICA), as well as proximal segments of the Anterior Carotid (ACA) and Middle Carotid Artery (MCA). There were multiple extensive serpiginous collateral vessels identified that were supplying distal segments and cortical branches of ACA and MCA territories. A dominant collateral supply is seen on vertebral angiogram through multiple distal branches of bilateral posterior cerebral arteries. According to the Suzuki Grading System, these findings demonstrate advanced Moyamoya disease stage VI [5]. Hence, due to the presence of Moyamoya disease with ECA to ICA shunt, embolization was not performed to avoid the high risk of stroke.

## Discussion

Exclusively affecting males from the age group 5 to 25 years, Juvenile Nasal Angiofibroma (JNA) is a rare, progressive, age and sex-linked disease mostly seen in adolescent males. The tumor grows extensively, causing proptosis, cranial neuropathy, diplopia with speech disturbance, and conductive hearing loss [6]. Our patient presented with three symptoms; a mass in the nasopharynx, recurrent epistaxis, and nasal obstruction over a period of 2.5 years. Angiography, MR imaging, and CT scans can all be used to diagnose angiofibroma. The most crucial preoperative test is a Computed Tomography (CT) scan Additionally, CT can be used to detect sphenoid invasion and choose how aggressively to operate. CECT findings in our patient show the tumor's aggressive development and osseous erosion led to extension into the frontal and middle cranial fossae involving both the sphenoid sinuses and other high-value regions, complicating therapy which is consistent with the previous literature [7]. The preferred and recommended course of treatment is surgical resection of the tumor. Many patients receive preoperative embolization due to the extensive vascularity of the tumor to reduce the risk of intraoperative hemorrhage and associated complications. In the case of advanced tumors that involve critical regions or those with intracranial extension that may not be resectable, radiation therapy can be used as adjuvant therapy for residual or recurrent illness [8].

During the angiography procedure intended for embolization, our patient's imaging results were unexpectedly indicative of Moya disease, with complete occlusion of both internal carotid arteries and the proximal segments of the anterior and middle cerebral arteries. On our patient, none of the surgical procedures were performed, neither for JNA nor for MMD. The Tumor Board at Jinnah Post Medical Centre unanimously agreed that embolization and any surgical intervention could not be done due to the elevated risk of stroke due to the presence of ECA to ICA shunt associated with MMD. Hence, surgical resection of the Juvenile Nasopharyngeal Angiofibroma (JNA) was not feasible without prior embolization due to the high risk of blood loss during surgery. The patient was kept on palliative care along with beta-blockers to control the episodes of bleeding. The patient passed away after a few months.

This case report highlights the occurrence of two rare diseases presenting concurrently in a young individual. It also shows the challenges encountered while managing the patient with these diseases, underscoring the need for further research in the treatment of nasopharyngeal angiofibroma. The current therapeutic approach, involving surgical intervention along with embolization [9], was precluded because of the potential risk of stroke.

## Conclusion

Juvenile Nasopharyngeal Angiofibroma (JNA) concomitant with Moyamoya disease represents an exceedingly rare occurrence. JNA, a locally aggressive and highly vascular tumor predominantly affecting adolescent males, Comprehensive evaluation through Contrast Enhanced CT (CECT) of the head and neck revealed the presence of a mass in the nasal cavity. The standard therapeutic approach involves a combination of angiography-guided preoperative embolization and subsequent surgical resection. However, the angiography procedure unexpectedly unveiled the diagnosis of Moyamoya disease, introducing an unanticipated twist to the case.

## References

1. Lloyd G, Howard D, Phelps P, Cheesman A. Juvenile angiofibroma: the lessons of 20 years of modern imaging. J Laryngol Otol. 1999;113(2):127-34.
2. Enepekides DJ. Recent advances in the treatment of juvenile angiofibroma. Curr Opin Otolaryngol Head Neck Surg. 2004;12(6):495-9.
3. Natori Y, Ikezaki K, Matsushima T, Fukui M. Angiographic moyamoya 'its definition, classification, and therapy. Clin Neurol Neurosurg. 1997;99:S168-72.
4. Gupta R, Agarwal SP. Juvenile nasopharyngeal angiofibroma: Combined approach for excision, transpalatal and endoscopic; a new perspective. Indian J Otolaryngol Head Neck Surg. 2018;70:125-9.
5. Rosi A, Riordan CP, Smith ER, Scott RM, Orbach DB. Clinical status and evolution in moyamoya: which angiographic findings correlate? Brain Commun. 2019;1(1):fcz029.
6. Gupta S, Gupta S, Ghosh S, Narang P. Juvenile nasopharyngeal angiofibroma: Case report with review on role of imaging in diagnosis. Contemp Clin Dent. 2015;6(1):98-102.
7. Tork CA, Simpson DL. Nasopharyngeal angiofibroma. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2024.
8. Ko C, Citrin D. Radiotherapy for the management of locally advanced squamous cell carcinoma of the head and neck. Oral Dis. 2009;15(2):121-32.
9. Maroda AJ, Beckmann NA, Sheyn AM, Elijovich L, Michael LM, DiNitto JM, et al. Trimodal embolization of juvenile nasopharyngeal angiofibroma with intracranial extension. Int J Pediatr Otorhinolaryngol. 2020;130:109805.