



Rare Atypical Radiological Appearance of Nasopharyngeal Angiofibroma: A Diagnostic Difficulty

Authors

Singh Rakesh Kumar^{1*}, Sinha Richi², Mishra Sarita Kumari³

Department of Otorhinolaryngology, Indira Gandhi Institute of Medical Sciences, Patna, India

*Corresponding Author

Singh Rakesh Kumar

Professor and Head, Department of Otorhinolaryngology, Indira Gandhi Institute of Medical Sciences,
Sheikhpura, Patna, India, Zip code: 800014

Email: rkstent5@gmail.com, Mobile: +91 7781912005

Abstract

Background: Juvenile angiofibroma is an uncommon, benign and extremely vascular tumor that presents as recurrent severe epistaxis with or without nasal obstruction in adolescent males. It is believed to arise from the posterolateral wall of the nasal cavity at the level of the sphenopalatine foramen. This typically produces characteristic radiological appearance that often renders biopsy unnecessary. However absence of classical radiological features can lead to misdiagnosis and inadvertent life threatening intraoperative complications.

Purpose: To highlight the importance of absence of classical radiological presentation of Juvenile nasopharyngeal angiofibroma and add to the current knowledge of diagnosis and management of this crucial disease.

Result: We came across 15 unique cases of nasopharyngeal angiofibroma with atypical radiological appearance amongst 34 cases over a period of 4 years. All of these cases had absence of nearly all the classical radiological findings. Only one such case was found reported on searching the available English literature on pubmed.

Conclusion: With strong clinical suspicion even in the absence of classical radiological signs a differential diagnosis of JNA should be considered when diagnosing any nasal or nasopharyngeal mass owing to the risk of associated life threatening hemorrhage. Furthermore, the absence of classical radiological signs emphasize the need for further research on the site of origin.

1. Introduction

Juvenile nasopharyngeal angiofibroma (JNA) is the most common, benign and extremely vascular tumour of nasopharynx. However, it accounts for merely 0.5 percent of all head and neck tumours. Most authors believe it to arise from the posterolateral wall of nasal cavity at the level of sphenopalatine foramen^[1]. Rarely, it is found at

other sites in the nasal cavity and paranasal sinuses. It is locally invasive, as it grows, the tumour extends into the nasopharynx, paranasal sinuses, pterygopalatine and infratemporal fossa. Larger tumors can involve the orbit and cavernous sinus. It typically affects adolescent males presenting with profuse, recurrent epistaxis and nasal obstruction. The diagnosis can be further confirmed

by its characteristic radiological appearance that often renders biopsy unnecessary. On CT and MRI the diagnosis of JNA is based on the following three characteristic features: the area of origin invariably located at the sphenopalatine foramen, widening of the pterygopalatine fossa and anterior bowing of the posterior maxillary sinus wall known as the antral sign^[2]. However, we came across a significant number of cases of angiofibroma with atypical radiological appearance that can mimic lesions other than angiofibroma like infected sinonasal polyp. Hence, absence of these signs can lead to misdiagnosis and inadvertent life threatening intraoperative hemorrhage. We analyzed all the cases of nasopharyngeal angiofibroma and highlight the importance of knowledge of atypical radiological appearance in the management of this crucial disease.

2. Patients and Methods

We retrospectively evaluated 34 operated and histopathologically confirmed cases of nasopharyngeal angiofibroma over a period of 4 years in the Department of Otorhinolaryngology of Indira Gandhi Institute of Medical Sciences. A note of detailed history and thorough clinical examination was made. The hematological and radiological profiles of the patients were studied. All patients had undergone contrast enhanced computed tomography of the paranasal sinuses. Surgical re-

moval of mass was accomplished in all the cases under general anesthesia without any preoperative embolization by open approaches followed by histopathological confirmation of the disease. An intraoperative note of site of origin of these tumors was also made.

3. Results

A total number of 34 cases of nasopharyngeal angiofibroma were evaluated. The clinical and radiological findings of the cases are summarized in Table 1 and 2 respectively. Out of total 34 cases, 15 (41.1%) cases showed absence of classical radiological features on CT (Fig 1). Table 3 summarizes the intraoperative site of origin of these cases. Based on our analysis and a strong possibility of missing the diagnosis due to absence of these classical radiological features, we formulated a protocol to diagnose angiofibroma even with slightest possibility of its presence (Fig.2)

Table 1: Summary of the clinical findings of the cases

Symptoms (n=34)	present	absent
recurrent profuse epistaxis	34	0
nasal obstruction	34	
-unilateral	21	0
-bilateral	13	
nasal endoscopy (fleshy pale to purple mass in nasal cavity and nasopharynx)	34	0

Table 2: Summary of the radiological findings of the cases

Classical Sign	presesnt (no. of cases)	absent (no. of cases)
Widening of sphenopalatine foramen	20	14
Widening of pterygopalatine fossa	21	13
Pterygoid plate erosion	21	13
Antral sign	19	15
Crescent sign	5	29

Table 3: Summary of the intraoperative site of origin

site of origin	no.of cases
posterolateral nasal wall	19
posterior nasal septum	11
nasopharyngeal vault	2
adjacent to torus tubaris	1
vidian canal	1

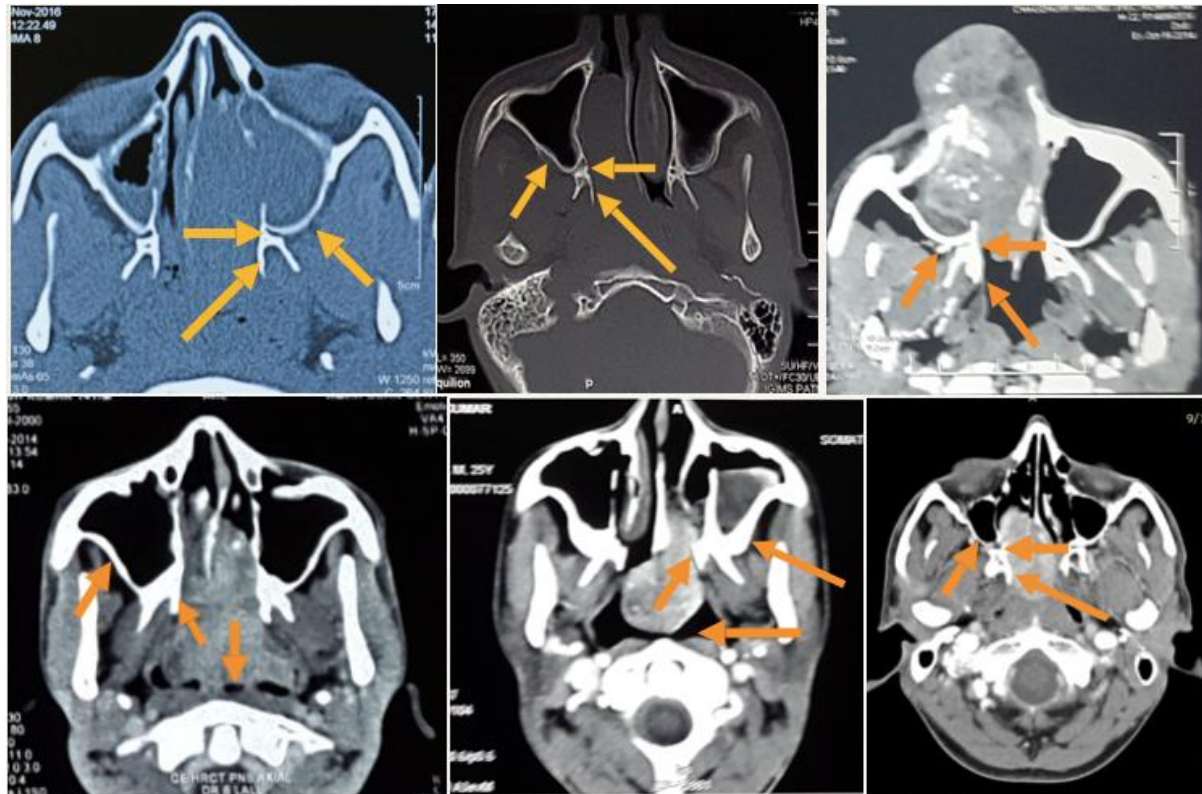


Figure 1: showing absence of classical radiological features on computed tomography of some of the cases

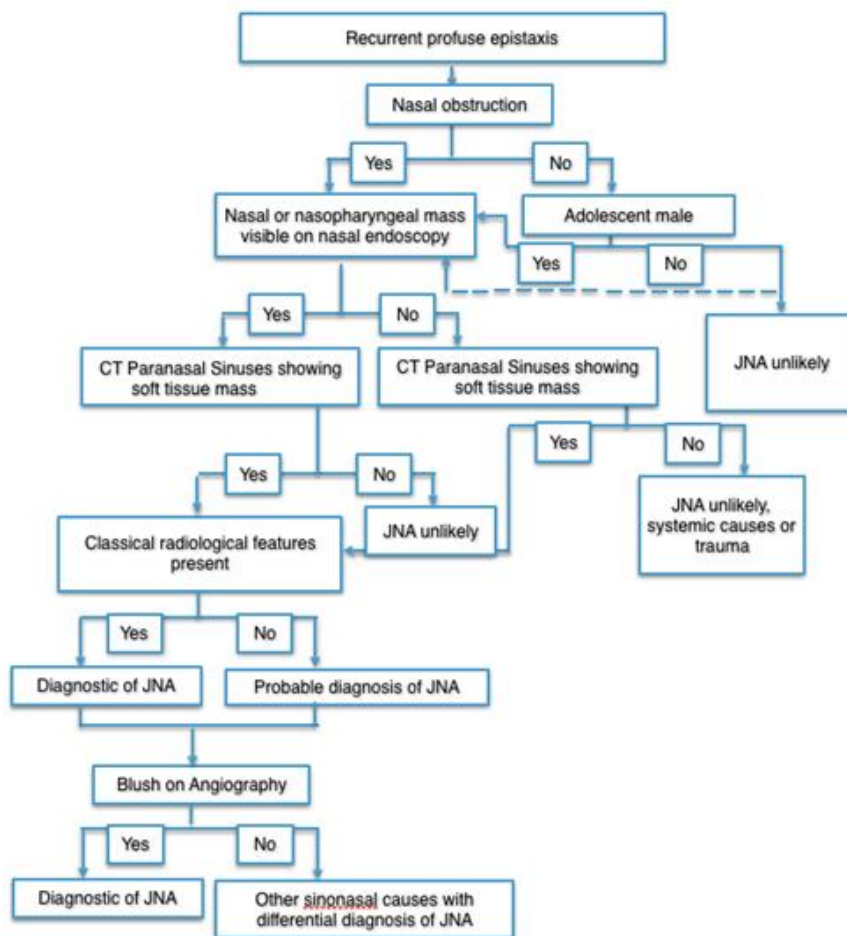


Figure 2: showing algorithm for diagnosis of juvenile nasopharyngeal angiofibroma.

4. Discussion

4.1 Background

JNA was first described in conjunction with nasal polyps by Hippocrates in the 5th century BC^[2]. Initially regarded as a fibrous nasal polyp at that time, the term “angiofibroma” was not coined until Friedberg did so in 1940^[2]. Juvenile nasopharyngeal angiofibroma is pathognomonically characterized as a benign highly vascular tumor located in the posterior nasal cavity and nasopharynx of adolescent males. The exact site of origin is controversial. Most authors believe it to arise from the posterolateral nasal wall at the level of sphenopalatine foramen. JNAs typically arise from the sphenopalatine artery, which is a terminal branch of the internal maxillary artery^[3,4]. Although not a malignant process, it is known for its locally invasive spread with progressive growth. This is associated with a significant degree of morbidity commonly related to either intracranial extension or massive hemorrhage. Thus, it becomes necessary to err on the side of caution while diagnosing any nasopharyngeal or nasal mass.

4.2 Clinical Features

The classical clinical presentation of juvenile angiofibroma is recurrent severe epistaxis associated with progressive nasal obstruction in almost exclusively adolescent males. Anterior rhinoscopy may show mass in the nasal cavity often covered with mucopurulent secretions. There may be bulge in the soft palate due to the bulk of the tumor which can be seen clearly as a pink or reddish mass that fills the nasopharynx. This mass effect also sometimes results in atypical symptoms including blockage of the eustachian tubes with middle ear effusions and conductive hearing loss, rhinolalia clausa, hyposmia or anosmia.

4.3 Histopathology

Microscopically, tumor consists of proliferating, irregular vascular channels within a fibrous stroma. The stromal compartment is made up of plump cells that can be spindle or stellate in shape

and give rise to varying amounts of collagen. It is this that makes some tumors very hard or firm. Tumour blood vessels typically lack smooth muscle and elastic fibers, this feature contributing to its character for sustained bleeding and for massive hemorrhage following even negligible manipulation^[5]. Therefore, one must steer clear of biopsy in such cases.

4.4 Radiology

The other diagnostic modality is radiology. Contrast enhanced CT is the gold standard diagnostic modality and remains as a widely used tool for diagnosing as well as defining the extent of angiofibroma. Magnetic resonance imaging (MRI) provides further assessment of the relationship of tumor with adjacent soft tissue and delineates its intracranial and cavernous sinus extension. It is also particularly useful to assess any residual or recurrent tumor. Digital Subtraction Angiography is critical in the evaluation of feeding vessels and allows for pre-operative embolization of JNAs.

Based upon its commonest site of origin from the level of sphenopalatine foramen, CT shows certain characteristic features:

(1) a mass in the posterior nasal cavity and/or nasopharynx, pterygopalatine fossa; (2) erosion of bone behind the sphenopalatine foramen and medial pterygoid plate; (3) anterior bowing of the posterior maxillary wall on axial CT slices known as the Antral or Holman-Miller sign^[1,2,6]. Lloyd et al investigated Seventy-two patients with juvenile angiofibroma by computerized tomography (CT) over a period of 20 years and provided evidence in support of these findings^[7]. As it grows, the tumor extends into the nasopharynx, paranasal sinuses, pterygopalatine and infra-temporal fossa. Larger tumors can involve the orbit and cavernous sinus. However, isolate extranasopharyngeal angiofibroma is rare. The commonest extra nasopharyngeal site of angiofibroma is maxillary sinus followed by ethmoid sinus and sphenoid sinuses. Angiofibromas at such a location seldom follow the classical radiological features^[8-10]. On the contrary, considering the fact that posterolateral nasal

wall at the level of sphenopalatine foramen is currently the most accepted site of origin, we found paucity of all the classical imaging signs in our cases. A similar case was reported by Ordóñez et al. in 2008 where the CT scan did not reveal any erosion of the sphenopalatine foramen but the biopsy later reported to be a juvenile nasopharyngeal angiofibroma^[11]. Patrocínio et al. in 2005 also reported a similar nasopharyngeal angiofibroma in an elderly female but not much was described about its radiological aspect^[12]. Another atypical case reported by Ezzat et al in 2014 showed that the lesion was located in pneumatized pterygoid air cell extending into the right nasal cavity. Here as well there was no erosion of posterior osseous margin of the sphenopalatine foramen, Holman-Miller sign or widening of the sphenopalatine foramen. However, there was erosion of the base of the medial pterygoid plate^[13]. Hence, even in the early stages of the disease some involvement of sphenopalatine area is expected in the form of erosion or widening. Intra-operatively we found in our study that the tumor in these 15 atypical cases had other sites of attachment as mentioned earlier. This generates a need for further research on the site of origin. More importantly, in the absence of classical radiological signs in cases with high clinical suspicion, a differential diagnosis of angiofibroma should be considered and appropriate management should be planned.

4.5 Management

Surgery is considered the mainstay in the management of juvenile angiofibroma. Several approaches are currently available, ranging from microendoscopic techniques to mid-facial degloving and infra-temporal fossa resection. Independent of the technique selected, the key steps to minimizing bleeding and achieving radical resection are the dissection of the lesion in the subperiosteal plane with the help of bipolar coagulation and ligation of the feeder vessel prior to any manipulation of the mass. We religiously followed these two steps in all our cases and achieved complete surgical excision of the tumor

without any preceding embolization. Complete surgical extrication is vital to definitive treatment of JNA as an incomplete resection is the leading etiology for recurrence. Moreover, a complete knowledge of its various clinical and radiological presentations is crucial for diagnosis.

5. Conclusion

JNA is a highly vascular neoplasm of nasopharynx in adolescent males that typically presents with profuse recurrent epistaxis with or without progressive nasal obstruction. Though in majority of the cases the diagnosis is based on its key radiologic features, yet in their absence a differential diagnosis of JNA should be considered when diagnosing any nasal or nasopharyngeal mass owing to the risk of life threatening hemorrhage when not vigilant. Moreover, this emphasizes a need for further research on the site of origin of angiofibroma.

Source of Support: Nil

Conflict of Interest: None declared.

References

1. Garça MF, Yuca SA, Yuca K. Juvenile nasopharyngeal angiofibroma. *Eur J Gen Med* 2010;7(4):419-25.
2. Mishra S, Praveena NM, Panigrahi RG, Gupta YM. Imaging in the diagnosis of juvenile nasopharyngeal angiofibroma. *J Clin Imaging Sci* 2013;3:S1 (Available from: <http://www.clinicalimagingscience.org/text.asp?2013/3/2/1/109469>).
3. Rogers D, Hartnick C, Fagan J. JUVENILE NASOPHARYNGEAL ANGIOFIBROMA SURGERY. *The Open Access Atlas of Otolaryngology, Head & Neck Operative Surgery*. (Available from: www.entdev.uct.ac.za).
4. Manjunath M, Deepak TA, Mayank KC. Juvenile Nasopharyngeal Angiofibroma. *Journal of Indian Academy of Oral Medicine and Radiology* 2011;23(3):S408-11.

5. Gleeson M. Juvenile angiofibroma. In: Gilbert RW, Gleeson M, Scott-Brown WG. Scott-Brown's Otorhinolaryngology Head and Neck Surgery. Hodder Arnold 2008;187:2237-45.
6. Lloyd G, Howard D, Lund VJ, et al. Imaging for juvenile angiofibroma. J Laryngol Otol. 2000;114(9):727-30.
7. Lloyd G, Howard D, Phelps P, et al. Juvenile angiofibroma: the lessons of 20 years of modern imaging. J Laryngol Otol. 1999 Feb;113(2):127-34.
8. R Singh, S Bhandary, A Sundas, S Panday. Extra Nasopharyngeal Angiofibroma Of Anterior Ethmoid In A Middle Aged Woman. The Internet Journal of Head and Neck Surgery. 2006;1(1).
9. Malvi G, Manestar D, Krstulja M, Corak D, Candrljic B, Kujundzic M, et al. Juvenile Angiofibroma of the Maxillary Sinus, Coll. Antropol. 2012;36(Suppl. 2):197-9.
10. Rodriguez LG, Rudman K, Cogbill CH, Loehrl T, Poetker DM. Nasal septal angiofibroma, a subclass of extranasopharyngeal angiofibroma. American Journal of Otolaryngology 2013;34(2):133-41.
11. Ordóñez LEO, Hernández JCV, Rueda RS. Juvenile Nasopharyngeal Angiofibroma: An Atypical Case. Acta Otorrinolaringol Esp. 2008;59(8):424.
12. Patrocínio JA, Patrocínio LG, Borba BHC, Bonatti BDS, Guimarães AHB. Nasopharyngeal angiofibroma in an elderly woman. American Journal of Otolaryngology 2005;26(3):198-200.
13. Ezzat AEM, Roshdy HM, Eid MI. Case Report: Atypical Clinical and Radiological Presentation of Juvenile Nasopharyngeal Angiofibroma. IOSR Journal of Dental and Medical Sciences (IOSR-JDMS) 2014;13(10)Ver. I:105-8.