Juvenile Nasopharyngeal Angiofibroma: Case report with review on role of imaging in diagnosis

Shikha Gupta, Sunita Gupta, Sujoy Ghosh, Poonam Narang[1]

Abstract

Juvenile nasopharyngeal angiofibroma is a locally aggressive benign vascular neoplasm, composed of vasogenic and myofibroblastic elements, accounts for 0.05–0.5% of all the head and neck neoplasms. There are very few case reports of nasopharyngeal angiofibroma involving the oral cavity; we report a case involving both the maxilla and mandible in a 17-year-old patient who reported with a large firm swelling on right side of face with recurrent epistaxis and headache. Magnetic resonance angiography revealed a large lobulated enhancing soft tissue mass, which was hypointense on T1-weighted image and heterogeneously hyperintense on T2-weighted image causing expansion of pterygopalatine fossa and sphenopalatine foramen with extension into the sphenoid sinus, ethmoid air cells, right nasal cavity, right infratemporal fossa and right maxillary sinus with remodeling of right zygomatic arch and part of body and ramus of mandible. It was supplied by the right external carotid artery. Patient was referred to the department of neurosurgery for further management. The diagnosis at an early stage is important because it is associated with high risk of morbidity, but advances in imaging, and surgical methods of treatment have changed the sites associated with high risk of morbidity.

Keywords: Angiography, computed tomography, juvenile nasopharyngeal angiofibroma, magnetic resonance imaging

Introduction

Hippocrates described this tumor in the 5th century BC, but Friedberg first used the term angiofibroma in 1940.[1] Juvenile nasopharyngeal angiofibroma (JNA) is a relatively rare tumor with an incidence of between 1:6000 and 1:55,000 of the population[2] and occur almost exclusively in male adolescent.[3] It invades the natural foramina and fissures usually present in the nasopharyngeal region.[4-6] The patients of nasopharyngeal angiofibroma presents with classical triad of epistaxis, unilateral nasal obstruction and a mass in the nasopharynx suggesting a diagnosis of nasopharyngeal angiofibroma and is supplemented by imaging.[3,7] Imaging such as computed tomography (CT), nuclear magnetic resonance, angiography and even nasal endoscopy clearly establishes the extent of tumor, its pattern of spread and consequently surgical planning.[8] Several classification methods based on CT, magnetic resonance imaging (MRI) and endoscopy have been employed to stage the tumor and assist in choosing the appropriate treatment. These include those developed by Radkowski (1996), Fisch (1983), Andrews (1989), Chandler (1984), although none is universally accepted.[9] But the classification by Fisch is mostly followed[3] [Table 1].

Here, we are reporting a case of JNA in a 17-year-old male presenting as an extraoral facial swelling with stage IIIb and reviews the pathogenesis and role of imaging in the diagnosis of the tumor.

Case Report

A 17-year-old male patient reported to the Outpatient Department of Maulana Azad Institute of Dental Sciences in July 2012 with the chief complaint of swelling on the right side of the face since 1 year that had rapidly increased in size since last 4–5 months. It was associated with occasional intermittent, spontaneous pain and continuous headache which aggravated on bending. Patient had a history of weight loss of over 15–16 kg with the loss of appetite over 1 year along with frequent epistaxis from the right nostril with feeling of stuffiness of nose and nasal discharge. There was a history of extraction of upper right second molar after the appearance of swelling.

The medical history was not significant. Physical examination along with cranial nerve examination did not reveal any abnormality. Extraoral examination [Figure 1] was suggestive of a lobular swelling on the right side of the

Department of Oral Medicine and Radiology, Maulana Azad Institute of Dental Science, [1]Department of Radio-Diagnosis, GB Pant Hospital, New Delhi, India

Correspondence: Dr. Shikha Gupta,
Department of Oral Medicine and Radiology, Maulana Azad Institute of Dental Sciences, New Delhi, India.
E-mail: Calmshikhs@yahoo.com

Access this article online	
Quick Response Code:	Website: www.contempclindent.org
	DOI: 10.4103/0976-237X.149301

face extending from infraorbital margin to the lower border of mandible supero-inferiorly and from ala of nose and corner of the mouth to tragus of ear and ramus of mandible anteroposteriorly approximately 7 cm × 8 cm in size. The overlying skin was normal in color and texture. The swelling was smooth surfaced with well-defined margins and resilient in consistency. It was compressible, pulsatile, and tender on palpation. The swelling reduced in size on opening the mouth and on clenching, swelling became more prominent adjacent to ramus of mandible. The mouth opening was reduced with deviation toward right on opening. The right submandibular lymph nodes were found to be enlarged, tender and mobile on palpation.

Intraoral examination [Figure 2] revealed generalized brownish discoloration of teeth in a linear fashion along the cervical third of teeth with high arched palate. The buccal mucosa on the right side was swollen and lobulated extending from the corner of the mouth to the ramus of mandible with indentations on its surface. An ulcer was present in relation to maxillary right first molar, which had everted margins and an erythematous surface about 1 cm in size. Based on history and clinical findings, a provisional diagnosis of benign soft tissue tumor (nonodontogenic) was given. However, benign tumor of the parotid gland, low grade malignancy of parotid gland and lymphoma were considered as the other differentials. Routine hematological investigations were found to be within the normal limits. Fine-needle aspiration of the swelling and the lymph node was performed, which revealed only blood.

Table 1: Fisch classification of JNA based on its extension in CT and MRI

Stage I	The tumor is limited to the sphenopalatine foramen, nasopharynx and nasal cavity without bone destruction
Stage II	The tumor invades the nasal sinuses or the pterygomaxillary fossa with bone destruction
Stage IIIa	The tumor invades the infratemporal fossa or orbit without intracranial involvement
Stage IIIb	The tumor invades the infratemporal fossa or orbit with intracranial and extradural involvement
Stage IVa	The tumor shows intracranial, extradural and/or intradural invasion, without invasion of optic nerve, sella, or cavernous sinus
Stage IVb	The tumor in stage IVa with invasion of optic nerve, sella and/or cavernous sinus

JNA: Juvenile nasopharyngeal angiofibroma; CT: Computed tomography; MRI: Magnetic resonance imaging

Orthopantomogram [Figure 3] showed erosion of the alveolar bone in relation to maxillary first molar region and maxillary tuberosity on the right side along with resorption of the anterior border of ramus and the effacement of right maxillary sinus, zygomatic arch and adjacent maxillary structures by the soft tissue mass without the presence of any well-defined lesion. All findings were suggestive of malignancy.

Contrast enhanced CT revealed a large, ill-defined enhancing soft tissue mass filling the nasopharynx, bilateral nasal cavity [Figures 4 and 5], and pterygopalatine fossa and sphenopalatine foramen extending into ethmoid and sphenoid sinus with erosion of its lateral wall and floor. There was an extension into soft tissue component into right buccal space and the infratemporal space [Figure 6] with associated mass effects giving an impression of nasopharyngeal angiofibroma.

Magnetic resonance imaging angiography showed similar findings with a large lobulated enhancing soft tissue mass measuring 5.4 cm × 8 cm × 8 cm in size in the right base of the skull adjacent to nasopharynx. The mass was seen as hypointense on T1-weighted image and heterogeneously hyperintense on T2-weighted image [Figure 7] causing expansion of pterygopalatine fossa and sphenopalatine foramen with extension into the sphenoid sinus, ethmoid

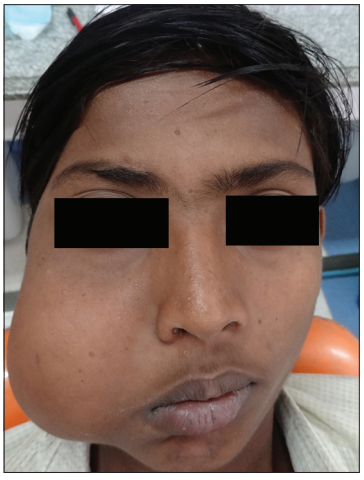

Figure 1: Extraoral photograph of the patient showing lobulated swelling on right side of face

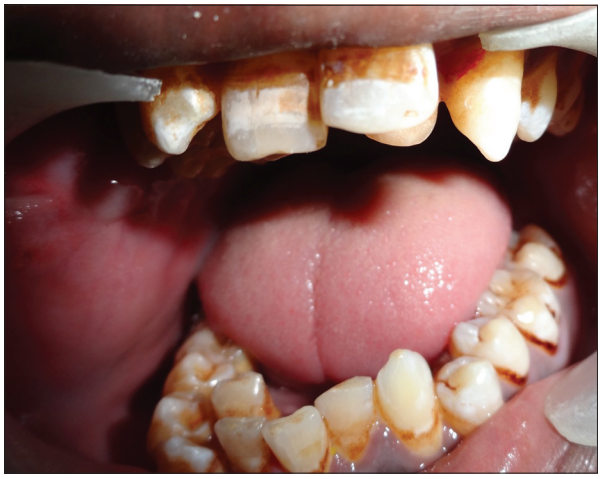

Figure 2: Intraoral photograph of the patient showing generalized brown discoloration of teeth lobulated surface of right buccal mucosa

air cells and right nasal cavity. Anteriorly the mass was seen to be extending into the right infratemporal fossa and right maxillary sinus with remodeling of right zygomatic arch and part of body and ramus of mandible with elevation of subcutaneous tissue of right cheek. The mass was supplied by the right external carotid artery [Figure 8].

Based on history, clinical presentation, CT and MRI findings, the lesion was finally diagnosed as nasopharyngeal angiofibroma. Patient was referred to the Department of Neurosurgery for further management where it was planned to carry out the initial embolization of the lesion to be followed by surgery.

Discussion

Angiofibroma is a relatively rare tumor. Only a few cases have been reported in female.[2,5,10,11] JNAs are age and sex linked. It affects almost exclusively male adolescents with median age of 15 years; raising suspicion about the role of sexual hormones in its pathogenesis.[9] They originate predominantly in the posterolateral wall of the nasopharynx, specifically at the trifurcation of the sphenoidal process of the palatine bone, the horizontal process of the vomer and the roof of the pterygoid process.[1] Patients usually present at late stage of the disease with typical complaints of nasal obstruction and recurrent epistaxis and rarely as swelling on face. Extensive growth of tumor may cause facial swelling, proptosis, diplopia with disturbance in speech and conductive hearing loss.[9] In the present case also, the patient was a male, 17 years old who presented with the right facial swelling along with the classical triad of recurrent epistaxis, nasal stuffiness and discharge.

Although the origin is still disputed, the medical literature reveals several consistent features like involvement of sphenopalatine foramen, erosion of the base of pterygoid plate; and secondary involvement of the nasopharynx.[2] In

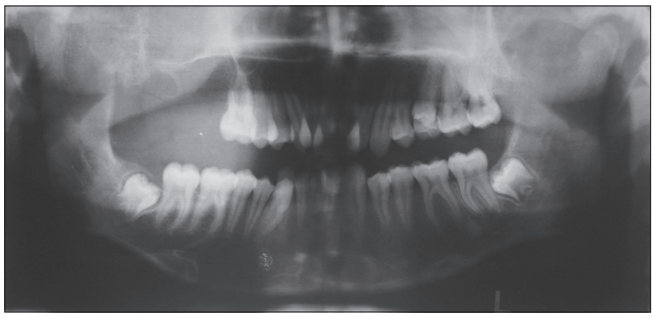

Figure 3: Orthopantomograph of the patient showing effacement of right maxillary alveolus, maxillary sinus, zygomatic arch and adjacent maxillary structures by the soft tissue mass with resorption of anterior border of ramus

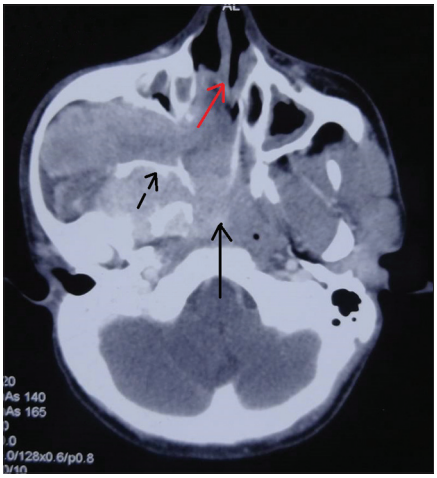

Figure 5: Axial computed tomography image showing ill-defined enhancing soft tissue mass filling nasopharynx (black arrow), bilateral nasal cavity with deviation of nasal septum (red arrow) and erosion of right medial pterygoid plate (black dashed arrow)

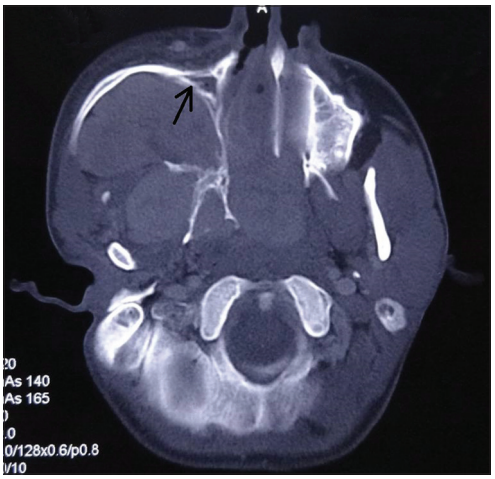

Figure 4: Axial computed tomography image showing bowing of anterior wall of right maxillary sinus (arrow) by the enhancing soft tissue mass known as Hollman Miller's sign

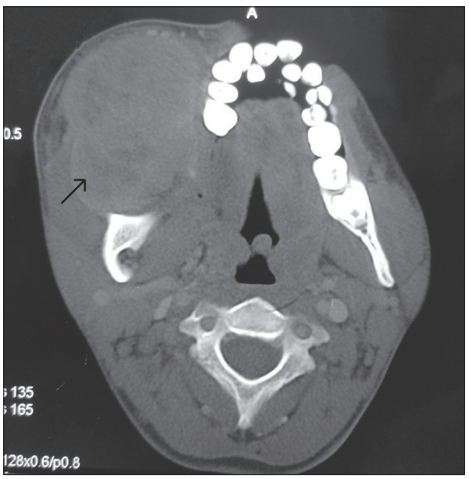

Figure 6: Axial computed tomography image showing extension of soft tissue component into right buccal space and infratemporal space (arrow) with remodeling of mandible

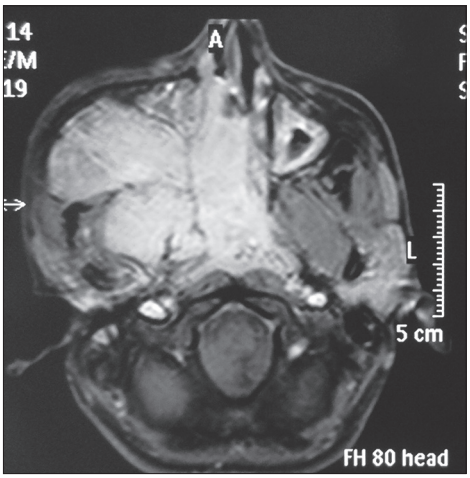

Figure 7: Axial T2-weighted magnetic resonance image showing hyperintense large lobulated enhancing soft tissue mass causing expansion of pterygopalatine fossa and sphenopalatine foramen with extension into sphenoid sinus, ethmoid air cells and right nasal cavity, maxillary sinus and infratemporal fossa

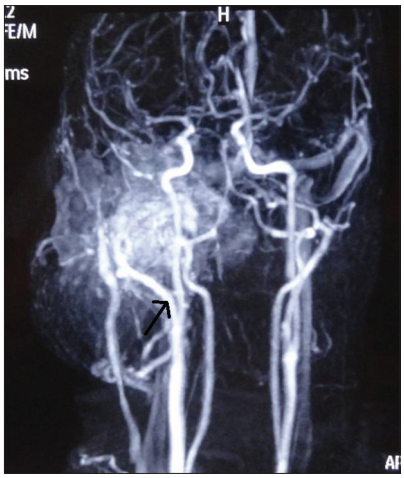

Figure 8: Magnetic resonance imaging angiogram showing the lesion to be supplied by external carotid artery (arrow)

separate studies, Brunner and Harrison found endothelial lined spaces in region of sphenopalatine foramen and base of pterygoid plates in male and female fetuses which suggests that JNA originates in a hamartomatous nidus of vascular tissue in the area of the sphenopalatine foramen that is stimulated by endogenous testosterone in early puberty.[2,12]

Lloyd *et al*.[13] reported three finding on CT and MRI imaging that should suggest a diagnosis of JNA:

1. A soft tissue mass in the nasopharynx or nasal cavity
2. A mass in the pterygopalatine fossa
3. Erosion of posterior osseous margin of the sphenopalatine foramen extending to the base of the medial pterygoid plate.

Angiofibroma can be diagnosed using CT, MRI, and magnetic resonance angiography.[1] CT is the most important preoperative test because it is useful in showing the destruction of bony structures and widening of foramen and fissures at the skull base.[1] Demonstration of the anterior bowing of the maxillary wall due to presence of a mass in the pterygomaxillary space on axial CT slices known as the Holman-Miller sign is a characteristic finding of JNA.[9] Moreover, tumor staging is done based on CT. MRI is useful to show the presence of intracranial extension of the tumor. MRI also helps discern between sinus invasion, obstruction and retention of secretions.[1] On MRI, JNA appears as a heterogeneous mass with signal voids that are consistent with the highly vascular tumor.[1] Selective angiography identifies the feeding vessels and allows for preoperative embolization for vascular control. It shows the size and site of lesion as well as the size and location of feeding vessel.[1,13] Vascularization arises most frequently from the external carotid branch of maxillary artery with background vascularization arising from blood vessels in the ascending pharyngeal artery and internal carotid artery.[4]

The preferred management includes surgery with preoperative embolization.[4] Other methods of treatment that have been employed are irradiation, hormone therapy, cryotherapy, arterial ligation, use of sclerosing agents.[2,4] Future therapy for JNA may include intra-arterial immunotherapy and the use of certain agents that inhibit angiogenesis.[2]

Conclusion

Significance of reporting the case lies in its involvement and extension into the oral cavity including mandible, which is rare and utilization of imaging as the sole modality to diagnose the lesion well in time.

References

1. Mehmet A, Orhan S, Suphi M. Juvenile nasopharyngeal angiofibroma: Radiological evaluation and pre-operative embolization. KBB Forum 2006;5:58-61.
2. McDaniel RK, Houston GD. Juvenile nasopharyngeal angiofibroma with lateral extension into the cheek: Report of case. J Oral Maxillofac Surg 1995;53:473-6.
3. Martins MB, Lima FV, Mendoca CA, Jesus EP. Nasopharyngeal Angiofibroma: Our experience and literature review. Int Arch Otorhinolaryngol 2013;17:14-9.
4. Dillard DG, Cohen C, Muller S, Del Gaudio J, Reichman O, Parrish B, *et al.* Immunolocalization of activated transforming growth factor beta1 in juvenile nasopharyngeal angiofibroma. Arch Otolaryngol Head Neck Surg 2000;126:723-5.
5. Shaheen OH. Angiofibroma. In: John NG. Scott and Brown Otolaryngology. 5th ed. London: Butterworth-Heinemann; 1987. p. 291-4.
6. Kabot TE, Goldman ME, Bergman S, Schwartz RD. Juvenile nasopharyngeal angiofibroma: An unusual presentation in the oral cavity. Oral Surg Oral Med Oral Pathol 1985;59:453-7.
7. Enepekides DJ. Recent advances in the treatment of juvenile angiofibroma. Curr Opin Otolaryngol Head Neck Surg 2004;12:495-9.
8. Nicolai P, Berlucchi M, Tomenzoli D, Cappiello J, Trimarchi M, Maroldi R, *et al.* Endoscopic surgery for juvenile angiofibroma:

When and how. Laryngoscope 2003;113:775-82.
9. Maurice M, Milad M. Pathogenesis of juvenile nasopharyngeal angiofibroma. Braz J Otorhinolaryngol 1981;95:1121-6.
10. Andrade NA, Pinto JA, Nóbrega Mde O, Aguiar JE, Aguiar TF, Vinhaes ES. Exclusively endoscopic surgery for juvenile nasopharyngeal angiofibroma. Otolaryngol Head Neck Surg 2007;137:492-6.
11. Neville BW. Soft tissue tumors. In: Neville BW, Damn DD, Allen CM, Bouquot JE. Oral and Maxillofacial Pathology. 1st ed. Philadelphia, Pennsylvania: WB Saunders Company; 1995. p. 362-415.
12. Harrison DF. The natural history, pathogenesis, and treatment of juvenile angiofibroma. Personal experience with 44 patients. Arch Otolaryngol Head Neck Surg 1987;113:936-42.
13. Lloyd G, Howard D, Phelps P, Cheesman A. Juvenile angiofibroma: The lessons of 20 years of modern imaging. J Laryngol Otol 1999;113:127-34.

How to cite this article: 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;6:98-102.

Source of Support: Nil. **Conflict of Interest:** None declared.