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Case Report

Dentistry Section

Juvenile Nasopharyngeal Angiofibroma Extending into the Oral Cavity: A Rare Entity

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ABSTRACT

Juvenile nasopharyngeal angiofibroma (JNA) is a rare vascular tumour which is benign but locally aggressive and occurs invariably in young and adolescent males. It seldom involves the oral cavity but has the tendency to invade the adjacent structures. Its characteristic features include slow progression, aggressive growth & an increased rate of persistence and recurrence due to its location in inaccessible areas. In literature, very few cases of JNA have been reported with extension into the oral cavity. Here, a case of JNA with extension into the oral cavity has been discussed who reported to our institute.

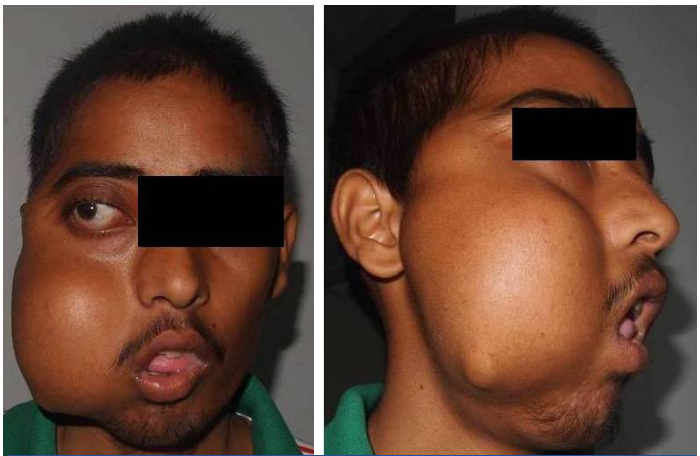
Keywords: Epistaxis, Locally aggressive lesion, Nasopharynx, Vascular tumour

CASE REPORT

A 15-year-old male patient reported to the Department of Oral & Maxillofacial Pathology at NIMS Dental College and Hospital, NIMS University with the chief complaint of painless swelling of the middle 3rd of the right side of face & nasal obstruction since four years. Patient had a history of trauma to face four years back; thereafter he noticed a small swelling at the middle 3rd of the right side of face which gradually increased to present size. He also gave history of epistaxis 2 to 3 times a day since the time of trauma. His past medical history was not contributory and on general examination he was found to be malnourished.

On extra oral examination, a large, solitary, expansile, diffuse swelling, of right middle 3rd of face approximately 5x3 cm in size extending superiorly from right eyebrow to angle of the mandible inferiorly & antero-posteriorly from right side of ala of nose to pretragus area was noticed. Patient was a mouth breather due to nasal obstruction & deviation of nose to the left with proptosis of right eye was also observed [Table/Fig-1]. The swelling was non tender & painless, soft, compressible with normal overlying skin. At the lower border of swelling a punctum was present without sinus opening [Table/Fig-2].

Intraorally, swelling involving right buccal mucosa, buccal cortical plates of right maxillary alveolus & right side of the hard palate in molar region was noticed [Table/Fig-3]. The swelling of the buccal mucosa was approximately 2x2 cm in size, non tender, soft, compressible, diffuse, and ovoid with normal overlying mucosa. Palatal swelling was present with respect to right maxillary second premolar, 1st molar & second molar, did not cross the midline & was firm in consistency.



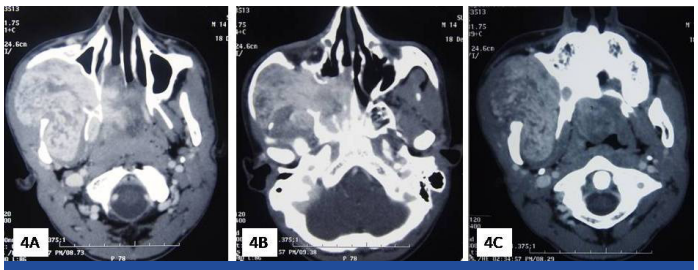
[Table/Fig-1]: Extraoral large solitary expansile diffuse swelling causing deviation of septum towards left **[Table/Fig-2]:** Punctum formation at the lower border of swelling. Proptosis of the right eye is also evident

[Table/Fig-3]: Intraoral swelling involving buccal mucosa & hard palate

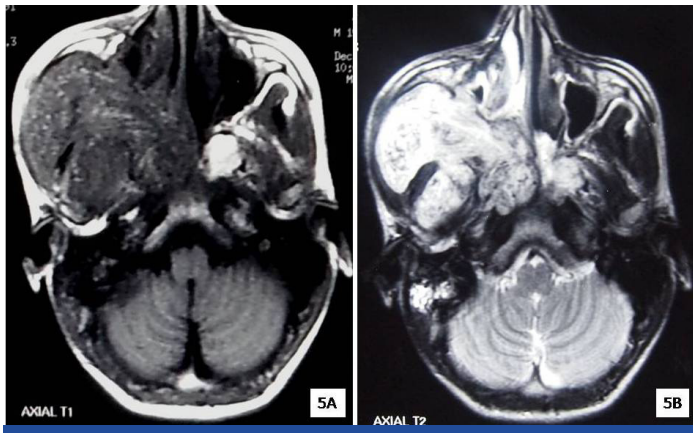
Computed Tomography showed a large infiltrating lesion with moderate heterogeneous contrast enhancement in right nasal cavity with extension into right infra temporal fossa & into subcutaneous fat of right cheek, lateral to body of mandible [Table/Fig-4a]. Bony erosion of wall of right nasal cavity, pterygoid plates, walls of sphenoid sinus, orbit & maxillary sinus wall were also seen [Table/Fig-4b] along with intracranial extension involving bilateral cavernous sinus [Table/Fig-4c].

Magnetic Resonance Imaging showed large mass lesion measuring 8cm x 8cm x 10.5cm, seen in right pterygopalatine fossa causing widening of fossa. Bowing & compression of right maxillary sinus with extension of the lesion into nasopharynx & right posterior nasal cavity through nasopalatine foramen was also seen [Table/Fig-5a]. Extension into orbit & right cavernous sinus with multiple flow voids are representative of a vascular lesion [Table/Fig-5b]. Angiogram showed multiple small & medium vessels supplied by right facial & lingual artery measuring 80cm x 54cm x 72 cm [Table/Fig-6a&b].

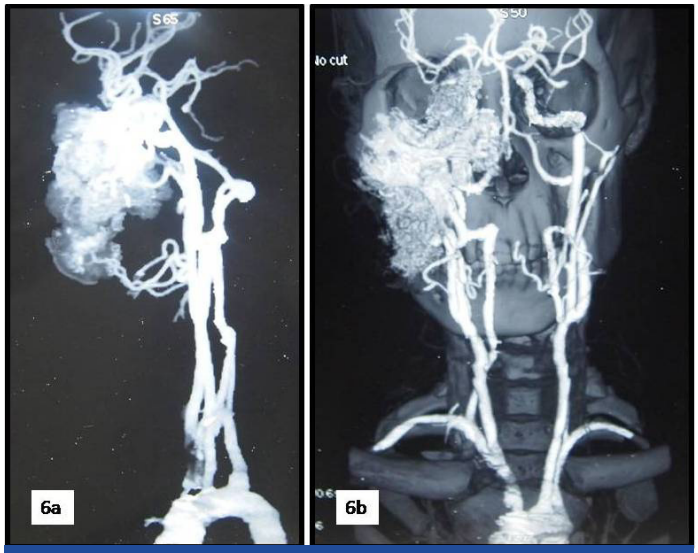
Fine needle aspiration cytology of the lesion was done which showed purulent fluid on aspiration. Numerous neutrophils & few chronic inflammatory cells against a necrotic background were seen, that were suggestive of acute or chronic inflammation.



[Table/Fig-4a-c]: Axial sections of contrast enhanced computed tomography scan reveals a) Large infiltrating lesion in right nasal cavity & nasopharynx. b) Bony erosion of wall of right nasal cavity, pterygoid plates, walls of sphenoid sinus, orbit & maxillary sinus wall. c) Intracranial extension involving bilateral cavernous sinus



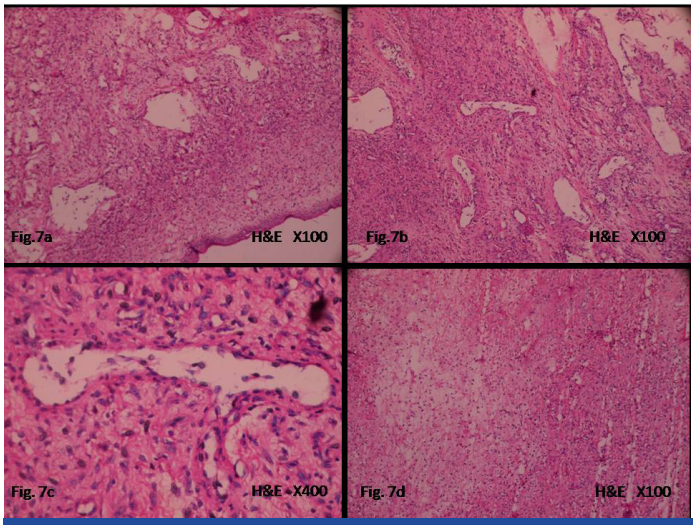
[Table/Fig-5a,b]: a) Axial section of T1 weighted Magnetic Resonance imaging reveals a large, ill defined heterogeneous enhancing mass lesion involving right pterygopalatine fossa. b) Axial section of T2 weighted Magnetic Resonance imaging reveals large ill defined hyper intense mass lesion involving right pterygo-palatine fossa. Multiple flow voids are also noted



[Table/Fig-6a,b]: ANGIOGRAM – a) Right external carotid artery angiogram reveals multiple feeding vessels of right facial and lingual artery and the hypervascular lesion. b)Three dimensional reconstruction

Histopathologically the incisional biopsy revealed a thin parakeratinised stratified squamous epithelial lining with almost flat epithelium & connective tissue interface [Table/Fig-7a]. Numerous & dilated vascular channels lined by single layer of endothelial cells & surrounded by dense paucicellular fibrous tissue were evident [Table/Fig-7b&c]. The cells in the fibrous tissue were cytologically spindle & stellate shaped with absence of hyperchromasia & had small nucleoli. Few myxomatous areas were also evident [Table/Fig-7d]. Extravasation of blood to the adjoining site & muscle bundles were also seen. Based on the clinical, radiographic and histopathological features diagnosis of Juvenile nasopharyngeal angiofibroma was made.

After explaining the surgical procedure to the patient & his parents, surgical resection of the lesion was planned. However,



[Table/Fig-7a-d]: a: Thin parakeratinised stratified squamous epithelial lining covering the lesion. b: Numerous dilated vascular channels lined by endothelial cells. c: Slit like vascular channel lined by single layer of endothelial cells. d: Myxomatous foci seen in the fibrous component

the patient did not report thereafter and so further follow-up could not be done.

DISCUSSION

Juvenile nasopharyngeal angiofibroma (JNA) is a rare vascular tumour, accounts for 0.05% of all head and neck tumours [1]. It originates in the posterior-lateral wall of the nasopharynx, where the sphenoid process of palatine bone meets the horizontal lamina of vomer and part of the pterygoid process of sphenoid bone. Growth of the lesion may occur beneath the mucosa, extending initially to the posterior nasal cavity and nasopharynx. Its blood supply is done by the internal maxillary artery, & may also by branches of the ipsilateral internal carotid artery [2].

Clinically JNA often present as a mass occupying nasopharynx and adjoining nasal cavity, producing nasal obstruction and epistaxis. Profound facial swelling, proptosis or diplopia, may be caused due to excessive growth of the tumour [3]. Very rarely intraoral features are seen, as seen in our case.

The pathogenesis of this tumour is still controversial & includes theories like congenital, genetic, hormonal & vascular. Several authors considered it as a vascular malformation while according to others it is a type of haemangioma. Immunohistological and electron microscopic studies suggested that this is a hamartoma rather than a true neoplasm. Role of chromosomal alteration are also detected in JNA. Gains at chromosomes 4, 6, 8, and X and losses on chromosomes 17, 22, and Y are the most frequent chromosomal abnormalities detected [4-8]. Schick also described AURKA gene (20q13.2) with a possible role in chromosomal and genetic instability in JA [6]. Andrade et al., stated that tumour selectivity by males may be explained by intra-nuclear accumulation of androgen receptor and beta-catenin, a co-activator that enhances tumour sensitivity to androgens. The genetic alterations observed more often involve sex chromosomes. Numerous growth factors like insulin-like growth factor, vascular endothelial growth factor (VEGF) and transforming growth factor beta (TGF-ß) have been implicated in the pathogenesis of tumour [9].

JNA has typical histological pattern, composed of the angiomatous and fibrous components. Sinus like vascular channels with small gapping that is lined by single layer of endothelium, surrounded by mostly incomplete rim of smooth muscle cells. Change in cellularity and fibrous connective tissue, associated with myxoid foci are seen in the fibrous component. Hyalinization is seen in the older lesions. Immunohistochemically, the endothelial cells of the tumour express positivity for the endothelial markers like CD34 and CD31. The perivascular cells show positivity for smooth muscle actin (SMA).

Androgen receptors also detected in nuclei of endothelial and stromal cells. Spindle cells show positivity of β-catenin virtually in all cases, while expression of c-Kit is observed variably [10].

Computerized Tomographic studies and Magnetic Resonance Imaging are the most efficient imaging techniques for determining the vascular nature of the tumour and delineating its borders, before undergoing any surgical procedures including biopsy [11].

The most widely accepted treatment modality for JNA is surgical resection. Other treatment modalities include radiation therapy, cryotherapy, hormone therapy, embolization, arterial ligation, use of sclerosing agents and observation with the hope of spontaneous regression. Preoperative hormone therapy with diethylstilbestrol 5 mg t.i.d for thirty days was suggested by Schiff. Patterson suggested an alternative regimen of ethinyl estradiol, given in a dosage of 0.1 mg t.i.d orally for thirty days. He also did a comparative study between the usage of estradiol and stilbestrol and found a dramatic improvement with estradiol [3,12].

JNA showed high rate of recurrence between 0% and 57% following treatment. The lesion mostly re-occurs due to the inadequate removal of the lesion because of its inaccessible location. Advent of sophisticated radiographic techniques leads to the assessment of the extension of the lesion & complete removal of the lesion for reducing the recurrence rate [13].

CONCLUSION

Although JNA is rare head and neck tumour but it should always be considered as a differential diagnosis when there is unilateral nasal obstruction, especially in young adolescent males. Our intention of presenting this case was to familiarize the dental surgeons with JNA and to keep in mind the lesions of JNA as one of the differential diagnosis of any maxillary soft tissue lesions with oral extension.

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