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Juvenile Angiofibroma: Tale Of A Rare Nasopharyngeal Soft Tissue Tumor

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Abstract

Nasopharyngeal angiofibroma (NA) is a locally aggressive, benign neoplasm arising predominantly in the nasopharynx or posterolateral nasal cavity wall of adolescent and young males. It is a rare condition with an estimated incidence is 0.4 cases per million in the general population. Patients usually present with epistaxis and nasal obstruction. The etiopathogenesis of NA is not definitively known. However, evidence of its hormonal dependency can be considered with disease presenting around the onset of puberty. Specific NA subsets are suggested to be associated with APC/beta-catenin pathway. Immunohistochemical nuclear expression of beta-catenin in stromal cells can be seen in more than 90% cases. Its highly vascular nature and anatomical complexity comes across as a surgical dilemma. Hence, imaging modalities are relied upon for diagnosis, early detection and effective management and preoperative biopsy is not recommended.

Keywords: Juvenile angiofibroma, Nasopharyngeal angiofibroma, Beta-catenin, Adolescent, Vascular

Introduction

Nasopharyngeal angiofibroma (NA) constitutes less than 0.5% of all head and neck tumors with an estimated incidence of 3.7 per million in the at-risk population (including 10- to 24-year-old males).[1] This tumor was first reported in the fifth century by Hippocrates and later in 1897, Chelius described the first case of this benign tumor as a polyp occurring in adolescence and early adulthood.[2,3]. The term juvenile angiofibroma was coined by Friedberg in 1940.[3]

NA is a benign mesenchymal, locally aggressive neoplasm arising typically in the nasopharynx or posterolateral nasal cavity wall and often shows extension into the surrounding tissues [3,4]. It may extend through the superior orbital fissure into the middle fossa of the skull causing loss of vision and cranial nerve palsies.[4,5] The extension can be accurately judged by imaging techniques like computed tomography (CT) scan and magnetic resonance imaging (MRI).[6] The vascular endothelial cells or fibroblasts are involved in the histologic origin of NA.[3]

Here, we describe a rare case report of NA in a 15-year-old male.

Case History:

A 15-year-old male presented to ENT OPD with chief complaints of a difficulty in breathing, foreign-body sensation in the nasal cavity and sporadic episodes of epistaxis for 4-5 months. Clinical examination revealed bilateral hypertrophy of inferior turbinate. The cold spatula test showed reduced misting on both sides and the cotton wool test showed reduced movement bilaterally. Investigations including blood counts were within normal range and no significant abnormality was noted on the chest X-ray.

On CT paranasal sinus (PNS) plain and contrast, a heterogeneously enhancing mass was noted in the

sino-nasal region with epicentre in the nasopharynx. Anteriorly, the mass projected into the nasal cavity with deviation of nasal septum and involving ethmoid sinus region. Medially, it crossed midline with nasopharyngeal involvement. Laterally, it extended into right spheno-palatine fossa, extending to superior and inferior orbital fissure and to deep masticator space with bowing of posterior maxillary wall on right side (Holman-Miller sign). Posteriorly, it extended to sphenoid sinus. Inferiorly, extension up to C3 was seen with involvement of left pharyngeal aspect. On angiography, the mass showed intense contrast enhancement predominantly from spheno-palatine branch of maxillary artery [Figure 1a]. The clinical differential diagnoses were juvenile nasopharyngeal angiofibroma and sinonasal carcinoma.

Endovascular embolization was done subsequently and the mass was sent for histopathological examination. Grossly, a single polypoidal, irregular grey-white soft tissue fragment was received in 10% neutral buffered formalin, measuring 11 cm x 4.5 cm x 2.5 cm [Figure 1b]. The tissue was processed following the standard protocol and hematoxylin and eosin-stained sections were examined.

On microscopic examination, tumor tissue revealed an intricate mixture of blood vessels and fibrous stroma. The partially collagenized stroma exhibited numerous irregular vascular spaces of variable sizes ranging from slit-like capillaries to large dilated vessels [Figure 2a and b]. The nuclei varied from small pyknotic structures to large vesicular forms with prominent nucleoli. The central portion of the tumor comprised of predominantly stromal elements and the peripheral portion comprised vessels that were closely packed, smaller and uniform in size with little stromal fibrosis [Figure 2c]. There was no evidence of mitosis or malignancy in the sections studied. Hence, a histopathological diagnosis of NA was considered, which was confirmed by immunohistochemistry (IHC).

On IHC, the tumor cells showed positivity for beta-catenin [Figure 3]. Stromal cells showed nuclear expression of beta-catenin and endothelial cells showed membranous and cytoplasmic expression of beta-catenin.

The patients' condition post operative period was uneventful and was discharged.

Discussion:

Juvenile nasopharyngeal angiofibroma is a rare, benign but locally destructive neoplasm that originates in the lateral wall of the nasal cavity, close to the superior border of the sphenopalatine foramen. It accounts for <1% of head and neck tumors and predominantly affects adolescent males with a high recurrence rate of 0–57%.[3,7] There is evidence of hormonal dependency of nasopharyngeal angiofibroma.

Molecular profile of NA includes loss of the Y chromosome with gain of the X chromosome. Up to 75% of these tumors show somatic mutation in exon 3 of the beta-catenin gene (CTNNB1), although beta-catenin nuclear localization is seen in > 90% of cases.[1] The beta-catenin may be relevant in adolescent males as it is suggested to function as androgen receptor coactivator protein.[8,9] The genetic profile of NA involving mutation in APC gene has also been reported. However, the association of FAP (familial adematous polyposis) in Indian population seems restricted to only a subset of NA, compared to majority of sporadic NA in the West.[9]

Any lesion, including inflammatory polyps, angiomatous polyps, nasopharyngeal cysts and carcinomas, soft tissue neoplasms such as papilloma, lymphoma, neurofibroma etc., presenting with unilateral nasal obstruction with or without epistaxis may be confused with NA. The clinical diagnosis of NA is achievable by complete history, clinical examination, radiography, nasal endoscopy and imaging techniques like arteriography, computed tomography scan and magnetic resonance imaging (MRI). These techniques make the precise staging of NA possible by establishing the exact site, extension and relation of the tumor to the adjacent structures. A histopathologic examination of tissue sections helps to obtain final diagnosis.[3]

Grossly, a variety of appearances may be seen, including polypoid and lobulated to rounded, circumscribed, non-capsulated mucosa covered mass, with an average tumor size ranging from 4 cm to 22 cm. The vascularity decides the color of the lesion varying from pale white in less vascular lesions to pink in highly vascular lesions [1,3] Histologically, some studies suggest that NA originates from myofibroblasts whereas some suggest that the spindle cells of NA are fibroblastic in origin. [3,5] The lesion

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exhibits a fibrocellular stroma comprising spindle cells and haphazardly arranged collagen. An irregular interspersed vascular pattern is observed with slit-like or dilated blood vessels of various calibers, organized in clusters. A higher density is seen commonly towards the periphery, which was also seen in our case. Mitosis is rare. Numerous inflammatory cells such as mast cells and T-lymphocytes are also observed.[3,5] The age of the patient in our case was 15 years, which correlates with a productive age group and a tumor in this age becomes a big hurdle in education / work.

Surgical excision of the tumor is considered as the gold standard treatment. However, endoscopic surgery is not advised, if the tumor extends laterally in the fossa infratemporalis or deep into the skull base.[2] Prognosis depends on multiple factors including the size and extent of the tumor, the presence of multiple feeding vessels and the completeness of surgical resection.[1]

Conclusion

NA is an uncommon tumor having high vascularity. It should be considered as a differential diagnosis during clinical evaluation of a painless nasal obstruction with or without epistaxis and rhinorrhoea to provide early diagnosis, plan treatment and determine prognosis.

Ethics Statement

Informed consent was obtained from all individual participants included in the study. The case report was approved by the Institutional Review Board of Ramaiah Medical College and Hospital. Informed consent was waived by the Institutional Review Board. Institutional Review Board approval was waived due to the use of retrospective, de-identified data. This study adhered to the guidelines enacted by the Office of Human Research Protection that is supported by the U.S. Department of Health & Human Services. All information included in this report is de-identified and no personal details that may be used to identify the patient are included in this report.

Author Contributions

Conceptualization: SM, SK.

Data curation: SM, SK, RS.

Formal analysis: SM, SK .

Investigation: SM, SK, RS.

Methodology: SM, SK, RS.

Project administration: SM, SK.

Resources: SK.

Software: SM, SK, RS.

Supervision: SM, SK.

Validation: SM, SK, RS.

Visualization: SM, SK, RS.

Writing - original draft: SM, RS.

Writing - review & editing: SM, SK

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Figures

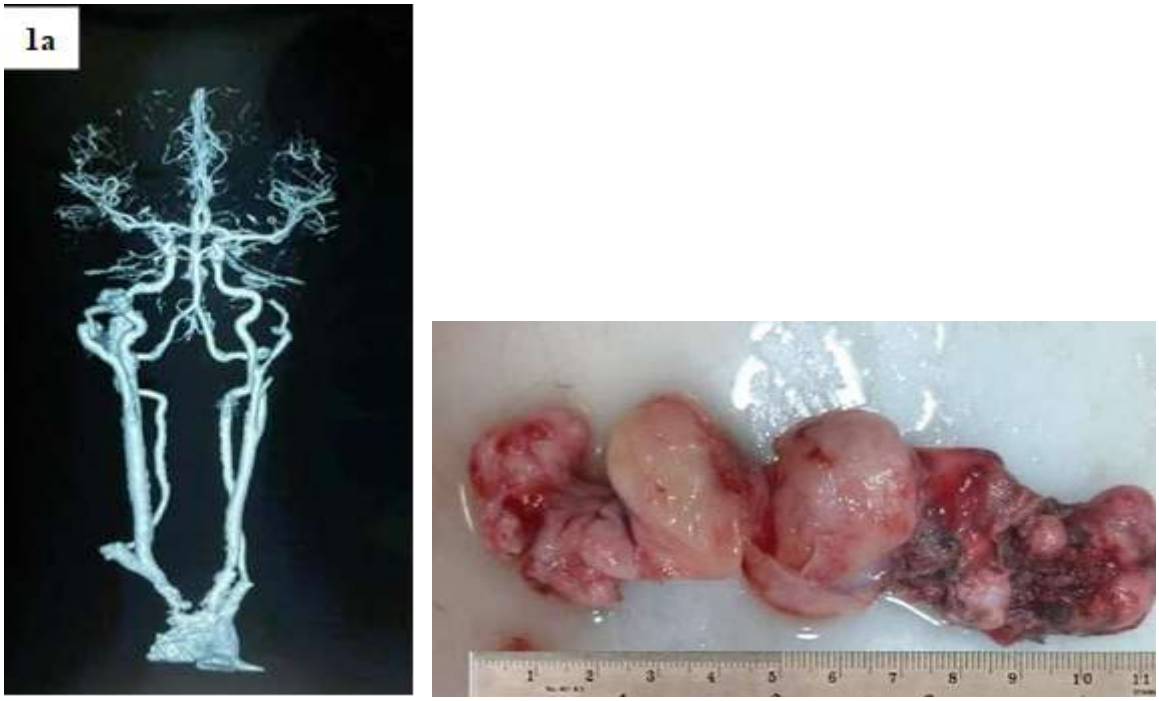

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Figure 2a

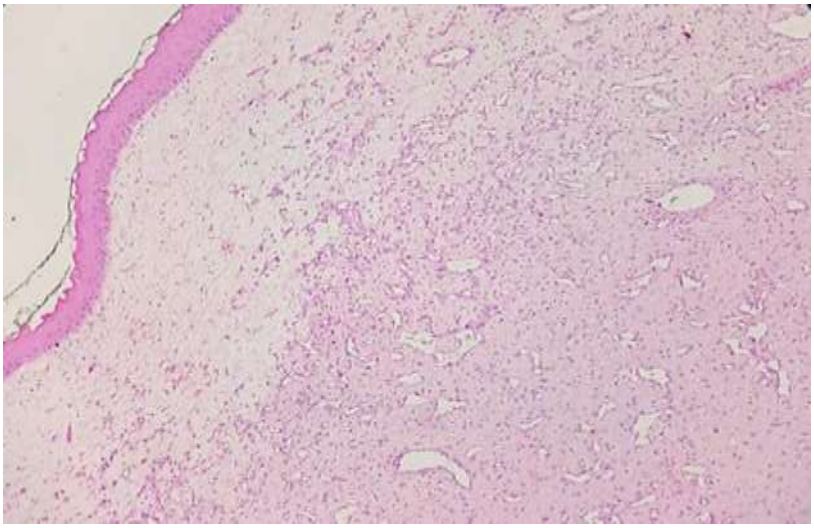
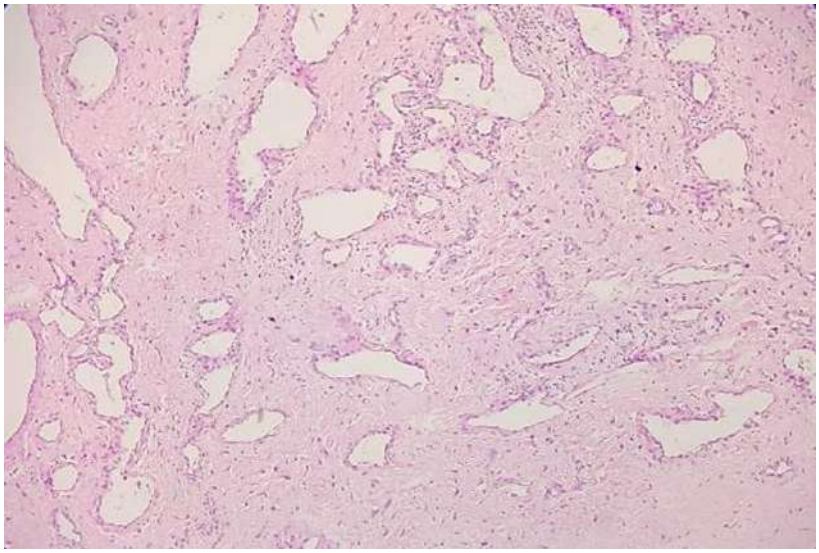


Figure 2b



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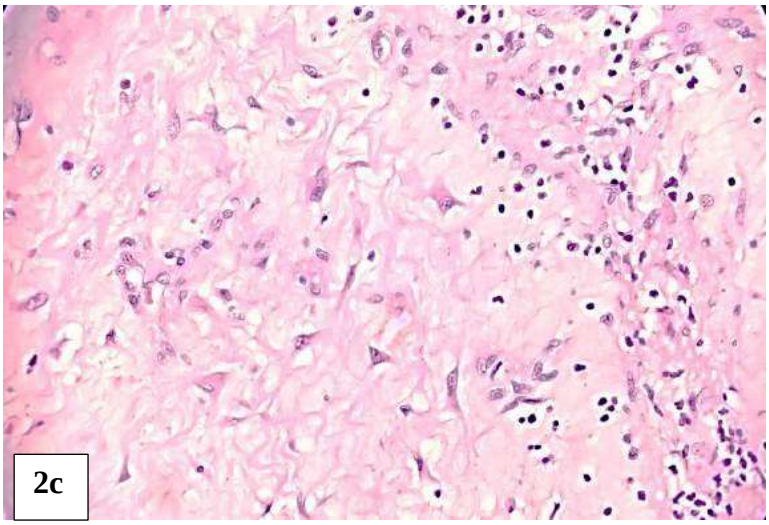

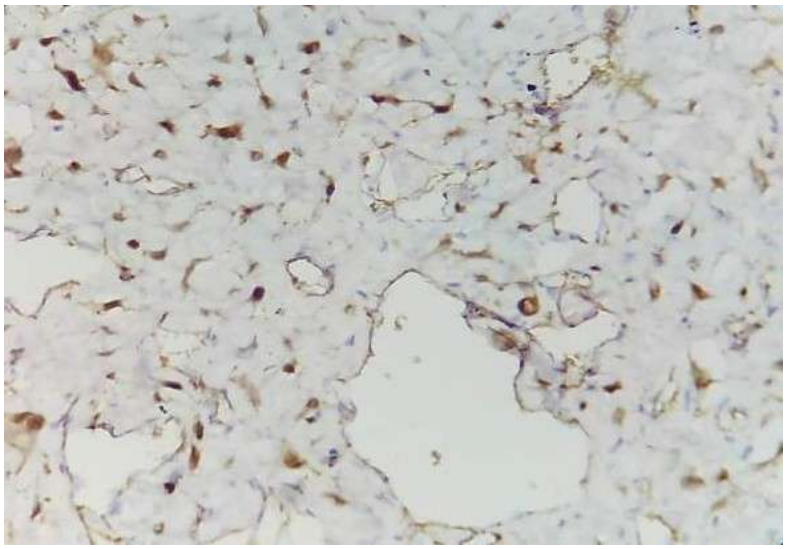

Figure Legends

1a – Intense contrast enhancement of NA predominantly from sphenopalatine branch of maxillary artery.

1b – Gross specimen showing polypoidal tumor mass with vascular areas.

2a – Photomicrograph from the tissue showing a vascular tumor underlying the nasopharyngeal respiratory mucosa in a cellular fibroblastic stroma (H&E, x100).

2b – Photomicrograph showing variably sized blood vessels ranging from slit-like branching capillaries to large dilated vessels (H&E, x400).

2c – Photomicrograph showing stellate & angulated fibroblasts (H&E, x400).

3 – Photomicrograph showing nuclear localization in stromal cells with membranous & cytoplasmic localization in endothelial cells (immunostaining beta-catenin, x400).

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