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An Unusual Presentation of Juvenile Nasopharyngeal Angiofibroma- A Case Report

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Abstract

Objective: To publish this rare case of Juvenile nasopharyngeal angiofibroma in a 24year old adult male with no epistaxis.

Methods: A 24 year old male presenting with left sided nasal obstruction since 9 months without epistaxis was evaluated thoroughly by clinical examination, hematological, radiological and histopathological evaluation.

Results: Clinical examination revealed a smooth pinkish globular mass in left nasal cavity; Hematological investigations were within normal limits. Radiological evaluation with contrast enhanced CT showed significantly enhancing globular mass with lobulated margins extending from pterygopalatine fossa in to left nasal cavity. Patient was managed surgically with transnasal endoscopic excision of the tumor. Histopathological examination showed pseudostratified columnar epithelium with multiple vascular areas of varying sizes and loose fibro collagenous tissue containing stellate cells and mast cells in subepithelium. With all the above evaluations, diagnosis was concluded as juvenile nasopharyngeal angiofibroma.

Conclusion: It is an unusual presentation of nasopharyngeal angiofibroma in an adult with 9 months history of nasal obstruction without epistaxis.

Introduction

Juvenile nasopharyngeal angiofibroma is a benign, highly vascular, locally aggressive tumour arising from nasopharynx. It most commonly occurs in adolescent males. Almost all cases present with nasal obstruction of insidious onset and unprovoked repeated epistaxis. Here we report an unusual presentation of a case of JNA in a 24 year old adult male presenting with left nasal cavity obstruction and without epistaxis.

Case Presentation

History

A 24 year old male presented with chief complaint of progressive left sided nasal obstruction for 9 months associated with mucoid nasal discharge, intermittent unilateral (left sided) headache which subsided with mild analgesics, mouth breathing which was evident specifically during sleep. Not associated with epistaxis, no external facial deformities, no fever, no diplopia, and no decreased vision. No history of trauma. No ear or throat complaints (Figure 1).

Clinical examination

General examination did not reveal any significant abnormality. Anterior rhinoscopy revealed deviated nasal septum to right with pinkish smooth globular mass in left nasal cavity. Posterior rhinoscopy showed smooth pinkish mass arising from choana of left nasal cavity occupying the nasopharynx completely on the left and crossing the midline slightly.

Investigations

Diagnostic nasal endoscopy findings: Deviated nasal septum to Right, single smooth pinkish globular mass in posterior part of left nasal cavity occupying entire left choana. On suctioning mass was not bleeding and insensitive to touch (Figure 2).

The Differential diagnosis of the mass is

1) Infected antrochoanal polyp

2) Hemangioma

3) Grossly enlarged adenoid mass

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Figure 1: 24year old Male with complaint of left sided nasal obstruction.

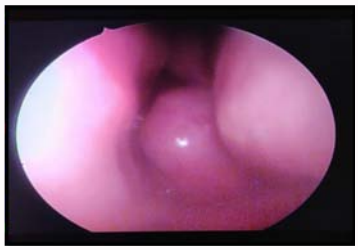

Figure 2: Preoperative Diagnostic Nasal Endoscopic examination showing pinkish smooth globular mass in the posterior part of left nasal cavity.

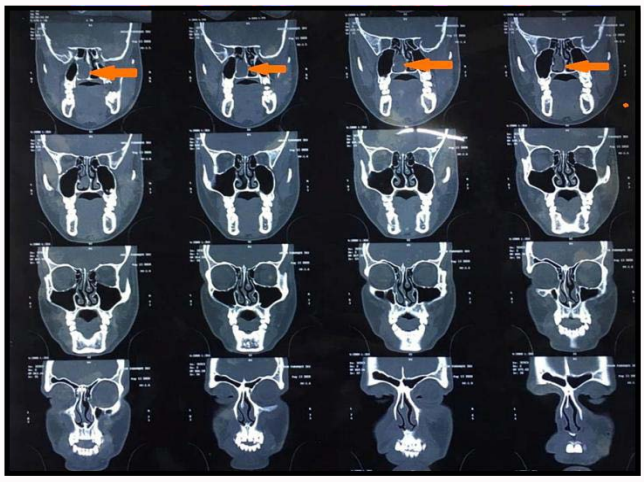

Figure 3: Coronal views of CT scan of nose and paranasal sinuses showing soft tissue mass in the posterior part of left nasal cavity.

4) Nasopharyngeal malignancies

5) Meningoencephalocele

6) Nasal glioma

7) Nasopharyngeal angiofibroma in young adult

Routine hematological investigations were found to be within normal limits.

Computed tomography of nose and paranasal sinuses showed a soft tissue mass in the posterior part of left nasal cavity extending into nasopharynx with widening of left sphenopalatine foramen. Enhancement of the mass was present in contrast enhanced CT scan, suggestive of nasopharyngeal angiofibroma with no intra cranial or intra orbital extension (Figure 3 and 4).

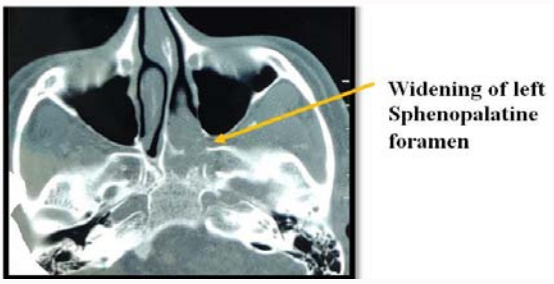

Figure 4: Axial view of CECT scan showing homogenously enhancing lesion in the posterior part of left nasal cavity extending into nasopharynx with widening of left sphenopalatine foramen.

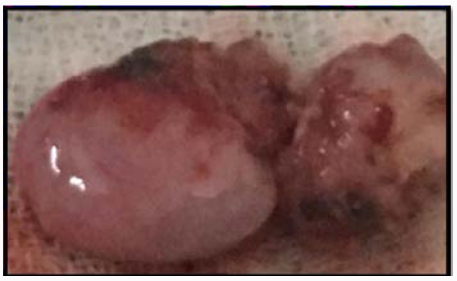

Figure 5: Post surgery specimen of the excised mass.

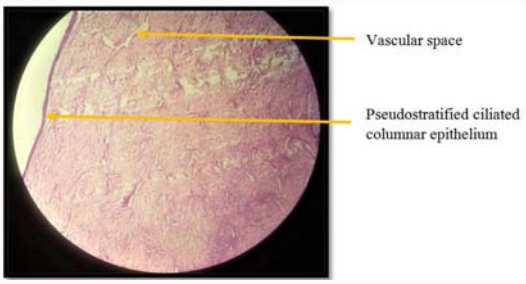

Figure 6: Histopathological image.

Treatment

Transnasal endoscopic excision of the tumor was done under general anesthesia. A pinkish globular mass of size about 2 cm × 1 cm was visualized in left nasal cavity arising just behind middle turbinate (sphenopalatine region) extending up to anterior end of middle turbinate and completely occluding left choana was delivered out of nasal cavity after releasing all attachments using coblator and monopolar cautery and was sent for histopathological examination. There was profuse bleeding during procedure and 1unit packed cell RBC was transfused. Postoperative period was uneventful (Figure 5).

Histopathology

The cut sections from mass showed pseudostratified ciliated columnar epithelium, subepithelium with loose fibrocollagenous connective tissue containing elongated spindle cells, areas with stellate cells and mast cells and having vascular spaces of variable sizes. Some vessels are showing smooth muscles in their walls. Features were suggestive of nasopharyngeal angiofibroma (Figure 6).

Follow up

Patient was followed up for two months. There were no signs of recurrence (Figure 7).

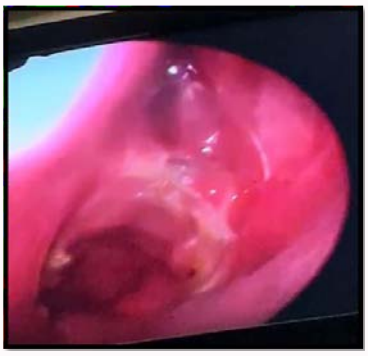

Figure 7: Postoperative diagnostic nasal endoscopy image of left nasal cavity.

Discussion

The commonest benign tumour of the nasopharynx is the juvenile angiofibroma [1]. It was first described by Hippocrates in 5th century AD and he called it as 'hard polyp'. It affects almost exclusively male adolescents with median age of 15 years; raising suspicion about the role of sex hormones in its pathogenesis [2]. It rarely involves the young adults; patient is a young adult in our case report. Goutham et al. [3] reported a JNA case of 39 years old whose age was higher than our case. Literature reports a few of these masses occurring in adults [4-6]. 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 [7].

Patients usually present with nasal obstruction and unprovoked epistaxis in most of the cases. This neoplasm is a combination of vascular and fibrous elements where symptoms predominate depending on the proportion of elements. In our case patient did not have epistaxis. As per literature almost all the cases reported with epistaxis. Abdelrahman EM Ezzat et al. [8] reported a case with single episode of epistaxis with long duration of nasal obstruction.

Patients are usually diagnosed based on history and clinical findings aided by imaging. In a study of 72 patients, Lloyd et al. [9] reported, three findings on CT and MR 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, erosion and widening of vidian canal. As there was no epistaxis, we have confirmed our diagnosis of JNA after CT imaging where it showed all the three findings mentioned above with homogenously enhancing mass on contrast enhanced CT-PNS.

Surgery remains the main stay in management [10]. Transnasal Endoscopic nasal surgical excision of the mass is preferred for small tumors. Other surgical approaches are, Transpalatal approach, lateral rhinotomy, midfacial degloving may be considered along with preoperative embolization, arterial ligation or use of sclerosing agents in large tumors. Incomplete excision of the mass leads to residual mass and recurrence. Other methods of treatment that have been employed are irradiation, hormone therapy and cryotherapy [11,12].

Conclusion

JNA though being a tumor of adolescence, it should be considered as a differential diagnosis of nasal mass even in young adults. JNA always may not present as nasal obstruction with recurrent epistaxis and nasal mass. Hence, radiological investigations like contrast enhanced CT, MRI and MR Angiogram are pathognomonic in diagnosis by exposing the high vascular nature of the tumor and also show origin and extension of tumor which is necessary for accurate staging of tumor, for choosing proper surgical approach and estimating the prognosis. This is a rare presentation of juvenile angiofibroma in a 24 year old adult male without epistaxis. Hence it is being reported.

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