CASE REPORT

Extranasopharyngeal Angiofibroma from the Nasal Septum: A Case Report and a Minireview

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Abstract

Extranasopharyngeal angiofibromas are rare tumors with only 65 cases being reported in the world literature. The most common sites involved are maxillary sinus, ethmoid sinus, sphenoid sinus, nasal septum, middle turbinate, inferior turbinate, cheek, conjunctiva, pterygomaxillary fissure, infratemporal fossa and laryngotracheal tree. We report on a case of 18 years old male patient of extranasopharyngeal angiofibroma of nasal septum.

Keywords: Juvenile nasopharyngeal angiofibroma, Vascular neoplasm, Extranasopharyngeal angiofibroma, Endoscopic excision.

INTRODUCTION

Juvenile nasopharyngeal angiofibroma comprises about 0.5% of all head and neck neoplasms and predominantly occurs in adolescent males.[1] They are pathologically benign but potentially locally destructive tumors. They are unencapsulated neoplasms composed of rich vascular network within a fibrous stroma.[2] In 1980, out of 704 cases of angiofibroma reviewed by de Vincentiis and Pinelli; only 13 cases occurred outside the nasopharynx, suggesting that extranasopharyngeal angiofibroma is a rare clinical entity.[3] Medical literature has been reviewed and only 65 patients of extranasopharyngeal angiofibroma have been reported. Among them, maxillary sinus is the most common site involved.[4] Nasal septum is an extremely unusual site and to the best of our knowledge less than 10 cases have been reported.[4-6] We report here a case of angiofibroma originating from nasal septum because of its unusual site of origin.

CASE REPORT

An 18-year-old male presented to the ENT outpatient department with history of left sided nasal obstruction and recurrent epistaxis for 3 months. Epistaxis was moderate in amount, spontaneous and used to stop by pinching of nose alone. The patient had no previous history of infection, trauma and any hematological disorder. On anterior rhinoscopy, a pinkish-red fleshy mass was seen filling the whole of left nasal cavity, reaching upto the vestibule. Nasal septum was found to be deviated to the right side. On posterior rhinoscopy, choanae were found to be free of mass. Keeping a diagnosis of a nasal vascular mass in mind, we got a CECT scan of nose and paranasal sinuses (PNS) done.

CECT of nose and PNS revealed an irregularly heterogeneously enhancing mass lesion occupying anterior part of left side nasal cavity causing its expansion with displacement of septum to right side. It was causing blockage of left osteomeatal complex (OMC) and was extending superiorly in left anterior ethmoid cells as seen in Figure 1. The posterior choanae and sphenoid sinus were completely free of mass as seen in Figure 2. There was no intraorbital or intracranial extension of lesion. The routine blood and urine investigations and X-ray chest were found to be within normal limits. A preoperative differential diagnosis of septal hemangioma/vascular mass/bleeding polypus was made.

The patient was taken for surgery and endoscopic excision of mass was performed. The mass was found to be attached to anterior 2/3rd of nasal septum and was removed in toto as shown in Figure 3.Total blood loss during surgery was around 120 ml and bleeding was controlled using electrocautery and packing. Postoperative course was

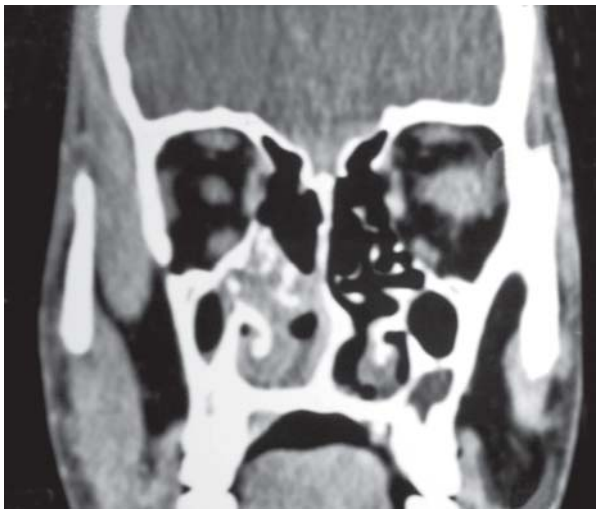

Fig. 1: CECT scan coronal cut showing mass in the nasal cavity blocking left OMC

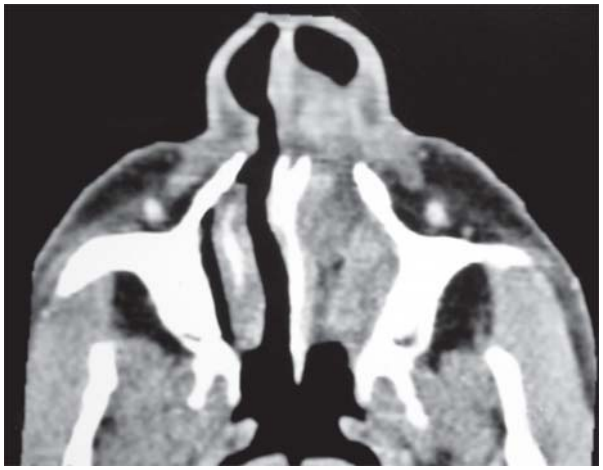

Fig. 2: CECT scan axial cut showing mass in left nasal cavity with free posterior choanae

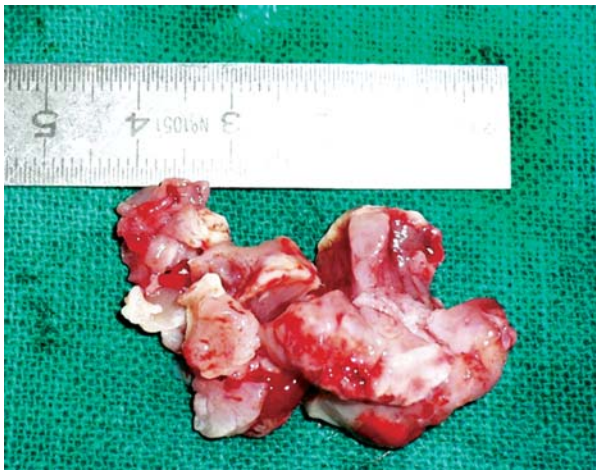

Fig. 3: Photograph of specimen

uneventful. Histopathology of the mass revealed ulcerated angiofibroma. There has been no recurrence in the follow-up of one year.

DISCUSSION

Juvenile nasopharyngeal angiofibroma (JNA), a well-known clinical entity, is the most common benign neoplasm of nasopharynx. Other benign pathologies include adenoids, Thornwaldt and mucosal cysts, fibromyxomatous polyps, choanal polyps, fibromas, papillomas, osteomas and fibrous dysplasia. The most common symptoms are nasal obstruction and epistaxis occurring in more than 80% of patients.[7]

There are various theories of origin of angiofibroma. Ringertz (1938) suggested that it originated from periosteum of nasopharyngeal vault. Bensch and Ewing (1941) thought that it arose from embryonic fibrocartilage between basiocciput and basisphenoid, whereas Brunner (1942) suggested origin from conjoined pharyngobasilar and buccopharyngeal fascia.[8] The etiology of JNA is debatable but predilection for adolescent males suggests a relationship with sex hormones. Recent studies suggest that nasopharyngeal angiofibromas originate from the superior margin of sphenopalatine foramen, formed by trifurcation of palatine bone, horizontal ala of vomer, and the root of pterygoid process.[7]

Extranasopharyngeal angiofibroma (ENA) is a totally separate and a rare clinical entity. Recent review of medical literature revealed 65 cases of ENAs till date. Among them, the maxillary sinus is the most common site involved. Other sites being involved are: ethmoid sinus, sphenoid sinus, nasal septum, middle turbinate, inferior turbinate, cheek, conjunctiva, pterygomaxillary fissure, infratemporal fossa and laryngotracheal tree.[4]

As compared to JNA, ENAs occur at slightly older age, i.e. 2nd decade with mean age being reported as 22 years as compared to 17 years for JNA. ENAs have been reported to have a higher incidence in females.[9] The clinical features of ENA will depend on the site and extent of tumor. Tumor originating from nasal septum mostly presents with nasal obstruction and epistaxis.

Our present case of ENA originated from perpendicular plate of ethmoid bone. This may be because of presence of fascia basalis which is not present in the cartilaginous part of nasal septum.[11] Contrast enhanced CT scan helps to differentiate between JNA and ENAs as JNA produces strong and homogenous enhancement as compared to heterogeneous or even no enhancement by ENA due to its poor vascularity.[8,10]

Table 1: Reported cases of extranasopharyngeal angiofibroma from nasal septum

Year	*Age/Sex*	*Site*	*Symptoms*	*Treatment*
1984[11]	13/M	Left side jn of bony and cartilaginous septum	Nasal obst'n, epistaxis- 2 months	Transnasal resection
1987[12]	9/M	Right side jn of bony and cartilaginous septum	Nasal obst'n, epistaxis- 6 weeks	Transnasal resection
2001[9]	8/M	Left side jn of bony and cartilaginous septum	Nasal obst'n, epistaxis- 6 months	Transnasal resection attempted- abandoned and then removed thru alar crease incision
2005[8]	27/M	Anterior 2/3 right nasal septum	Nasal obst'n- 6 years, epistaxis- 2 years	Transnasal resection
2008[13]	57/F	Posterior 1/3(vomerian bone) right nasal septum	Nasal obst'n- 1 year	Endoscopic resection
2009[14]	22/M	Antero-inferior cartilaginous left nasal septum	Nasal obst'n. Epistaxis- 2 months	Endoscopic resection

The first case report of ENA arising from nasal septum was a 13 years old Japanese male with complaints of recurrent epistaxis and left sided nasal obstruction for 2 months duration. The tumor was found attached to anterior 2/3rd of nasal septum by a small pedicle.[11]

We present a mini-review of cases of ENAs arising from nasal septum, reported in the literature in Table 1.

None of the cases reviewed from literature have reported any recurrence.[5] Surgery has been advocated as the mainstay of treatment of ENA; the approach may vary depending on site of origin and extent.

Because of the propensity of recurrence in case of incomplete excision and tendency of excessive bleeding; Extranasopharyngeal angiofibromas must be kept as a differential diagnosis in cases of vascular neoplasms of the nose.

SUMMARY

1. Extranasopharyngeal angiofibroma is a rare clinical entity.
2. Very few cases of angiofibroma arising from the nasal septum are reported in the world literature.
3. Angiofibroma should be kept in mind as a differential diagnosis in vascular septal masses.

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