e - ISSN - 2349 - 8005

INTERNATIONAL JOURNAL OF ADVANCES IN CASE REPORTS

Journal homepage: www.mcmed.us/journal/ijacr

EXTRA-NASOPHARYNGEAL ANGIOFIBROMA OF NASAL SEPTUM: A CASE REPORT

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Article Info
Received 09/03/2015
Revised 15/03/2015
Accepted 20/03/2015

Key words:
Angiofibroma, Extra-nasopharyngeal, Nasal septum.

ABSTRACT
Angiofibromas (AFs) are histologically benign but potentially locally aggressive vascular neoplasms originate predominantly in the nasopharynx. Extra-nasopharyngeal angiofibromas (ENAFs) are those originating from other sites such as the paranasal sinuses and nasal cavity which are less frequent, angiofibroma of the nasal septum is extremely rare,. Here I am reporting a case of extra-nasopharyngeal angiofibroma (ENAF) of nasal septum.

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a neoplasia that is histologically benign, locally aggressive, non-encapsulated, extremely vascularised, commonly located in the nasopharynx and predominantly occurs in adolescent males [1]. Its incidence is 0.5% among all head-neck tumours. They rarely originate from outside the nasopharynx which is called extra nasopharyngeal angiofibroma (ENAFs) [2]. The most common sites involved are maxillary sinus, ethmoid sinus, sphenoid sinus, nasal septum, middle turbinate, inferior turbinate, cheek, conjunctiva, pterigomaxillary fissure, infratemporal fossa and laryngo-trachial tree. These ENAFs are clinically distinct from the JAFs and therefore be misdiagnosed. A high level of suspicion is essential for an adequate diagnosis and treatment. Septal ENAF is a rare one up to my review about15 cases, for whom the main diagnosis and treatment is complete surgical resection with no recurrence [3].

Case Report

A 14 year old boy presented with history of recurrent right nasal bleeding for 3 months with frequency of about 3-4 times per month, mostly spontaneous. Bleeding stops either with minimal pressure if it is mild or with nasal packing if it is severe. Now with progression, bleeding is associated with nasal obstruction, right more than left, hyposmia, headache, post nasal drip, and snoring and sleep disturbance. There is no history of fever, change of vision, neck swelling or hearing impairment. No history of trauma, bruises or bleeding from any other areas. No history of any chronic disease. He has history of adenoidectomy at the age of 4 years. Family and social history are unremarkable. On examination, patient was looking healthy, stable and not bleeding. Nasal examination showed right nasal mass, pushing the septum to the other side. The left nasal cavity was narrow. The mass easily bleed on touching. Other ENT examination was unremarkable. The suspicion of angiofibroma was made clinically.

CT paranasal sinuses (PNS) showed thickening of nasal mucosa in the anterior inferior part of right nasal cavity (Figure 1), associated with acute sinusitis changes in the left maxillary sinus (Figure 2).

Patient was taken to the operation room after general routine laboratory investigations which were unremarkable. The plan was to do complete nasal

endoscopy with excisional biopsy if it is possible. During the operation , the mass was bigger than what it was appearing on CT scan, extending from the anterior end of inferior turbinate till the nasopharynx, attached with a small pedicle to the cartilaginous septum.

After infiltration of xylocaine with adrenalin around that pedicle, the mass was released with part of mucopericondrum without an unusual bleeding. Post operatively the patient did well and discharged within 2 days.

Histopathology of the mass showed prominent stroma, associated with dilated vascular channels with abnormal muscular layer. (Figure 3 a-b).

Fig 1. Coronal CT image showing soft tissue mucosa in the right nasal cavity

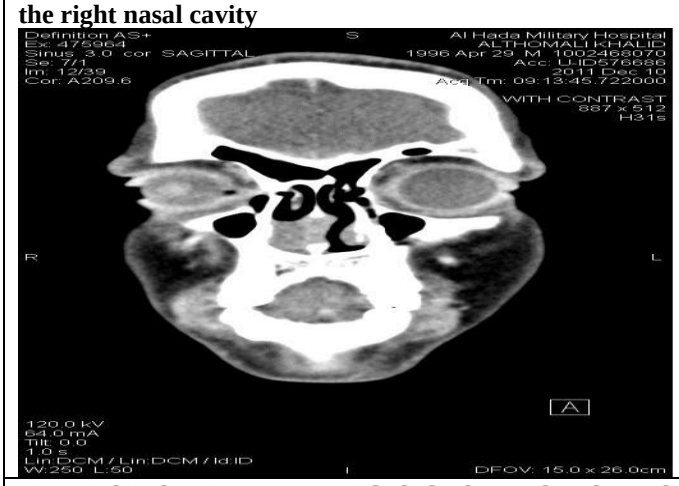


Fig 2. Axial CT image of showing left maxillary sinusitis

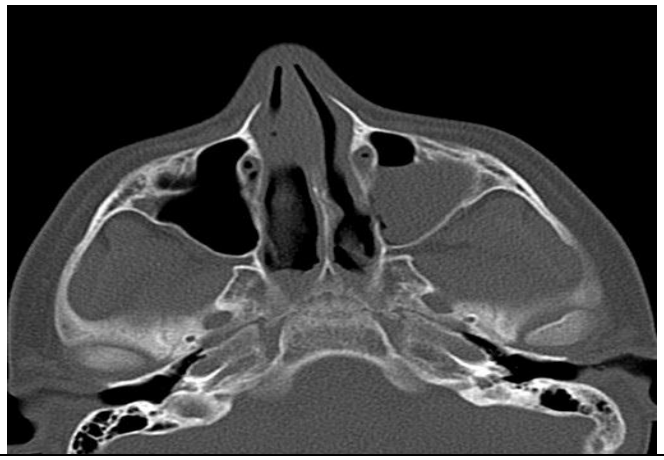
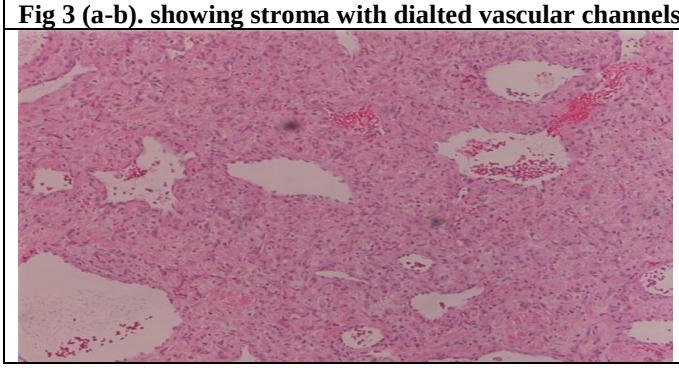
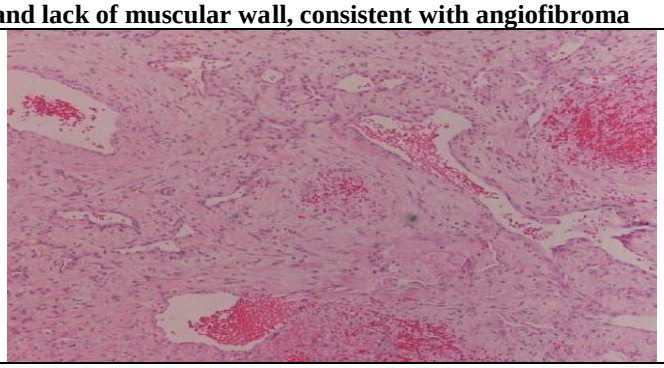


Fig 3 (a-b). showing stroma with dialted vascular channels and lack of muscular wall, consistent with angiofibroma





DISCUSSION AND CONCLUSION

JAF is a benign tumor but highly vascular with potential capability to expand locally causing serious morbidities either by destruction or sever bleeding. JAF arise typically in the nasopharynx specially at the trifurcation of the sphenoidal process of the palatine bone, the horizontal ala of vomer and the roof of the pterygoid process of the sphenoid at the posterolateral wall of the nasal cavity, near the superior margin of the sphenopalatine foramen from a facia called facia basalis [4]. JAF predominantly affecting adolescent males, which is part of criteria in the diagnosis of JAF, and diagnosed by clinical presentation, radiological features in CT scan & angiography [5] and confirmed with tissue diagnosis after biopsy which may cause serious bleeding. Treatment is surgical excision which may needs embolization before and special precautions like standby blood for the possibility of severe bleeding. The recurrence rate is sometimes high up to 50-60%.

ENAF is a separat and rare clinical entity. Review of the literature revealed that to date about more than 65 cases have been reported, most frequently (32%) localized in the maxillary sinus followed by ethmoid sinus, sphenoid sinus, nasal septum, middle turbinate, inferior turbinate, cheek, conjunctiva, pterigomaxillary fissure, infratemporal fossa and laryngo-trachial tree.

Septal subtype of ENAF is rarer; about 15 cases have been reported so far in the literature. Most of these originating from bony carilagenous junction and some from the anterior septum or from posterior bony part. They affect both sex (male: female 2:1), age is broad from 13-57, and presented all with nasal obstruction and epistaxis [6]. Radiological evaluation with contrast enhanced CT is essential which is mostly moderately enhancing in this type because of less vascularisation of these tumours. All the cases treated with surgical excision mostly endoscopically and the cure is completed without recurrence [7]. In

histopathological examination a dense acellular stroma and excessive collagen tissue is pathognomonic for the diagnosis [6, 7].

In conclusion, angiofibroma should be suspected for any vascular nasal mass regardless the age, sex or site of the lesion. Complete work up including nasal endoscopy, contrast enhanced CT scan and essential laboratory investigation should be done. Histo-pathological diagnosis may need more effort to find subclasses classification of angiofibroma if it is possible.

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