

Juvenile Nasopharyngeal Angiofibroma in a Woman: A rare case report

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ABSTRACT

Background: Juvenile nasopharyngeal angiofibroma (JNA) is rare otolaryngology disorder worldwide and in Saudi Arabia. It is a benign tumor that tends to bleed and occurs in the nasopharynx which most commonly occurs in men more than women.

Methods: A view of rare case report following a 27-year-old woman presented with recurrent attack of epistaxis for six months which was associated with nasal obstruction and snoring who was radiologically and surgically managed in Otolaryngology-Head and Neck Surgery (ORL-HNS) Clinic of Khamis Mushayt General Hospital and follow up for one year who was completely asymptomatic. **Conclusion:** Juvenile nasopharyngeal angiofibroma in a female is very rare, and if confirmed sex chromosome studies must be performed. Surgery is the mainstay of treatment of JNA. Preoperative embolization result in less blood loss and complete resection.

Keywords: Juvenile nasopharyngeal angiofibroma, rare, female, otolaryngology disorder

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a highly vascular, aggressive and locally invasive tumor. This tumor has a high potential to cause serious illness from severe epistaxis, involvement of intracranial structures and high-rate of recurrence. The age of presentation varies between 7 and 19 years ⁽¹⁾, with isolated patients presenting earlier or later. It is characterized morphologically by irregular, proliferating vascular channels within a fibrous stroma, which consists of plump, spindle or stellate cells. It has a peculiar propensity for local extension into the adjacent tissues that often precludes complete surgical resection and likely is responsible for tumor persistence and recurrences in 21 to 34% of the affected patients ⁽²⁾.

Topographically, this tumor frequently originates in the posterolateral wall of the nasal cavity, close to the superior margin of the sphenopalatine foramen. Subsequently, JNA may cause bone erosion and displacement of adjacent structures, thus involving the nasopharynx, paranasal, ethmoidal and maxillary sinuses, and orbit and skull base, with possible intracranial extension ⁽²⁾.

A review of the literature reveals rare cases of female and older male patients with JNA ^(3, 4, 5, 6). Malignant transformation of the tumor is also a rare finding and reported to be associated with recurrent radiotherapy ⁽⁷⁾.

This article aims to report an uncommon case of Juvenile nasopharyngeal angiofibroma.

CASE DESCRIPTION

A 27-year-old woman presented to the Otolaryngology-Head and Neck Surgery (ORL-HNS) Clinic of Khamis Mushayt General Hospital,

complaining of recurrent epistaxis for six months. Khamis Mushayt General Hospital is a tertiary care hospital in Khamis Mushayt City, Aseer Region, South western area of the Kingdom of Saudi Arabia. The patient complaint was mainly from right side with small amount. Symptom was progressive with no aggravating or relieving factors. It is associated with right nasal obstruction, snoring and headache. No history of visual loss, or diplopia.

On examination, the patient was generally stable, but she had right reddish nasal mass with nasal discharge Fig (1). No external affection, normal vision, and normal extraocular movement.

Basic laboratory investigations were normal. CT scan revealed expansile enhancing right nasal mass filling and ipsilateral maxillary and projecting posteriorly onto nasopharynx. Associated thinning out of the medial wall of the right maxillary sinus and medial wall of the right orbit Fig (2).

The decision was made to treat the patient operatively with preoperative embolization. Patient prepared for embolization by 48 hours, intervention radiologist detect the main blood supply of the lesion, which is right sphenopalatine artery, he did embolization of distal right maxillary artery with its branches. Fig(3A) shows conventional digital subtracting angiography with iv contrast that shows right nasal enhancing lesion, while Fig (3B) shows the lesion post embolization of distal right maxillary artery and its branches (nasopharyngeal angiofibroma) showing normal flow in right ICA and their branches, no flow through right external carotid artery mainly in the maxillary artery with no mass blush, and non-opacifying left common carotid artery appropriate with occlusion. Endoscopic sinus surgery was performed, there is right friable nasal mass, bleed on touch, all the lesion removed Fig (4) and sent for histopathology,

which revealed hemangioma with prominent fibrous stroma and ulcerated surface. Immunostaining positive result of CD 34, Actin, Vimentin and PgR are pointing toward angiofibroma Fig (5).

Follow-up of the case after one year revealed complete disappearance of her symptoms. Chromosomal study was not done for our patient due to lack of facility in our hospital.



Fig (1): Right friable reddish nasal mass.

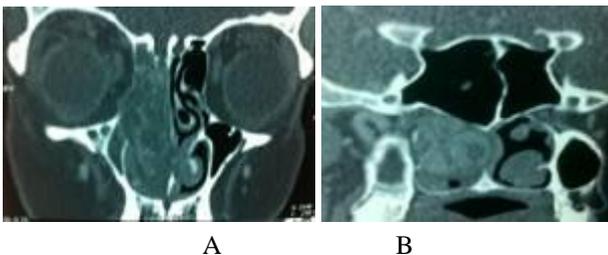


Fig (2 A,B): Enhancing right nasal mass and ipsilateral maxillary and projecting posteriorly onto nasopharynx.

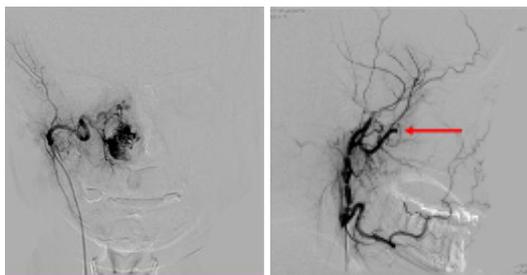


Fig. (3): (A): Angiography shows the enhancing mass. (B): Angiography shows the lesion post embolization.



Fig (4): Right nasal lesion.

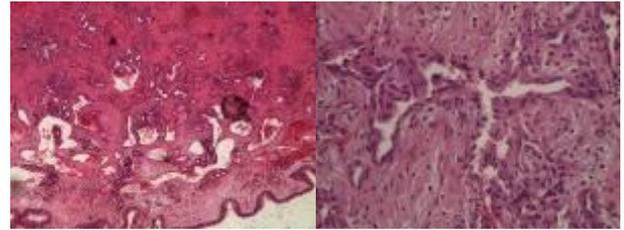


Fig. (5): Hemangioma with prominent fibrous stroma and ulcerated surface.

DISCUSSION

Juvenile angiofibroma (JNA) is a benign tumor associated to young males between 9 and 19 years of age. It makes about 0.05% of head and neck tumors, with an incidence between 1:5,000-1:60,000 (528) and 1:6,000-1:16,000 in the USA (529,530) being among the most frequent of the adolescence⁽⁸⁾.

Juvenile nasopharyngeal angiofibroma is a locally aggressive, polypoid tumor of the sphenopalatine foramen that originates from the primitive mesenchyme. It is a highly vascular tumor and occurs in adolescent males. Due to its intense vascular supply, diagnostic biopsies are mostly avoided. Thus, the patient history and the clinical and imaging findings play an important role in diagnosis.

Our patient was a 27 year old female presented with recurrent epistaxis and right nasal congestion for six months.

Szymanska et al reported that JNA is a histopathological benign tumor, but it extends into adjacent foramina, fissures, and sinonasal spaces and demonstrates a locally invasive behavior^(9, 10). Schicket al., added that JNA may extend into unexpected locations, it usually follows a predictable spread pattern. Originating from the sphenopalatine foramen, the pterygoid process, and the sphenoid sinus, JNA mostly extends medially and laterally, largely because it encounters less resistant barriers in these directions⁽¹¹⁾. Medially, the tumor usually invades the nasopharynx, the nasal cavity, and the maxillary and ethmoid sinuses. Laterally, it invades the pterygopalatine fossa and causes an anterior bowing of the posterior wall of the maxillary sinus (Holmann-Miller sign). Through the pterygomaxillary fissure, the tumor extends into the infratemporal fossa and laterally into the cheek^(9, 11).

Lloyd et al., stated that CT and MRI are the two primary imaging modalities used in the diagnosis and staging of these tumors. CT or MR angiography studies may also be used to detect the feeding vessels^(12, 13, 14). CT is superior in demonstrating the bone erosions and the invasion of the sphenoid bone, and MRI is more beneficial for the evaluation of the soft tissue, medullary bone marrow, and intracranial involvement⁽¹²⁾.

Szymanska et al., argued that the gold-standard treatment of JNA is the surgical resection of the tumor, and surgical options include endoscopic surgery. Endoscopic surgery is advantageous for cosmetic reasons and also has the main advantage of less intraoperative blood loss. However, the use of the endoscopic surgical approach for locally invasive advanced-stage lesions is limited. Preoperative tumor embolization performed with direct or trans-arterial injections of the liquid embolic agents are now frequently used to reduce the intraoperative blood loss. Radiotherapy, alone or combined with surgery, may be used in the treatment of partially respectable or advanced-stage tumors. The reported recurrence rates for JNA are between 20 per cent and 50 per cent, and advanced-stage or large-sized lesions carry the risk of higher recurrence rates. For all the above mentioned reasons, recognition of the tumor's imaging characteristics and its extension pathways is essential for the accurate preoperative staging of these tumors and determines the choice of treatment, including the postoperative radiotherapy^(9, 15).

In 1965, Apostol and Frazell reported 40 cases of JNA in male patients and suggested that if this diagnosis is confirmed in a female, sex chromosome studies must be performed, to investigate for androgen insensitivity syndrome (formerly testicular feminization) of a phenotypic female but genetic male⁽¹⁶⁾.

CONCLUSION

In Conclusion, the reported case is different from a classic case of juvenile nasopharyngeal angiofibroma in many aspects. The disease occurred in a female which is a rarity. In a female patient presenting with epistaxis, Juvenile angiofibroma is a rare albeit important differential diagnosis, as it challenges the hormonal theory of angiofibroma etiopathogenetic. Our diagnosis was confirmed by histopathological examination and was revised by the

Department of Pathology at our hospital. Follow up of patient is regular by means of nasal endoscopy to ensure the absence of tumor persistence or recurrence.

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