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Endoscopic Removal of a Rare Case of Extranasopharyngeal Angiofibroma: A Case Report and Review Article

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Abstract

Introduction: Angiofibroma is a benign, un-encapsulated, and highly vascular tumor which typically arises from the pterygopalatine fossa in the young adolescent males. It predominantly extends into the nasopharynx and rarely develops extranasopharyngeally. However, the most common origins of extranasopharyngeal angiofibromas are the maxillary sinus, ethmoid sinus and nasal cavity.
Case Presentation: Here, we report a 40-year-old male who presented with headache and epistaxis and was diagnosed as an extranasopharyngeal angiofibroma originated from the sphenoid sinus. The tumor was completely resected endoscopically. The 24-month follow-up showed no recurrence.
Conclusions: Angiofibromas should be considered as a possible diagnosis in any unilateral vascular mass of the nasal cavity, even in the older patients without typical imaging patterns.

Keywords: Nasopharynx, Angiofibroma, Nasopharyngeal Neoplasms, Paranasal Sinuses, Sphenoid Sinus, Endoscopy

1. Introduction

Angiofibromas are presented as rare vascular tumors, composed of 0.05% - 0.5% of all head and neck tumors (1). These benign un-encapsulated tumors with potential local destructive effect, occur almost exclusively in the male adolescent (2). Angiofibromas predominantly originate from the pterygopalatine fossa and extend into the posterolateral wall of the nasopharynx called as juvenile nasopharyngeal angiofibromas (JNAs). When the tumors arise outside the nasopharyngeal region they are termed as extranasopharyngeal angiofibromas (ENAs). Only few cases of ENAs have been reported. Due to the fact that they are extremely rare and mostly seen in the maxillary and ethmoid sinuses (1-3).

We report a 40-year-old male who was referred because of headache and long-term epistaxis and was diagnosed as ENAs.

2. Case Presentation

A 40-year-old man with a two-month history of headache, nasal obstruction presented to the emergency department with long term, recurrent, and massive epistaxis. There was no history of bleeding disorders, digital manipulation, or trauma. Family, drug, and habitual history were negative. Endoscopic examination revealed a large, vascular mass in the left part of the nasopharynx and left nasal cavity. According to the patient's documented medical records, an endoscopic biopsy of the nasal mass had been done about one month before referring to our department. The pathologic report confirmed angiofibroma as the definite diagnosis.

Computed tomography (CT) showed a large mass that partially had occupied the sphenoid sinus and was extended into the lateral wall of the nasal cavity and the posterior ethmoidal cells in the left side (Figure 1). There was not expansion and bony erosion of the left pterygopalatine fossa. According to the Chandler's staging of JNA, the tumor was classified as stage III. Magnetic resonance imaging (MRI) further delineated a soft tissue mass in the left sphenoid sinus measuring 32 × 26 × 23 mm with the same extension pattern of CT scan (Figure 2). The mass was demonstrated as intense contrast enhancement containing numerous punctate flow voids consistent with a vascular tumor with no intracranial extension.

The patient underwent an endoscopic surgery under general anesthesia, using image-guided navigation system. Local vasoconstriction was performed by 1/100,000

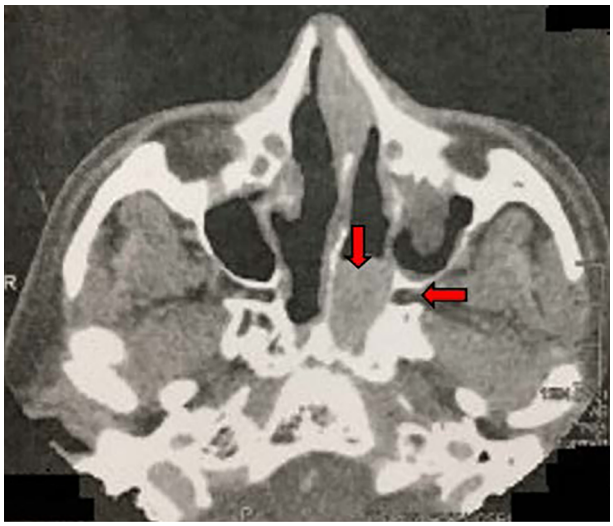

Figure 1. Axial CT scan with contrast shows a mass (vertical arrow). There isn't any pterygopalatine fossa widening or bowing of the posterior maxillary wall in the left side (horizontal arrow).

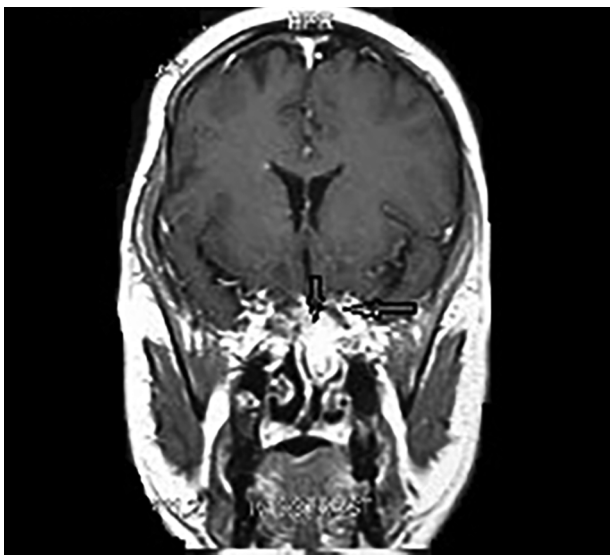

Figure 2. Coronal MRI with Gadolinium enhancement shows an enhanced mass in the left sphenoid sinus (vertical arrow) with some adhesions to the internal carotid artery (horizontal arrow).

epinephrine injection. Initially the tumor bulk was pushed into the nasopharynx using a long 1.5 centimeter- width wick. After partial resection of the middle conchae, endoscopic uncinectomy, antrostomy of the maxillary sinus, ethmoidectomy and esphenoidotomy in the left side were performed. The sphenopalatine artery in the medial part of the pterygopalatine fossa was exposed and ligated. The tumor was precisely dissected from the lateral nasal wall. The anterior wall of the left sphenoid sinus was removed completely. Then the tumor adhesions to the lateral wall of the sphenoid sinus and the left carotid artery were carefully released. Moreover the tumor was pushed out the left sphenoid sinus into the nasopharynx. Therefore, the en-block resection of the mass was performed and was sent for definite tissue diagnosis. The bony origin of the tumor in the anterolateral wall of the sphenoid sinus was drilled to reduce the rate of recurrence. Two expert pathologists confirmed the diagnosis of angiofibroma. The postoperative MRI with contrast revealed no residue of the tumor. The patient was followed by diagnostic endoscopy every month. Two years post-operatively, endoscopic examination and MRI imaging with contrast showed no recurrence.

3. Discussion

JNAs are highly vascular benign tumor which mainly occur in adolescent males with unclear etiology. JNAs usually present with the classical triad of nasopharynx mass, epistaxis and unilateral nasal obstruction. However, ENAs can spread deep into the cranial fossa and involve intracranial and major cranial base structures, meaning that they can be presented with various neurologic symptoms such as headache. This poses a considerable challenge and risk in the management of these lesions (4-6). The most prominent clinical feature is progressive unilateral nasal obstruction with or without epistaxis and nasal discharge. Eustachian tube dysfunction, facial deformity, pain, proptosis, impaired visual acuity, and cranial nerve palsy are other features which can be seen in advance stages. It seems that JNA mostly affects 14 - 25 year-old males. It appears as a firm and friable nasal mass on physical examination. It and should be differentiated from other causes of unilateral nasal obstruction such as: angiomatous polyps, nasopharyngeal carcinoma, and other sinonasal neoplasms. This study demonstrated that early diagnosis and treatment were associated with good prognosis. It is noteworthy to mention that surgical resection with preoperative embolization is the standard treatment option. Moreover, Lesions with orbital or intracranial involvement are difficult to manage with more recurrence rate. Furthermore higher recurrence rates are seen in cases with posterolateral extension of the tumor (7).

Angiofibromas predominantly originate from the sphenopalatine foramen and extended laterally into the pterygopalatine fossa (8, 9). The most common origins of ENAs groups are maxillary sinus, ethmoid, nasal cavity, and nasal septum. Also, there are rare reports of ENAs that were originated from cheek, retromolar area, middle turbinate, inferior turbinate, tonsils and parotid region

(10-12). Only few ENA cases originating from sphenoid sinus have been reported until now (12-14). In comparison to patients with JNAs; ENAs are extremely rare and also more common in female adults. ENAs should be accepted as a different entity. less vascularized, it could be diagnosed earlier and the patients are older than those with JNAs. In fact, the ENAs have different growth pattern. Although CT scanning is the initial imaging choice, angiography and MRI are helpful to define the vascularity and extension of the tumor. The typical landmark of JNA on the axial plane of CT scan is the so-called Holman-Miller sign which is defined as expansion of the pterygopalatine fossa and anterior bowing of the posterior wall of the maxillary sinus (15). While in ENAs, there is no typical imaging pattern, therefore; the diagnosis in this case would be more difficult.

The reported case was a 40-year old man with a rare ENA of the sphenoid sinus that was originated from the most anterior part of the lateral wall of the sphenoid sinus with some adhesion to the carotid artery. The patient's primary symptoms were nasal obstruction and massive recurrent epistaxis. He complained about headache due to involvement of the skull base in the sphenoid sinus, too. Although the clinical presentation was somehow typical, the patient was older than typical JNAs cases. Also, the posterior wall of the maxillary sinus was unchanged and there was no expansion in the pterygopalatine fossa. Despite ENAs, this sphenoidal angiofibroma was highly vascularized according to pre-op MRI and caused massive hemorrhage before and during surgery. Although, no embolization was performed before surgery, the sphenopalatine artery was ligated at the beginning of the operation to reduce bleeding.

Surgical interventions are the main treatment of JNAs. Various surgical methods have been identified. The most appropriate approach should be determined by various factors such as tumor size, vascularity and location, the patient age and the experience of the surgeon. The traditional methods include: transfacial, transoral, transpalatal degloving, and craniofacial approaches. Recent progresses in the field of endoscopic sinus surgeries have provided a minimally invasive approach to achieve excellent visualization and complete resection. The most important advantages of endoscopic approaches are low postoperative morbidity, significant reduction in the duration of the operation and low rate of recurrence (16). Furthermore, endoscopic removal ensures excellent preservation of physiology and anatomy of the nose and paranasal sinuses. Which is helpful especially in the management of recurrent tumors. Nowadays, endoscopic resection of JNAs is widely accepted for both early (16) and advanced stages of JNAs (17, 18).This method is associated with a low rate of residual lesions, which are reported in 6% - 17% of patients.

The tumor was resected endoscopiccally and the precise magnification of the endoscope and its wide-angle field of view, helped us to gently detach the tumor from the carotid artery and do en-block complete removal of the tumor.

3.1. Conclusion

However, angiofibromas commonly occur in young males and arise from pterygopalatine fossa, it should be considered as a possible diagnosis in any unilateral vascular mass of the nasal cavity, even in the older patients without typical imaging patterns (19).

Endoscopic surgery is the preferable approach considering the tumor extension and the surgeon experience.

3.2. Ethical Consideration

According to the ethical committee of Shahid Beheshti University of Medical Sciences., all medical ethics have been considered in reporting the case. The patient signed the informed consent. The authors had no conflict of interest or financial support to declare.

Footnote

Authors' Contribution: Study concept and design, Matin Ghazizadeh; acquisition of data, Fatemeh Mokhtarifar; analysis and interpretation of data, Matin Ghazizadeh, Fatemeh Mokhtarifar; drafting of the manuscript, Fatemeh Mokhtarifar, Matin Ghazizadeh; critical revision of the manuscript for important intellectual content, Matin Ghazizadeh, Fatemeh Mokhtarifar; administrative, technical, and material support, Fatemeh Mokhtarifar, Matin Ghazizadeh; study supervision, Matin Ghazizadeh.

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