Case Report

Juvenile nasopharyngeal angiofibroma: a case report

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ABSTRACT

Introduction: Juvenile nasopharyngeal angiofibroma (JNA) is a rare and locally invasive benign hypervascular neoplasm, representing 0.05 to 0.5% of all neoplasms in the head and neck. Though it is benign, it has an aggressive nature. The clinical presentation of this tumor is a triad of epistaxis, unilateral nasal obstruction, and a mass in the nasopharynx. It could cause massive epistaxis or intracranial involvement, which both lead to fatal consequences. It is most commonly seen in adolescent males because it is considered an androgen-dependent tumor.

Case Presentation: This report introduced a case of a 13-year-old Saudi male patient who presented two months after surgical removal of JNA with a complaint of progressively left-sided nasal blockage, snoring, rhinorrhea, hyposmia, and oral fullness with no epistaxis. Computed tomography (CT) with contrast of the sinuses was done and confirmed the recurrence of angiofibroma.

Conclusion: This report revealed a recurrent period of two months after the surgical removal of a primary tumor, which is shorter than the typical period of recurrence.

Keyword: JNA, Clinical presentation, Diagnosis, Management, Recurrence.

Introduction

Juvenile nasopharyngeal angiofibroma (JNA) is a benign, highly vascular neoplasm [1]. The lesion is almost exclusively seen in adolescent males, representing 0.05 to 1% of all neoplasms in the head and neck [2]. It has a triad of a mass in the nasopharynx, epistaxis, and unilateral nasal obstruction, suggesting the diagnosis of this tumor [1].

The tumor typically originates from the superior margin of the sphenopalatine foramen and spreads into the submucosal plane [1]. Despite being benign, juvenile nasopharyngeal angiofibroma has an aggressive evolution, especially in the youngest patients, which can result in massive epistaxis and intracranial involvement, which can have potentially fatal consequences. Thus, it should be treated as

Access this article online	
Quick Response Code:	Website: www.smh-j.com
	DOI: 10.54293/smhj.v4i2.95

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Received: 23 Oct 2023 **Received in revised form:** 22 Mar 2024 | **Accepted:** 3 Apr 2024

Please cite this article as: AlAli D, Alnefaie B, Al Qattan F, Alabdulmuhsin H, Alrasheed M, Almomen A. Juvenile nasopharyngeal angiofibroma: a case report. SMHJ. 2024;4(2): 112-115.

SMHJ
Saudi Medical Horizons Journal

Radically as possible [3, 4]. Here we present a case of recurrent juvenile nasopharyngeal angiofibroma after two months of surgical excision.

Case Presentation

A 13-year-old Saudi male patient from Alahsa presented with a complaint of progressively left-sided nasal blockage and snoring for two years, rhinorrhea, hyposmia, oral fullness, normal vision, and no epistaxis. On examination, there was a left bluish vascular polyp obstructing the left nasal cavity, pushing the septum to the right side. In addition, there was bulging in the soft palate. On the otoscope, both ears showed a slightly retracted tympanic membrane. Standard hematological tests revealed results that were within normal ranges. Radiological and tympanometry studies are shown in (Figures 1, 2), respectively. The surgery was done with a complete resection of the tumor; however, the patient presented with similar signs and symptoms to his primary complaint two months postoperatively. The patient was diagnosed with a recurrence of angiofibroma at the same site as the first tumor. The patient underwent another surgical resection of the mass. Nonetheless, the patient had another recurrence even after the second surgery after almost 4 months. Currently, the patient is followed up regularly to ensure there is no recurrence.

Surgical procedure: Under general anesthesia, an examination was done by a rigid scope zero & 30 degree that showed a left vascular nasal mass filling the nasal cavity. The mass was cauterized and dissected. The pedicle at the sphenopalatine foramen was identified, cauterized, and dissected. There was a medial attachment to the septal mucosa, which was removed. Posteriorly, the mass was attached to the nasopharyngeal wall, which was cauterized and removed. The posterior wall of the maxillary sinus was taken away using Kerrison Rongeur, and the extension of the mass into the pterygopalatine and infratemporal fossa was removed as well. Sphenoidotomy was done. Hemostasis is secured with diathermy and SURGIFLO. The procedure was done endoscopically, which was painless, simple, and cost-effective compared to an open procedure. The mass was sent to histopathology lab. It appeared as highly vascular fibrous proliferation with characteristic plump, angulated and stellate cells, categorized as fibroblasts, consistent with JNA.

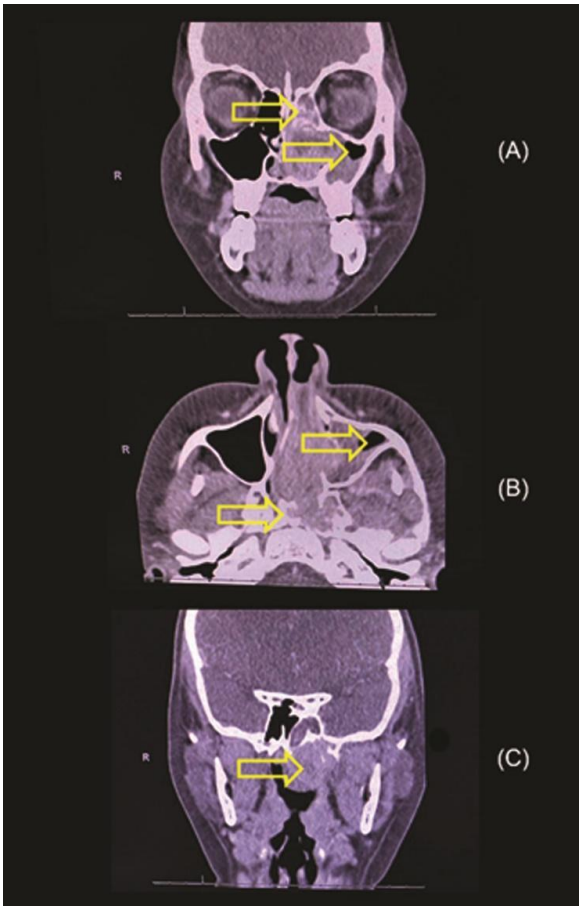

Figure 1: Computed tomography with contrast.

Computed tomography with a contrast of (A) sinuses was done and showed enhancing mass occupying and expanding the left nasal fossa with extension to the left maxillary sinus, (B) sphenoid sinus, and nasopharynx with underlying bone erosions of the medial wall of the maxillary sinus and the inferior wall of left sphenoid sinus, (C) the mass result in complete obstruction of the nasopharyngeal airway. There is the opacification of the ethmoid air cells. The left frontal and sphenoid sinus represent retained secretions. The nasal septum had deviated to the right. The cribriform plate of the ethmoid is intact. The image portion of the brain and orbit appears normal.

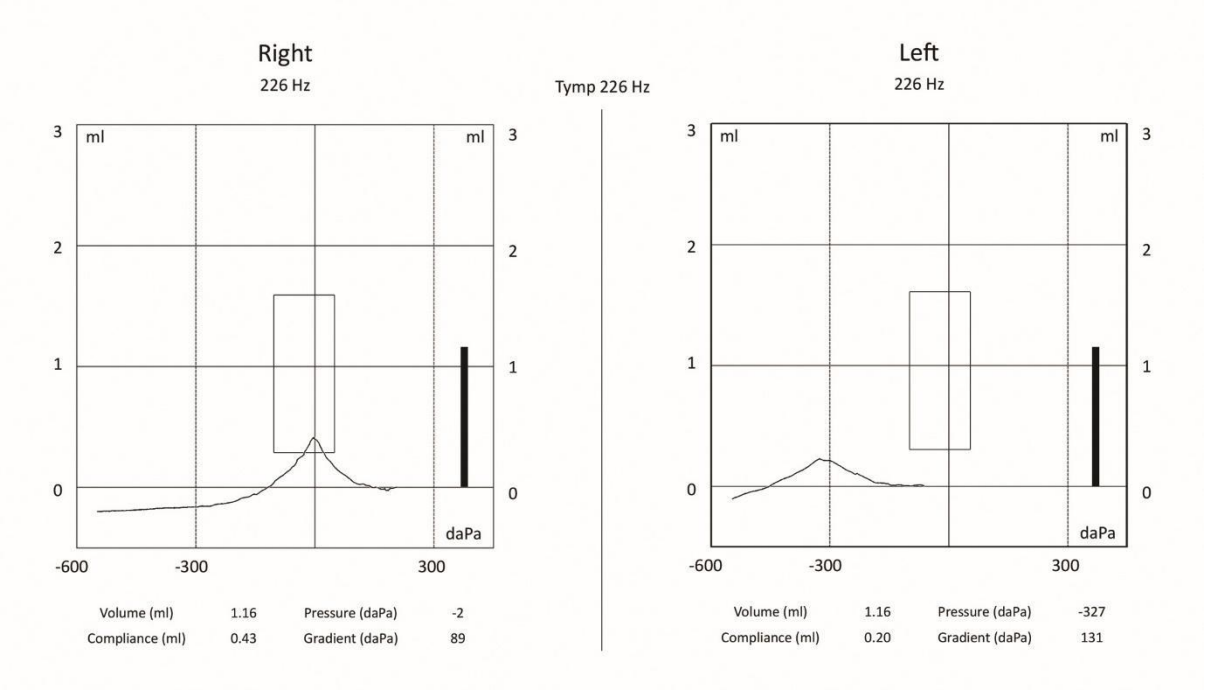

Figure 2: Tympanometry revealed right ear type A and left ear type C.

Discussion

Nasopharyngeal angiofibroma is a rare tumor commonly present among adolescent males. The tumor is linked to a specific age group, which is 14–15 years old [5, 6]. JNAs have a general incidence of 1:1,500,000 person [7]. As far as it known, there are no many studies on the duration of recurrence of angiofibroma postoperatively. However, a study conducted between 1997 and 2019 on 123 patients revealed that the recurrence of JNA mainly occurred between 6.4 and 16.6 months after primary surgical treatment [8]. Juvenile nasopharyngeal angiofibroma is a tumor that has a rich, irregular vascular network, which makes it bleed easily and is completely free of any muscular layer [9]. Furthermore, JNA originates within the sphenopalatine foramen, which is triggered by pathogenic hormones during the puberty period and continues to expand into the nasal cavity, paranasal sinuses, and pterygopalatine fossa [6, 10]. Almost all cases typically present with recurrent epistaxis, a mass in the nasopharynx and nasal cavity, and increasing nasal obstruction [6]. Moreover, the tumor has the potential to induce proptosis, which can damage the orbit and intracranial extension and eventually result in cranial nerve palsy and vision loss [3]. In our case, the patient complained of hyposmia, which can be caused by obstruction of the tumor itself or the extension of the tumor to the ethmoid bone affecting the olfactory nerve. Contrast computed tomography (CT) is considered the diagnostic modality for JNA because it shows accurate per-tumor anatomy, the extension, and staging of the tumor, which is beneficial for selecting the surgical technique, predicting the prognosis, and reporting results. Magnetic resonance imaging (MRI) is also used for the diagnosis of JNA, but it is more important to be done postoperatively to reveal any residual or recurrent tumor [6]. Another modality is angiography, which is commonly used preoperatively to embolize any feeding vessels to achieve optimal vascular control [5]. Surgery is the mainstay of management. The selection of the surgical technique depends on several factors, depending on the case. Surgical techniques include either open surgery or endoscopic nasal microsurgery [6]. However, the endoscopic technique is preferable in view of the fact that it has fewer postoperative complications [6].

Conclusion

This case report presented a case of recurrent juvenile nasopharyngeal angiofibroma after the first two months postoperatively. This case is inconsistent with previous literature reports, where JNAs recurrence cases were mainly reported after more than six months of surgical intervention. The cause(s) of recurrent JNA are not fully understood, although there are some risk factors, including residual tumors that are not fully dissected and the stage of the tumor itself. Further

retrospective studies should be conducted to assess the prospect of modifying the recurrence period and the possibility of unidentified contributing factors.

Conflict of Interest

None

Funding

None

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