Case Study

Extranasopharyngeal Angiofibroma of Maxillary Sinus: A Diagnostic Dilemma

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Abstract

Nasopharyngeal angiofibroma is a benign neoplasm of the nasopharynx with a propensity for aggressive local spread. It is responsible for 0.5% of head and neck cancer cases. Primarily affected are teenagers and young adults in the age range of 14–25 years. It is an aggressive tumor that frequently spreads locally, destructive, including the base of the skull and the cranium. Evidence points to an androgen-dependent tumor given the substantial preference for young guys. Usually, the tumor originates around the sphenopalatine foramen. We present a case of angiofibroma in a young adult arising from the lateral wall of the nasal cavity around the maxillary sinus, masquerading as an antrochoanal polyp. Diagnostic nasal endoscopy was suggestive of an antrochoanal polyp. NCCT nose and PNS showed that the tumor originated from the lateral wall of the nasal cavity around maxillary sinus ostium with and left maxillary sinus opacification. Histopathological examination shows dense fibrocollagenous stromal proliferation interspersed with staghorns of various sizes and numerous thin-walled vessels, characteristic features of juvenile nasopharyngeal angiofibroma. After surgical resection, the person was followed for up to six months without tumor recurrence.

Key words: Nasopharyngeal angiofibroma, antrochoanal polyp, sphenopalatine foramen, pterygoid, rhinorrhoea, epistaxis.

Introduction

Nasopharyngeal angiofibroma is a benign tumor of the nasopharynx with a tendency for aggressive local spread. It accounts for 0.5% of all head and neck malignancies [(1).] Young adults between the ages of 14 and 25 are the most affected[(2).] The prevalence in men is explained by the presence of high androgen receptors in the tumor, suggesting that it is androgen dependent. The tumor usually arises from the lateral wall of the posterior nasal cavity, near the sphenopalatine foramen and the base of the pterygoid [(2).] We present a case of angiofibroma in a young adult, in which the tumor arose from the lateral wall of the nasal cavity around the left maxillary sinus ostium.

Case Report

A 24-year-old adult male presented with complaints of progressive left-sided nasal obstruction with rhinorrhoea of four months duration. There was no history of epistaxis, anosmia, diplopia, blurring of vision, or cheek swelling. Nasal endoscopy revealed a grayish-white polypoidal mass completely filling

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the left nasal cavity. NCCT nose and PNS revealed the tumor to be arising from the lateral wall of the left nasal cavity with opacification of the left maxillary sinus (Fig. 1). Based on the symptoms endoscopic appearance of the mass, and radiological signs, a clinical diagnosis of antrochoanal polyp in the nasal cavity was made. A complete endoscopic excision of the mass was done under general anaesthesia. During the excision, there was substantial bleeding, which was unusual for an antrochoanal polyp. The whole mass was sent for histopathological examination, which was consistent with features of juvenile nasopharyngeal angiofibroma (Fig. 2). After surgical resection, the individual was followed for up to six months with no recurrence of the tumor.

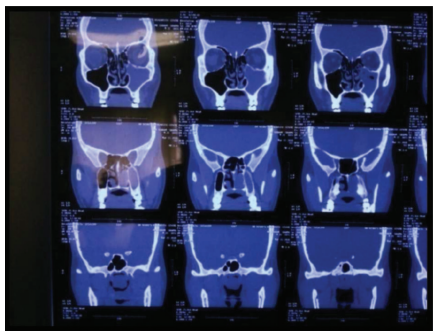

Figure 1: NCCT of Nose and PNS showing the origin of the tumour with opacification of maxillary sinus

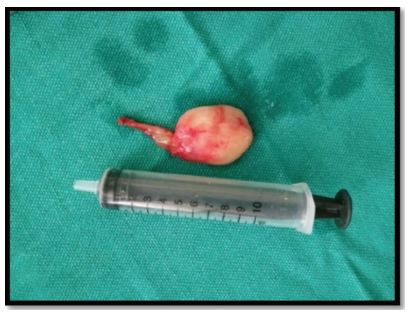

Figure 2: Resected Specimen

Discussion

Juvenile angiofibroma is a rare benign highly vascular tumor arising in the tissues of the sphenopalatine foramen. The first comprehensive study of nasopharyngeal angiofibroma was carried out by Bensch in 1878 in a review of the general subject of nasal polyps[3]. He compiled scattered case reports and produced an excellent morphological and clinical description of this tumor. JNA is almost exclusively seen in young men. Although not a malignant tumor, JNA is known for its high local invasiveness, which can contribute to significant morbidity associated with either intracranial extension (10–36%) or massive hemorrhage. The most common symptoms of JNA are unilateral nasal obstruction, rhinorrhea, and severe paroxysmal epistaxis. Late clinical manifestations include maxillary swelling, proptosis due to eye damage, orbital involvement, and increased ICH due to intracranial spread. Routine biopsy is contraindicated because of the risk of massive epistaxis.

Despite many proposed theories, the origin of the tumor is not well understood. Androgen receptors are present in 75% of cases. Proliferative cell nuclear antigen (PCNA), vascular endothelial growth factor (VEGF), and transforming growth factor β (TGF-β) may contribute to the pathogenesis of JNA by promoting angiogenesis and proliferation[4]. The nuclear accumulation of mutated beta-catenin suggested that the APC/beta-catenin pathway may be involved in the pathogenesis of JNA. Beta-catenin can act as a co-activator of androgen receptors and thus increase tumor androgen sensitivity, which may explain why JNA develops in young men [5, 6]. Microscopically, the image shows vascular spaces of different shapes and sizes against the background of the fibrous tissue (Figure 3). The relative proportion of vascular and stromal components changes with the age of the tumor. Tumor vessels typically lack smooth muscle and elastic fibers, which contributes to their reputation for constant bleeding.

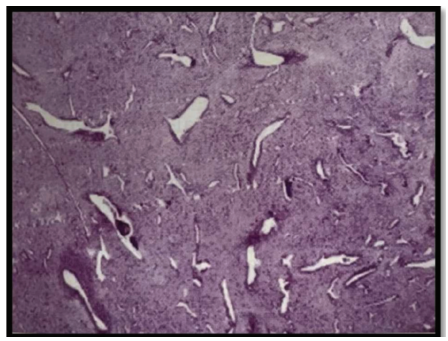

Figure 3: 20X magnification showing the staghorn and numerous thin wall vascular spaces within a stroma of fibrous tissue

Isolated case reports of angiofibromas arising outside the nasopharynx have been reported, most commonly in the maxillary sinus (32%) with ethmoid sinus and nasal septum are less reported sites [7-9]. Other rare sites include the larynx, sinus, cheek, conjunctiva, oropharynx, middle turbinate, and inferior turbinate. These tumors differ clinically from nasopharyngeal angiofibromas, being less vascular, affect older men or women, disappear completely with surgical resection, and have a lower tendency to recur [8]. As with typical nasopharyngeal angiofibromas, small tumors can be removed using an endonasal endoscopic approach.Large and extensive tumors require a midfacial degloving, lateral rhinotomy, or transpalatal approach. Radiation therapy may be used for inoperable tumors. In conclusion, extranasopharyngeal JNAs differ significantly in clinical and radiological presentation. They lack typical clinical and radiological features because they occur in all age groups, arise from different locations, may be less vascularized, cause different symptoms depending on the origin, and are often misdiagnosed at initial evaluation [10]. Extranasopharyngeal JNA should be in the differential diagnosis of an extranasopharyngeal mass with an unusual clinical picture.

Conclusion

Nasopharyngeal angiofibroma is a rare, highly vascular, locally invasive, unencapsulated tumor of the nasopharynx in teenage males. It appears as a unilateral, painless obstruction of the nasal passage, with or without rhinorrhea and epistaxis. Clinical examination, radiography, nasal endoscopy, and imaging methods like CT and MRI are used to make the diagnosis. Confirmation is done on histopathological examination. Because it is an aggressive tumor, nasopharyngeal angiofibroma may return after treatment. In order to effectively manage this lesion, early identification, precise staging, and appropriate treatment are therefore crucial.

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Conflict of Interest: None

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