CASE REPORT
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Intraoral Presentation of Juvenile Nasopharyngeal Angiofibroma: A Case Report

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ABSTRACT

Juvenile nasopharyngeal angiofibroma (JNA) is a vascular, locally destructive, and histologically benign tumor typically affecting the nasopharynx of adolescent males. This rare neoplasm has the potential to permeate through natural foramina and spaces and extend into neighboring structures. However, the intraoral extension of this tumor is extremely uncommon. Dental surgeons should remain alert to this possibility while evaluating such lesions. This article describes a case of JNA with intraoral extension in an 11-year-old boy and outlines the treatment procedures. JNA may rarely manifest intraorally as a submucosal swelling. This neoplasm should be considered in the differential diagnosis of intraoral lesions in adolescent males, especially when associated with nasal complaints.
Keywords: Oral, angiofibroma, adolescent

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a benign but locally aggressive vascular tumor that occurs almost exclusively in the nasopharynx of adolescent males. The tumor is uncommon and constitutes 0.5% of all head and neck neoplasms (1). This type of tumor often infiltrates through the surrounding tissues and has the capacity to extend widely. However, the intraoral presentation of JNA is exceedingly rare, with only a few cases having been reported in the literature (2, 3). Such an unusual presentation of a tumor may present the clinician with diagnostic difficulties while evaluating these types of lesions. This report describes a case of JNA with an extension in the oral cavity, alongside the relevant imaging and histopathological findings. The report also discusses treatment modalities and stresses the importance of an interdisciplinary approach in managing this unusual tumor.

CASE PRESENTATION

An 11-year-old boy reported in with the chief complaint of a large swelling in the right cheek that had been increasing steadily for the last six months, as well as an intraoral mass that had appeared recently, causing difficulty eating (Figure 1). The boy also presented with a history of episodic epistaxis, nasal obstruction, and noisy respiration over the last 6-12 months. The intraoral examination revealed a pinkish mass protruding from the posterior right maxillary alveolus just distal to the permanent first molar involving the pterygomandibular raphe and obliterating the buccal sulcus. The mass was found to be extruding through the tooth socket into the oral cavity. The mass was 2 cm in size, non-hemorrhagic, lobulated, and smooth surfaced. Significant bulging of the soft palate was also seen, as well as a mass to be present in the right nostril and toward the choana of the left nostril. A diffuse soft-to-firm swelling was palpated, encompassing the entire right cheek, extending from the malar process to the inferior border of the mandible.

Computed tomography (CT) showed a large mass in the nasopharynx and oropharynx, nasal cavity, posterior nares with superior extension to the roof of the nasopharynx, and apex of the orbit. Bowing of the posterior wall of the maxillary antrum (Holman-Miller sign) with erosive changes

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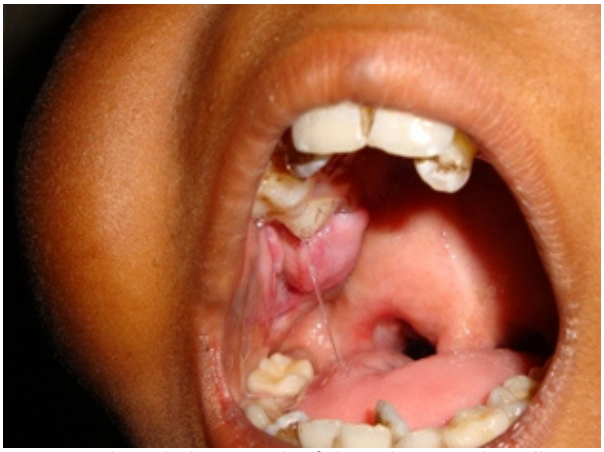

Figure 1: Clinical photograph of the submucosal swelling in the posterior maxillary alveolus.

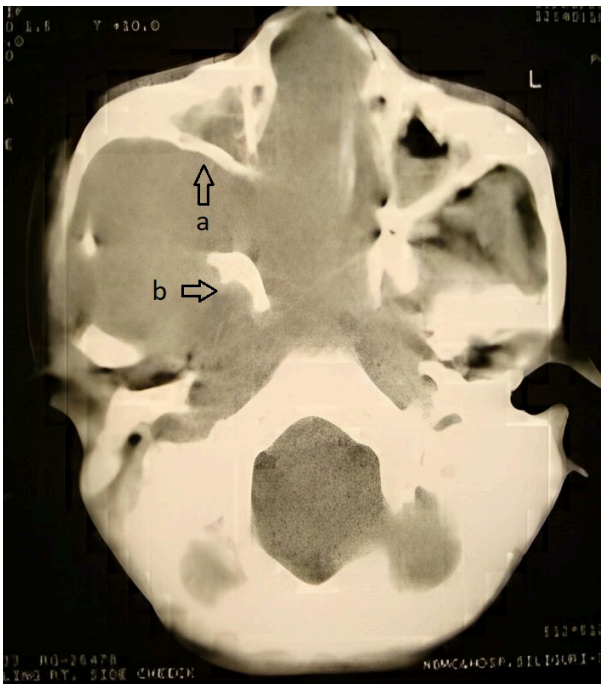

Figure 2: CT scan showing the extent of the lesion. (a) Holman-Miller sign; (b) Erosion of the pterygoid lamina.

in the right pterygoid lamina was also noted (Figure 2). The patient was then referred to the Ear, Nose, and Throat (ENT) Department, and a treatment plan was formulated following interdisciplinary discussions. A provisional diagnosis of JNA was made, and the patient was prepared for surgery. A sublabial incision was applied, extending from the upper central incisors to the retromolar region to expose the tumor. The tumor was removed through a transantral approach, along with midfacial degloving after releasing the attachments, and then removed orally. Brisk blood loss was encountered, which was managed by ligating the vessels in the tumor bed. The excised specimen measured approximately 12 cm along the anteroposterior dimension. Overall, the tumor appeared pale and rubbery firm with indentations from the pterygoids. The cavity was packed with sucralfate metronidazole in a roller pack that was brought

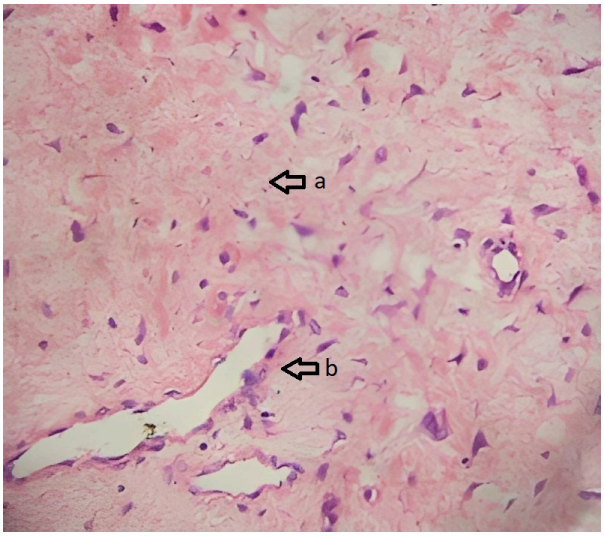

Figure 3: Photomicrograph of H&E stained sections (10x) showing a few vascular spaces in a mature fibrous stroma. (a) fibrous stroma; (b) thin-walled vascular spaces.

out through a widened middle meatus, and the wound was closed with chromic catgut. A histopathological examination of the excised specimen, which was stained with hematoxylin and eosin, revealed stellate fibrocytes in a mature fibrous stroma interspersed by thin-walled vascular spaces of variable calibers. These findings are consistent with the diagnosis of JNA (Figure 3). Postoperative healing was uneventful, other than transient conjunctival congestion and facial edema. The patient was advised to apply a routine nasal alkaline douche and follow-up was maintained with a nasal endoscope. The patient remained free of any recurrence three months post-operation and is scheduled for periodic check-ups.

DISCUSSION

JNA constitutes 0.5% of all head and neck tumors. Though this vascular lesion is benign, it however may show local aggressive behavior by extending through natural foramina and fissures. JNA is infrequently seen in dental clinics because of its rarity in intraoral locations and may cause confusion in diagnosis. Thus, the primary objective of this paper is to familiarize the clinician with this unusual presentation of JNA and to consider the possibility of JNA while evaluating lesions with similar characteristics. The clinical presentation of this neoplasm is often distinctive. It occurs almost exclusively in young males and is associated with rhinologic symptoms such as epistaxis, nasal blockage, and frequently a nasal mass. The site of origin of this tumor has been debated, but evidence suggests that it probably arises in the posterolateral wall of the nasal cavity at the junction formed by the sphenoid process of the palatine bone and the ala of vomer (4). Even the pathogenesis of this tumor remains speculative. Various symptoms such as ambryonal chondro-cartilage, normal embryonal fibrovasculature, testosterone, trauma, and heredity have been implicated in its formation (5).

One characteristic feature of JNA is its ability to spread, sometimes assuming large sizes. Sessions and Fisch classified the tumor under various categories based upon the extent of its spread (6, 7). In 1996, Radkowski proposed a newer staging system that has become more widely used in recent times (8). Facial swelling, proptosis, diplopia, and cranial neuropathy are suggestive of an extensive tumor. Diagnosis of the tumor is suggested by combining the clinical features, age at presentation, male sex, nasal complaints, and the characteristic imaging findings. CT scans are excellent imaging modalities, as they offer a superior delineation of the extent of the lesion and associated bone changes. The Holman-Miller sign, which is the anterior bowing of the posterior maxillary wall, is often picked up in CT scans and lateral radiographs. Surgery remains the most effective form of treatment. Newer treatment methods are being evaluated for unresectable tumors. Hormone therapy and low dose radiotherapy have also been tried in some cases (9). Various surgical approaches (e.g., transpalatal, sublabial and frontoethmoidal) have been advocated with the principal aim of gaining adequate exposure in order to completely remove the tumor, which is essential for avoiding recurrences (10). The prognosis for JNA is excellent in cases that are adequately treated. Angiofibroma has an unmistakable histological appearance, which essentially is the proliferation of the vascular channels lined by a single layer of endothelial cells in a fibrous stroma. The present case demonstrates that the dental practitioner should remain alert to the possibility of JNA while evaluating intraoral maxillary masses in adolescent males. The characteristic clinical presentation and imaging results should suggest the diagnosis. An unnecessary biopsy procedure should be avoided, as this may prove to be dangerous in vascular tumors such as JNA due to the risk of massive bleeding. The present case also underlines the importance of an interdisciplinary approach involving otolaryngologists in order to effectively manage patients with this tumor.

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