Case Report

Angiofibroma of the mandible: Rare sight for juvenile tumor

Mohammad I. Khan[1], Abhisheik Khare[1], Iqbal Ali[2], Abdullah Nasir[1]

[1]Department of Oral and Maxillofacial Pathology and Oral Microbiology and [2]Oral and Maxillofacial Surgery, Career Post Graduate Institute of Dental Sciences and Hospital, Uattardhona, Uttar Pradesh, India

Abstract Angiofibroma also called juvenile nasopharyngeal angiofibroma are tumors of adolescence and the commonest site is the nasopharynx. Extra nasopharyngeal sites include upper respiratory and digestive tracts, oral cavity, tonsils, larynx, trachea, and esophagus. Intraosseous angiofibroma is the rarest of a rare entity.

Keywords: Angiofibroma, ectopic angiofibroma, feeder artery, juvenile angiofibroma

Address for correspondence: Dr. Mohammad I. Khan, Department of Oral and Maxillofacial Pathology and Oral Microbiology, Career Post Graduate Institute of Dental Sciences and Hospital, Uattardhona, Uttar Pradesh, India.
E-mail: imranplz@yahoo.co.in

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INTRODUCTION

Angiofibroma is a soft tissue tumor of the nasopharynx, benign in nature but locally invasive and can extend into adjacent anatomic structures. Their most common site is the nasopharynx and commonly occur in male adolescents. Commonly referred to as juvenile nasopharyngeal angiofibroma, it is also known as juvenile angiofibroma of the nasal cavity. It commonly extends into the nasal cavity and pterygopalatine fossa along with the involvement of the nasopharynx. The larger lesions may extend into extra pharyngeal sites as the sinuses including maxillary sinus, sphenoid, and ethmoid sinuses. They can also demonstrate extension through the inferior orbital fissure, and into the masticator space through the infratemporal fossa. The severe form of the disease has also been reported to involve orbit and intracranial invasion.[1]

CASE REPORT

A male patient of age 45 years reported with swelling on the left side of the lower jaw involving the posterior region of the gingiva in relation to mandibular molars extending to the retromolar area [Figure 1]. The duration of swelling was of 2-month history. The patient reported of having pain since the swelling started to enlarge and a small amount of pus discharge. He had some medicines which reduced the pus discharge but no resolution in swelling. On examination, an extra-oral swelling was noted on the left side of the face extending to the lower border of the mandible, which was moderately tender and firm on palpation. Intra-oral examination revealed a large lump associated with mandibular molars on the left side of the jaw, which were firm and obstructing the occlusion.

Orthopantomogram revealed unilocular radiolucency on the left side of the mandible involving the second and third molar periapical approximately 3.5 cm in anterior-posterior dimensions [Figure 2]. The lesion was excised completely along with the second molar tooth. CBCT revealed radiolucency in the left third molar area with osteolytic destruction of bone encroaching inferior alveolar canal

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and perforated lingual plate up to the mylohyoid ridge in relation to the third molar [Figure 3a and b].

HISTOPATHOLOGY

The specimen was of size 3.5 × 3.2 × 2.5 cm, firm in consistency, irregular in shape, and grayish in color [Figure 4]. The specimen was cut into two equal halves to assess the inner contents of the mass or any calcifications in the center of the specimen.

MICROSCOPIC EXAMINATION

Revealed para keratinized stratified squamous epithelium with dense connective tissue stroma. The connective tissue fibers are arranged in a haphazard pattern, made up of fibroblasts with varying cellular shapes as spindle, angular, or stellate-like cells. Connective tissue stroma was infiltrated with numerous blood capillaries and compressed blood vessels. The blood vessel linings varied throughout the lesion from thin fragile linings to vessels with distinct muscle layers suggestive of venous origin. The blood vessels were evenly spaced and some of the vessels are branched giving the appearance of "stag horn." Focal areas show dense chronic inflammatory cell infiltration mainly composed of lymphocytes and plasma cells [Figure 5a].

Immunohistochemical marker analysis with different markers revealed positivity for different cellular components. Stromal cells were strongly positive for vimentin [Figure 5b]. The endothelial linings of vascular channels were positive for CD34 [Figure 5c].

DISCUSSION

The significance of this case report is to be observed under its rarity of site that is intraosseous in the mandible which is rare to its common site of occurrence that is nasopharynx and till now only three cases have been published to the best of our knowledge.[2] Angiofibroma is rare tumors of the maxillofacial region, with the incidence of approx. 0.05–0.5% of head and neck tumors.[3,4] They are highly vascular lesions, considered to be benign in nature but locally invasive and have a high recurrence rate. The aggressive lesions may invade into nasal turbinates, nasal septum, and nasal cavity. The commonest site of extra

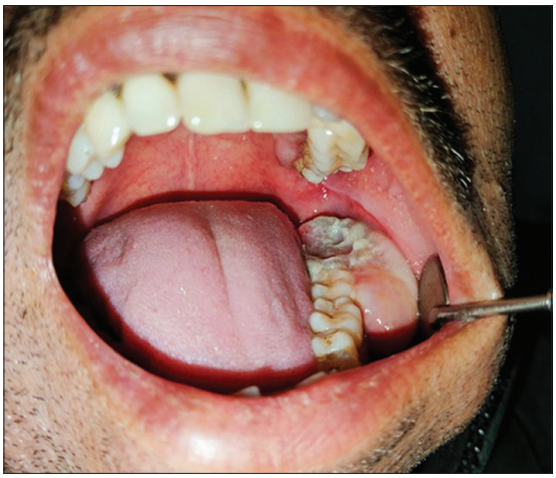

Figure 1: Clinical intra-oral photograph showing the extent of the lesion

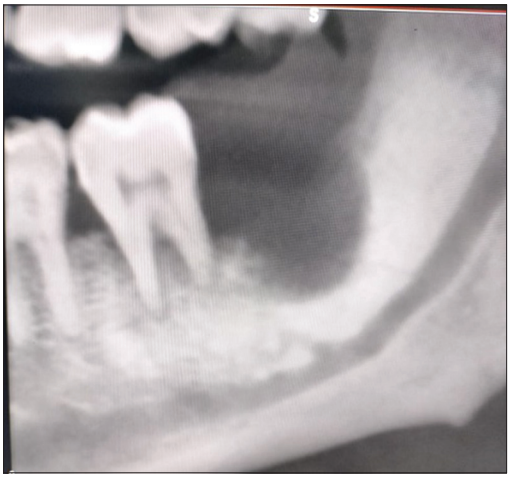

Figure 2: Radiological image showing the extent of the lesion toward the inferior alveolar canal

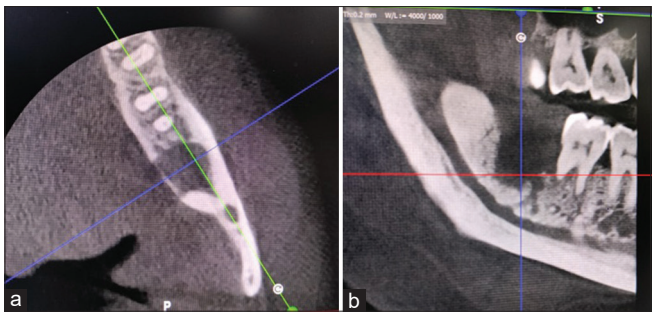

Figure 3: (a and b) CBCT images of the lesion showing the destruction of the lingual plate

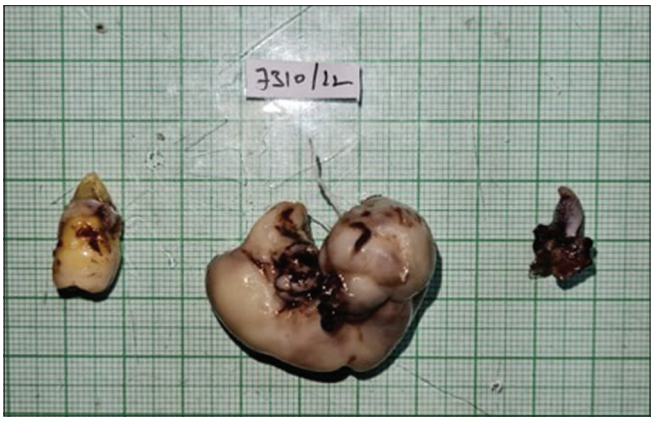

Figure 4: Specimen after complete excision

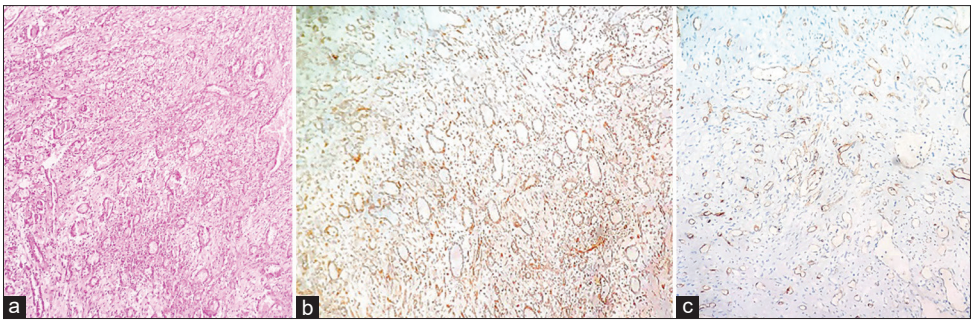

Figure 5: (a)—The H&E section shows the cellular stroma and endothelial linings. (b)—cellular stroma positive for vimentin. (c)—endothelial linings positive for CD 34

pharyngeal lesion is the maxillary sinus. Severe lesions may invade into orbit and intracranial involvement has also been reported.

As angiofibromas are highly vascular lesions, they have one or more arterial vascular associations. The common primary arterial supply is from twig of maxillary artery which is a branch of external carotid artery. The ascending pharyngeal artery is the second most common artery feeding the lesion. Larger lesions may have multiple feeder arteries with additional accessory arteries. The other hypothesis for the occurrence of lesions is remnant of first branchial arch, as incomplete regression of first branchial arch tissue leaves remnants in or near the sphenopalatine foramen.[5]

The site of origin involves sphenopalatine foramen, ala of vomer, and base of lateral pterygoid plate. The tumors that are not arising from sphenopalatine foramen or pterygoid plates are referred as extra nasopharyngeal tumors. Windfuhr *et al*.[6] reported that patients having extra pharyngeal angiofibromas are older in age compared to nasopharyngeal angiofibroma.

The origin of this type of tumor has also been hypothesized to arise from a hamartomatous nidus of hormone-sensitive fibrovascular tissue entrapped in the periosteum.[7] These hormone-sensitive tumors have been shown to express androgen receptors, which are stimulated during the pubertal period by endogenous androgenic hormones. The juvenile age of occurrence of this can be explained by this hypothesis.

The tumor has definitive sex predilection and is almost exclusively seen in males and commonly appears in the second decade of life. The possible cause for the occurrence of extra pharyngeal angiofibroma can be the ectopic tissue located away from the usual site and this may explain the occurrence in the mandible.[8]

Angiofibromas were considered to be benign tumors but the aggressive nature and molecular profiling of these tumors have revealed that the pathogenesis is more complex in nature and these tumors may act as neoplastic. Molecular profiling of these tumors has made an impact on an increased expression of neoplastic markers, including vascular endothelial growth factor-A, FGF, PDGF-A, KIT proto-oncogene receptor tyrosine kinase, avian myelomatosis viral oncogene homolog, and the tumor suppressor gene (TP53).[9]

Treatment

The treatment was done with complete excision of the lesion along with enucleation of the bone cavity. The bone cavity was filled with resorbable collagen and sutured. As in our case no major artery was involved, no frank bleeding occurred. After 6 months of follow up no recurrence was seen and the postsurgical phase is uneventful. Cases where larger blood vessel is involved embolization and ligation of the source artery is required.[10]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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