Case Report

Aggressive angiomyxoma of maxilla: A confounding clinical condition with rare occurrence!

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Abstract

The angiomyxomas are rarely reported in the head and neck region; this paper entails a case of aggressive angiomyxoma presenting as a growth in the maxillary anterior alveolar mucosa and lip region for 1 year, which was accurately identified and treated successfully by surgical excision. An attempt has been made to highlight the clinical and pathologic stand out features of this tumor with intricate emphasis, alongside a literary review.

Keywords: Aggressive angiomyxoma, abundant myxoid matrix, prominent vasculature

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INTRODUCTION

The ability of establishing an apt accurate diagnosis of a presenting ailment or pathology in contemporary clinical practice mandates attainment of ample amount of expertise, so as to harness the holistically acquired observations, clinical presentations investigatory findings into place, and merge them with the diagnosticians wit, wisdom and experience. Thereby unraveling the pathologies of rare occurrence is a furthermore herculean task, because of paucity of their prevalence, and their pathognomonic signs and symptoms, relatively difficult to identify as a stand out feature. One such confounding condition, putting the skills of the clinician to test is myxoma.

The term "myxoma" was coined by Virchow in the first edition of Die Krankhaften geschwülste in 1863.[1] Soft-tissue myxomas are the benign tumors of primitive indifferent mesenchyme, closely mimicking the structure of mucoid connective tissue of umbilical cord.[1-3] The aggressive angiomyxoma is an extremely rare clinical entity, locally infiltrative in nature and of myxoid mesenchymal connective tissue origin with specific predilection for the perineal regions.[4] These are characterized by frequent local recurrences (36%–72%)[5] and show lack of malignant potential.[4-6] Considering their locally aggressive nature, a comprehensive management plan, with a long-term follow-up is necessary.[4,5]

The angiomyxomas are rarely reported in the head and neck region. This paper entails a case of aggressive angiomyxoma presenting as a growth in the maxillary anterior alveolar mucosa and lip region for 1 year, which was accurately identified, and treated successfully by surgical excision. An attempt has been made to highlight the clinical and pathologic features of this tumor, with intricate emphasis on its with literary review.

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CASE REPORT

A 47-year-old male patient reported to the Department of Oral Diagnosis and Radiology, with a presenting complaint of a slowly growing swelling in the maxillary anterior alveolar and lip region for 1 year, which was otherwise asymptomatic, except for its facial asymmetry [Figure 1]. On further eliciting dental history, the patient revealed of having undergone an operation for a similarly presenting lesion 15 years ago, without any complication. His medical history was noncontributory. On examination, lesion was inspected to be single, sessile, well-defined, reddish, ulcerated growth measuring about 4 cm × 3 cm in size extending from the vermillion border of upper lip to the upper alveolus, overlapping incisal surfaces of anterior teeth and was firm in consistency, mildly tender to touch, with slight bleeding on palpation [Figure 2]. Radiographs showed an irregular bone loss pattern, with displacement of teeth in upper anterior region. Based on the history and clinical presentation, a preliminary diagnosis of fibrous epulis/peripheral giant cell lesion and malignancy of upper lip was put forth. When a further radiographic investigation (contrast-enhanced computed tomography scan) was carried out, it validated the presence of an expansile unilocular lesion, which extended into the right maxillary sinus. An altered marrow signal in the underlying alveolus prompted a possible intraosseous invasive component of the lesion [Figure 3]. Next, to substantiate the findings, an incisional biopsy was performed, which revealed hyperplastic stratified squamous epithelium, with underlying areas exhibiting poorly circumscribed tumor tissue, chiefly consisting of spindle- and stellate-shaped tumor cells, arranged in short fascicles, with abundant myxoid matrix and vasculature [Figures 4 and 5]. On immunohistochemistry was positive for vimentin (I: 200 biogenex, cloneV9), calponin (I: 30 biogenex, cone EP7984), CD34 [Figures 6 and 7] and negative for Actin, Desmin, S-100. This could confirm the diagnosis of the pathological entity being aggressive angiomyxoma. As a treatment resort, surgical excision was done under GA with no complications [Figure 8].

DISCUSSION

AA was first described by Steeper and Rosai in 1983 as a slowly growing but nonmetastasizing neoplasm.[4-7] There are three variants of it: aggressive, superficial and angiomyofibroblastoma.[5] The term "aggressive" specifically denotes its propensity for local aggression and recurrences after excision.[4] The etiopathogenesis is still a matter of literary dispute and unclear.[1,4,6] Women are affected more than men [6:1] with a peak occurrence in the fourth decade.[1,7] However, this case report narrated above was that of a male patient, aged 47 years. AA are most frequently found in the vulva, pelvic

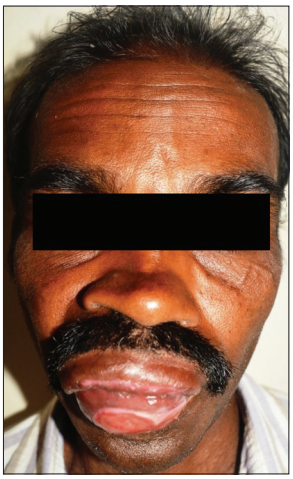

Figure 1: Extraoral preoperative view

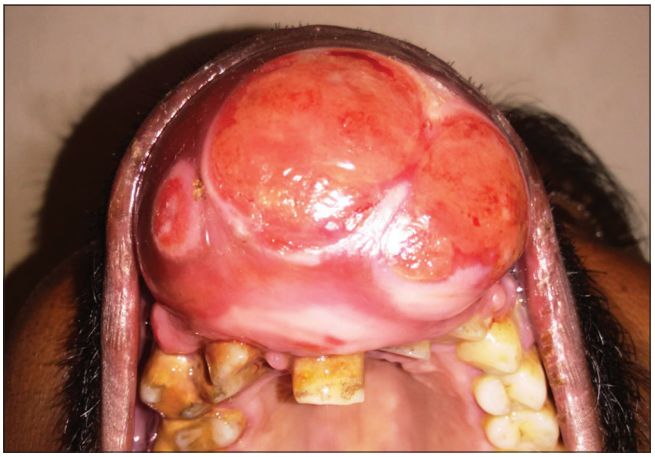

Figure 2: Intraoral preoperative view

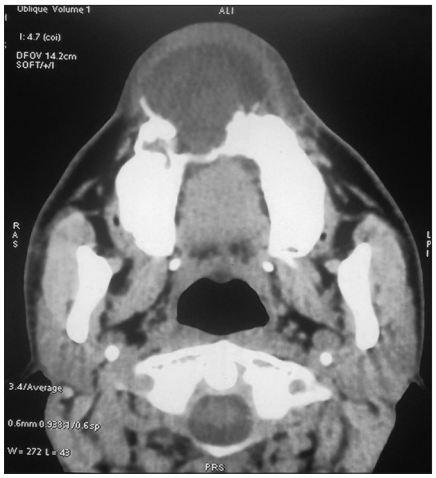

Figure 3: Contrast computed tomography scan showing hypodense anterior maxillary mass

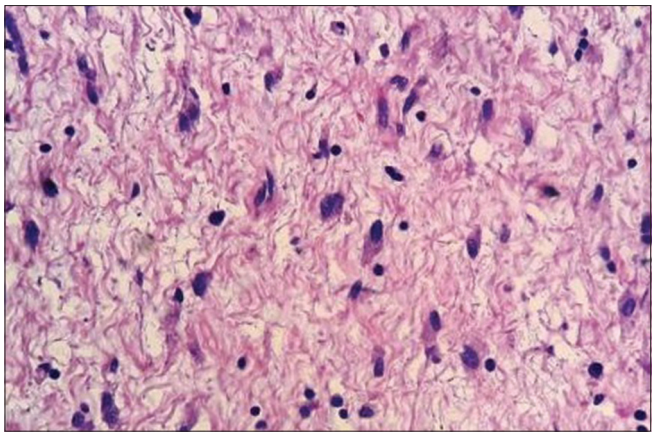

Figure 4: Photomicrograph showing spindle-shaped and stellate cells in a copious myxoid stroma (H & E stain, ×40)

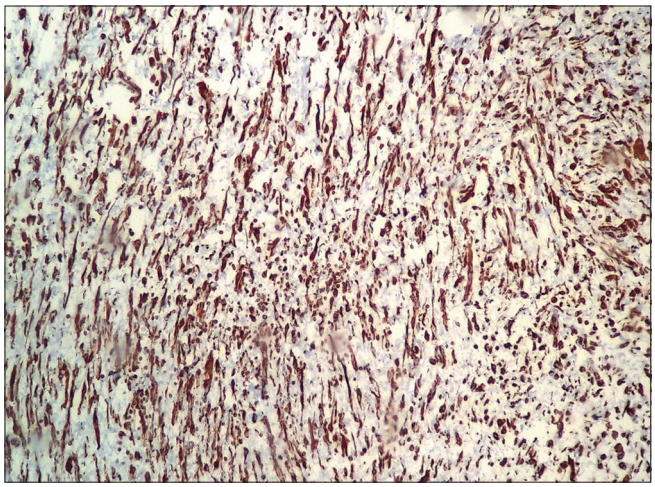

Figure 6: Photomicrograph showing strong immunoreactivity for vimentin (×40)

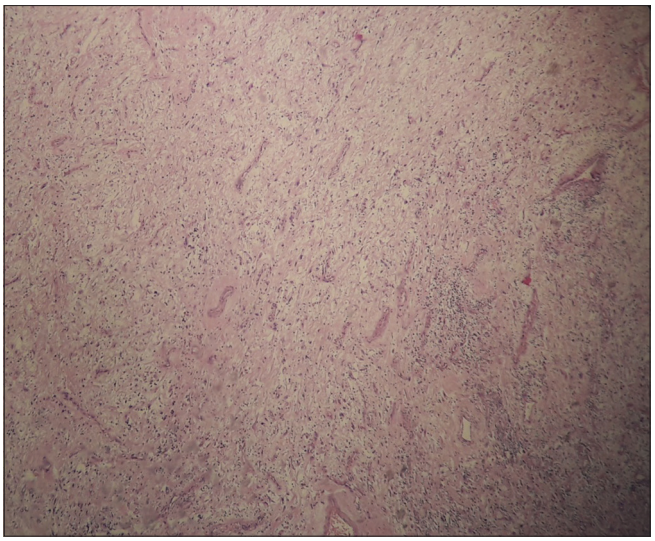

Figure 5: Photomicrograph showing intervening curvilinear blood vessels throughout the myxoid tumor (H & E stain, ×10)

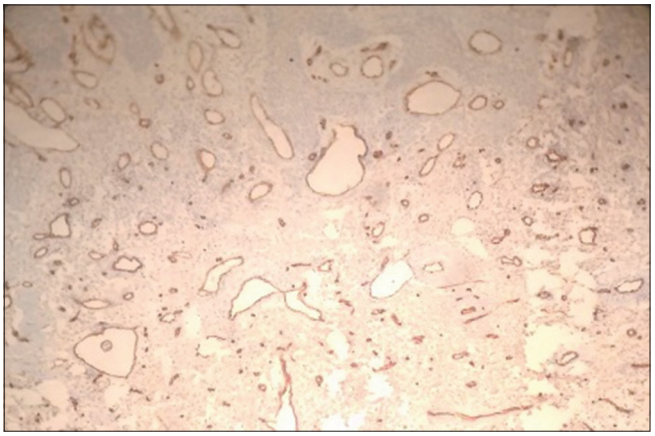

Figure 7: Photomicrograph showing strong immunoreactivity for CD34 (×10)

floor and perineum region[7] and rare in head and neck region. Thereby, a clinical occurrence in upper alveolus and lip region with invasion into surrounding structures conveyed through the manuscript is an instance of notably rarer propensity.

Clinically, these lesions bear clinical resemblance with traumatic fibroma, pyogenic granuloma, lipoma and hence often get misinterpreted. In this case, the lesion presented in the form of an exophytic growth with a sessile base, so the clinical diagnosis was thought of as fibrous epulis, while the differential diagnosis was peripheral giant cell lesion and malignancy of lip, based on its duration, appearance, consistency, tenderness and lymphadenopathy. The distant metastasis is not a common occurrence with aggressive angiomyxoma, while local recurrence is common finding. A similar finding was coherent in this case as well; the patient was operated for same the lesion 15 years before with no metastatic evidence.[6,8] Radiographic findings of this case revealed aggressive nature, with bony destruction in maxillary anterior alveolar bone, and concomitant invasion into surrounding structures, suggestive of its local infiltrative nature.[8]

Microscopically, bulk of the tumor is composed of spindle- or satellite-shaped fibroblasts with poorly defined, palely eosinophilic cytoplasm. The tissue surrounding the cells appears myxoid in some areas and densely collagenous elsewhere. It involves numerous blood vessels, particularly capillaries and medium-sized arteries with thick muscle layer.[1,5,6,8] Similar histological findings were noted in this case report also.

Immunohistochemical staining is consistently positive for vimentin, desmin, CD34, while S-100 protein staining is negative. Begin *et al*. claimed that vimentin stain positive by the intermediate filaments of the tumor, having a fibroblastic

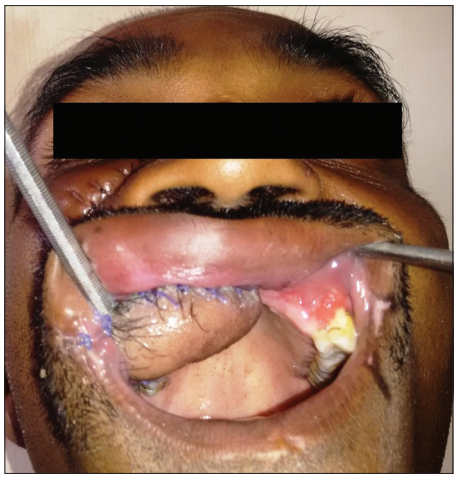

Figure 8: Intraoral postoperative view

origin. Steeper and Rosai showed ultrastructurally that the spindle-shaped stromal cells showed features consistent with myofibroblastic differentiation. Due to these features, the histogenesis of angiomyxomas was traced to be of myofibroblastic or fibroblastic origin precisely.[5,8,9]

The other histological differential diagnoses of possibly myxoid origin could be angiomyolipoma, myxoid lipoma liposarcoma and nerve sheath myxoma.[1,5,7,8] The distinctive histologic features such as prominent vascular component, extensive myxoid areas, absence of mitosis and immunohistochemical findings (strongly positive for vimentin, CD34) of this lesion distinguished it from other myxoid tumors.[5,10-13]

The lack of a capsule and infiltrative growth pattern is responsible for high rate of recurrence. Hence, conservative surgical resection is the treatment of choice with follow-up. The prognosis of this tumor is touted to be good.[1,5,8]

CONCLUSION

Aggressive angiomyxoma of oral cavity is a benign tumor with aggressive nature, and it is not well documented in the literature. Awareness of potential diagnostic pitfalls and careful evaluation of clinical, radiological, histological and immunohistochemistry data are indispensable to derive the correct diagnosis of this myxoid intraoral soft-tissue neoplasm.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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