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Angiomyxoma of the gingiva - A case report and literature review

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Abstract:
Intra oral soft tissue myxoma is an extremely rare, slow growing, benign mesenchymal tumor. Histopathologically, it may be difficult to differentiate from other tumors with myxoid stroma and occasionally misinterpreted as malignancy. We report a case of recurrent intra oral soft tissue swelling in the right maxillary alveolus in an adolescent.
Keywords: Aggressive angiomyxoma, soft tissue myxoma, cavernous hemangioma.

Introduction:
Myxomas of oral and para oral soft tissues have been well documented by Elzay et al.[1,2] Myxomas are benign tumors of primitive undifferentiated mesenchyme, closely mimicking the structure of mucoid connective tissue of umbilical cord. Most soft tissue myxomas are deep lesions, occurring in cutaneous or subcutaneous tissues, genito-urinary tract, gastrointestinal tract, or in organs such as liver, spleen and occasionally within the parotid. An extremely rare lesion, with very few cases documented in the medical literature - an aggressive intra-oral angiomyxoma has been described. Aggressive angiomyxoma mostly develops between the legs or in the pelvis. Nine out of ten people (95%) diagnosed with the aggressive variant are women of childbearing age.[1,2-6] A case of an angiomyxoma of the right maxilla in a 14 year old adolescent is reported here.

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Case report:

A 14- year old female reported to the dental outpatient department with a complaint of growth in the right upper premolar region (Figure 1). History dates back to a year ago when she noticed the swelling, progressively increased in size and had undergone an excision three months later. She was asymptomatic for a short period, but the growth recurred and a second surgical intervention was performed four months later. There were no associated symptoms except bleeding at irregular intervals. Intra orally, a single, sessile, firm, non-tender swelling was present in the right upper premolar region measuring 4 x 3 cms extending from the buccal sulcus, involving premolars and crossing over to the palatal side. Surface appeared erythematous with areas of blanching corresponding to the indendations of the cusps of lower premolars (Figure 2). IOPA revealed mild drifting of premolars proximally with alteration in the trabecular pattern at the alveolar crest (Figure 4). Based on the history and clinical presentation, a diagnosis of pyogenic granuloma / peripheral giant cell lesion was considered. Following an informed consent, an excision was performed. The excised soft tissue section showed hyperparakeratinised and orthokeratinised stratified squamous epithelium with fibrovascular connective tissue. The overlying epithelium was hyperplastic with variable thickness. Intra and inter cellular edema was present. The underlying connective tissue showed dense irregularly arranged collagen fibres with severe chronic inflammatory cell infiltrate predominantly of lymphocytes, macrophages and plasma cells. Focal areas show basophilic structure suggestive of bone. Vascularity was increased with large, dilated blood capillaries lined by plump endothelial cells engorged with RBCs suggestive of infected hemangioma. The post-operative period was uneventful.

Two months later, patient reported back to us with a recurrent growth at the same operated site. Intra orally, the growth was bright red on the buccal aspect and erythematous on the palatal side. Marked proximal displacements of premolars were evident (Figure 3). Ultrasound of upper alveolus and adjacent palatal region revealed a 3.7 x 1.3 cm mass consisting of dilated blood vessels of varying sizes from 1 to 3 mm suggestive of cavernous hemangioma (Figure 5). MRI study of maxilla showed a fairly well defined T2 W hyperintense lesion in the right alveolus of maxilla at the level of premolars measuring about 2 x 1.6 cm suggestive of hemangioma. An altered marrow signal in the underlying alveolus indicates a possible intraosseous component of the lesion (Figure 6). A second surgical intervention was performed and tissue specimen sent for microscopic examination. The lesional connective tissue was highly cellular showing large areas of hyalanized collagen fibres with numerous spindle and stellate shaped fibroblasts. Focal areas of connective tissue showed myxoid changes. Chronic infammatory cell infiltrate were predominantly lymphocytes. Areas of hemorrhage were evident. The overlying epithelium was two to three cell layer thicknesses with irregular rete ridges and edematous changes. These features were suggestive of aggressive angiomyxoma (Figure 7).

Discussion:

The term "myxoma" was coined by Virchow [2,7] in the first edition of Die Krankhaften GeSchwulster in 1863. He further described its histologic appearance in 1871, likening it to the Wharton's jelly of the umbilical cord.[2,8] Myxomas occur in all parts of the body; particularly the soft tissues and bone.[2] Soft tissue myxomas of the oral and perioral tissue are rare and considerably less common than odontogenic myxoma of the jaws (Shimoyama et al. 2000. Chang et al. 2001, Curran et al.2002). The intra-oral myxomas are benign, slowly growing, insidious and potentially infiltrative (Elzay and Dutz 1978, Ramaraj & Shah 2003); in contrast with osseous myxomas, they show a less-aggressive behavior and rarely recur following conservative excision (Barnes 2001, Ramaraj & Shah 2003).[9] The myxomas are derived from early

embryonic primitive mesenchyme which it resembles. According to general norms of tumor behavior, it should therefore occur relatively early in life and be highly malignant. This is true only for the extremely dedifferentiated neoplasms of early childhood, such as the so-called embryonal rhabdomyosarcoma, which consists mainly of primitive embryonal myxomatous connective tissue and displays only occasional better-differentiated cells with cross-striations. These

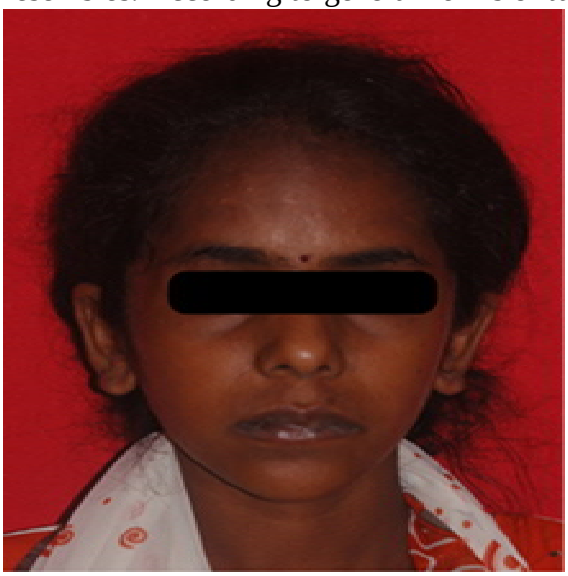

Fig 1: Clinical photograph

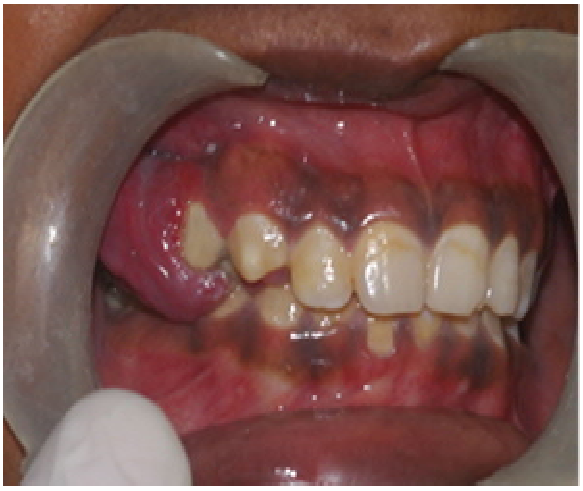

Fig 2: Intra oral primary lesion

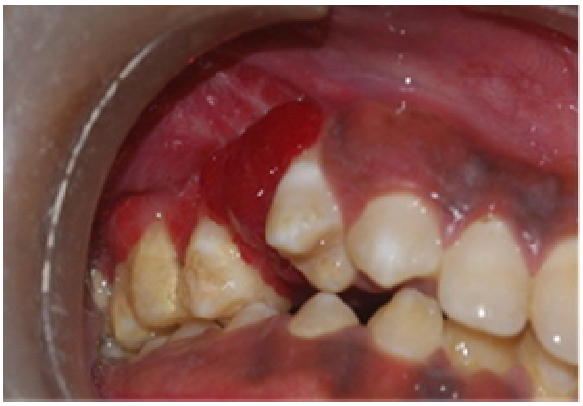

Fig 3: Intra oral recurrent lesion

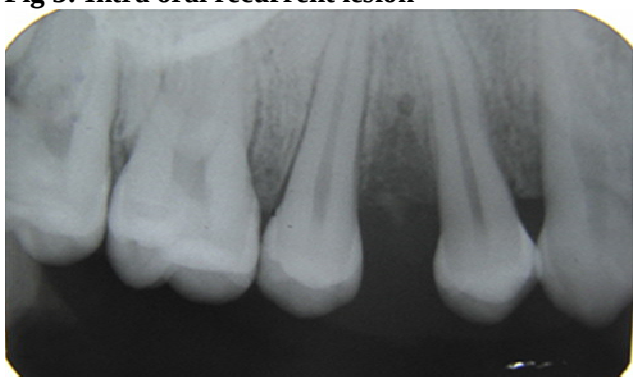

Fig 4: IOPA demonstrating drifting of premolars

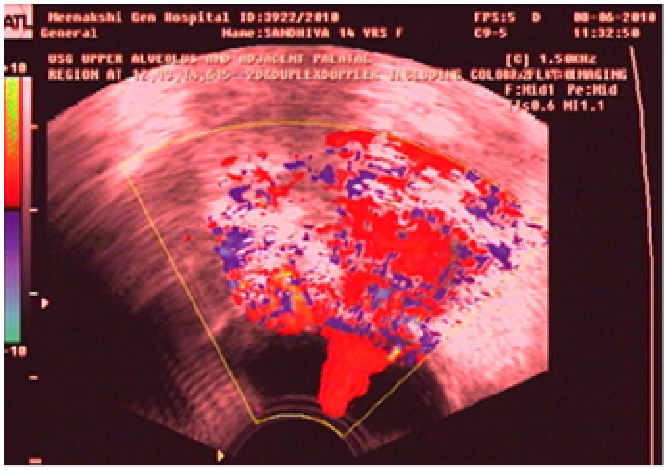

Fig 5: Ultrasonograph

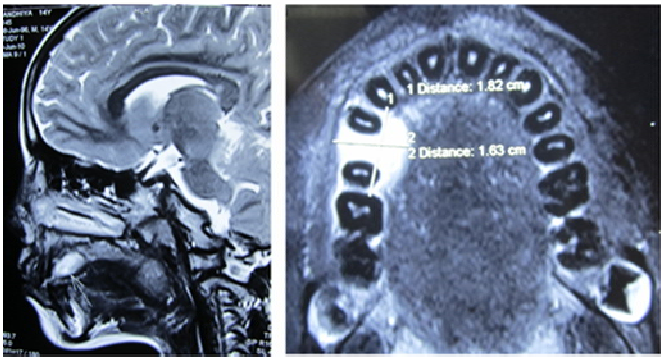

Fig 6: MRI--- T2W Axial and Sagittal

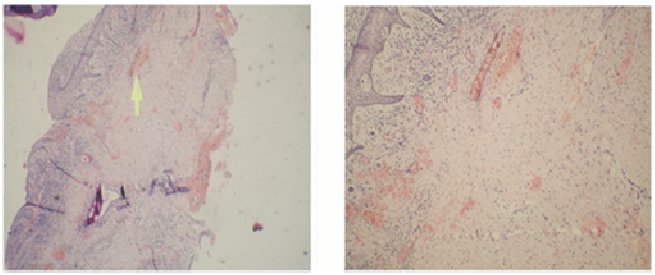

Fig 7: Photomicrograph 4X, 20X

tumors should be more logically classified as embryonal mesenchymal sarcoma.[2]

Tse and Vander (1985) reviewed 43 cases of soft tissue myxoma of the head and neck region. In their study, men were affected more than women (59-41%) and the most common location was the palate, followed by the parotid area. In other studies, other locations like the cheek and the floor of the mouth were reported (Elzay and Dutz 1978, Barnes 2001). These tumors may occur almost in every decade of life, with a peak occurrence in the fourth decade (Tse and Vander 1985. Regezi and Sciubba 1999). They present macroscopically as grey-white, mucoid masses with a smooth or multinodular external appearance and they are usually encapsulated or circumscribed (Barnes 2001).[9]

Several theories concerning the pathogenesis of this tumor were proposed. The prevailing opinion was that altered fibroblasts or myofibroblasts could produce an excess of mucopolysaccharides and were commonly incapable of forming mature collagen even if some cells could retain this capacity (Barnes 2001). Another theory attributed the origin of these tumors to mesenchymal elements derived from dental papilla, dental follicle or periodontal membrane (Tse and Vander 1985, Gunhan et al. 1991, Shimoyama et al. 2000, Chang et al. 2001). However, the histogenesis of these lesions remains obscure and further studies are necessary to clarify its origin. Pathologically, it may be difficult to differentiate from other tumors with a myxoid stroma and is occasionally misinterpreted as malignant (Ramaraj & Shah 2003).[9]

Many characteristics make this myxoma case unusual. First of all, the location: only very few cases of myxoma of gingiva have been reported in the literature (Tahsinoglu et al. 1975, Shimoyama et al. 2000, Chang et al. 2001); moreover, all these were located in the mandibular gingiva, and our case is in the maxillary gingiva. In our case, the clinical differential diagnosis was made in particular with traumatic fibromas (Regezi and Sciubba 1999), extra-osseous odontogenic fibromas, nerve sheath tumors and oral focal mucinosis (Lucas 1998, Barnes 2001). Traumatic fibroma is difficult to differentiate clinically; for this reason, it was our first clinical diagnosis. Microscopically, it is characterized by dense collagen fibres (Regezi and Sciubba 1999). Extra-osseous odontogenic fibroma contains strands of inactive odontogenic epithelium immersed in a stroma rich with collagen (Lucas 1998). However, odontogenic myxomas have a different pathogenesis from soft tissue myxoma, because they arise from primitive mesenchyme of tooth germ, after early induction of fibroblasts into odontoblasts.[9] The other differential diagnosis could include angiomyolipoma and various myxoid tumor such as myxoid lipoma and liposarcoma, nerve sheath myxoma, myxoid neurofibroma, myxoid type of embroyonal rhabdomyosarcoma, myxoid variant of malignant fibrous histiocytoma, and fibromatosis with focal myxoid areas.[1] Although distant metastasis has not occurred with aggressive angiomyxoma, local recurrences were common.[1,10] Conversely, local recurrence has also been reported in superficial angiomyxoma because of incomplete excision.[1,11] This suggests that angiomyxoma is a true neoplasm rather than a hamartoma or a reactive hyperplastic process.[1] Awareness of potential diagnostic pitfalls and careful evaluation of clinical and radiological data are necessary to narrow the differential diagnosis.[9]

Because soft tissue myxoma is a benign tumor, conservative surgical resection is the treatment of choice (Elzay and Dutz 1978, Tse and Vander 1985, Barnes 2001, Kumar et al. 2002, Ramaraj

and Shah 2003). The recurrence rate is 3-8%, and the neoplasm is most likely to recur within 2 years; hence, close follow-up is required (Kumar et al. 2002). The prognosis of this soft tissue tumor is good (Tse and Vander 1985).[9] The tumor is composed of a mixture of myxoid and vascular components, highly resembling an angiomyxoma, a rare mesenchymal myxoid tumor typically including aggressive[1,10] and superficial types.[1,11] Aggressive angiomyxoma, most frequently found in the female vulva, pelvic floor, and perineum[1,4] shows local infiltrative growth and has a propensity to recur.[1,10] Superficial angiomyxoma involves merely the cutaneous and subcutaneous tissues without infiltrating deeper structures; it has been reported in the skin and umbilical cord.[1,12]

Conclusion:

Myxomas present challenges in both diagnosis and treatment. Patients are generally asymptomatic and are first noticed because of progressive swelling and the resultant facial deformity. It is often mentioned that the jaw myxoma has to be distinguished from other tumor entities that may mimic myxoma by an extensive myxomatous degeneration[13-5] diagnostic confusion mostly arising from neurofibroma.[13]

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