Case Report

"Gigantic aggressive angiomyxoma" of the jaws: A rare case report

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Abstract

Angiomyxomas are a group of rare myxoid benign mesenchymal tumors prone to local recurrences. Two types of angiomyxomas are well recognized in the literature: superficial and aggressive angiomyxoma. Aggressive angiomyxomas are relatively uncommon, poorly circumscribed, locally infiltrative myofibroblastic tumors with a specific predilection for the perineal regions and exceedingly rare in the head and neck region. To the best of our knowledge, we report the first case of a gigantic aggressive angiomyxoma occurring in both the maxilla and mandible in a 30-year-old male patient. Clinicians should carefully evaluate the clinical, radiological and histological data to derive the correct diagnosis of this myxoid intraoral soft-tissue neoplasm.

Keywords: Aggressive angiomyxoma, gigantic, recurrence

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INTRODUCTION

Angiomyxomas are a group of rare myxoid benign mesenchymal tumors prone to local recurrences.[1] Two types of angiomyxomas are well recognized in the literature: superficial and aggressive angiomyxoma.[2] Some report a third type known as the angiomyofibroblastoma. Superficial angiomyxoma also known as cutaneous myxoma[3] was first described by Allen *et al.* in 1988. It occurs commonly in the head and neck region, including sites such as the chin, lip, cheek, nose, ear and forehead, but the intraoral occurrence is quite rare. Aggressive angiomyxomas on the other hand are relatively uncommon, poorly circumscribed, locally infiltrative myofibroblastic tumors with specific predilection for the perineal regions.[4] These are characterized by frequent local recurrences (36%–72%),[2] particularly if the tumor is incompletely excised and shows lack of malignant potential.[2,4,5] The occurrence of this type of angiomyxoma is exceedingly rare in the head and neck region. To the best of our knowledge, we report the first case of a gigantic aggressive angiomyxoma occurring in the jaws.

CASE REPORT

A 30-year-old male patient reported to our institution with a huge swelling of the left side of his face and neck for almost 10 years. He was unable to close his mouth, had difficulty in breathing and his chin was deviated to the opposite side. His head was weighed down by the immense weight of the mass and it had to be supported with his hand while sitting or standing. When asked why he turned up so late, he reported that the swelling caused no problems in his daily life until only a few days back when he suffered from difficulty in breathing and spontaneous intraoral

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bleeding. On examination, a gigantic extraoral swelling measuring around 28 cm × 24 cm × 20 cm was seen on the left side of his face and neck [Figure 1]. The lips were wide apart at rest and he was unable to bring them together. Extensive engorged blood vessels and areas of superficial erosion and crustation were visible [Figure 2]. On palpation, the mass was firm in consistency with some fluctuant areas. No pulsations or thrills were felt. The solid bone of the mandible could be felt posterior to the second premolar area on the right side. Intra orally, a lobulated, erythematous swelling was present on the entire alveolar ridge of the left side including the floor of the mouth, elevating the tongue. The swelling was soft and fleshy in consistency. On mild provocation, oozing of blood was observed which regressed after application of firm pressure. The aspiration test was negative. Computed tomography scan of the face revealed a well-circumscribed, multilobular lesion involving the maxilla and the mandible, compressing against the adjacent structures and crossing the midline [Figure 3]. The individual locules were enhanced homogenous areas arranged circumferentially, pointing toward an ill-defined and hazy central radiolucent area. Incisional biopsy was performed from an anterior intraoral site, but severe intraoperative hemorrhage was encountered which was arrested by application of a local hemostatic agent and firm digital pressure. Histopathological report revealed the presence of nondescript epithelium backed by myxoid connective tissue stroma and numerous vessels of varying shapes and sizes, some of which showed perivascular hyalinization. Few areas of myoid differentiation were also noted. All the features were suggestive of aggressive angiomyxoma [Figure 4]. The patient was advised to undergo a digital subtraction angiography (DSA) to determine the presence of feeder vessels. Unfortunately, the patient expired

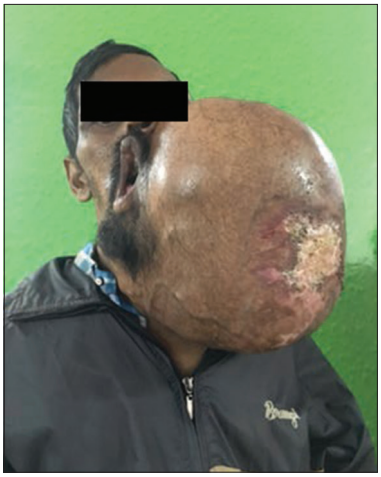

Figure 1: The extensive swelling on the left side of the face and neck

Figure 2: Superficial engorged blood vessels and erosions with crustations, swelling is supported by the patient's hand

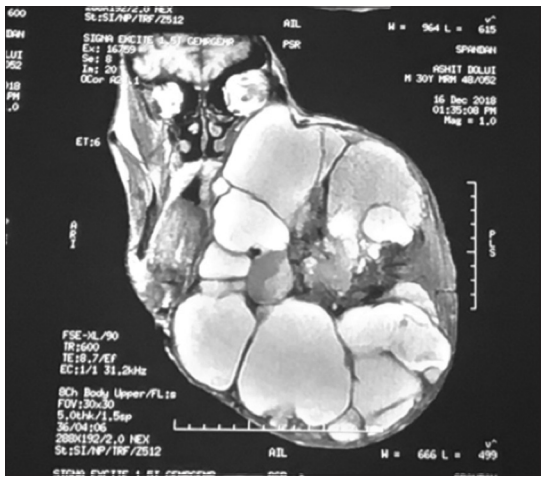

Figure 3: Computed tomography scan of the face

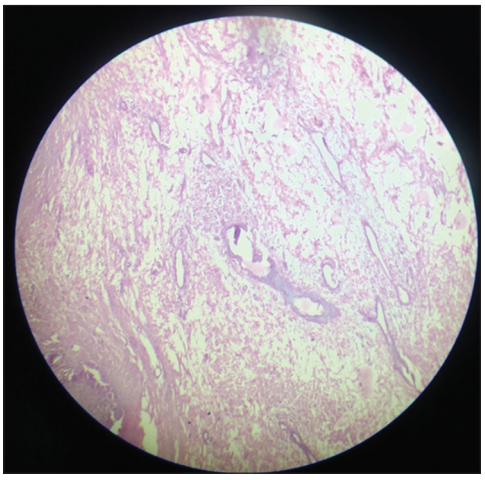

Figure 4: Histopathological features suggestive of aggressive angiomyxoma

during the procedure (DSA) due to a massive cardiac arrest caused most probably by high output cardiac failure.

DISCUSSION

Aggressive angiomyxoma was first described by Steeper and Rosai in 1983 as a slowly growing but nonmetastasizing neoplasm.[2,4-6] The term "aggressive" characterizes its propensity for local aggression and recurrences after incomplete excision.[4] The etiopathogenesis is still a matter of literary dispute and unclear.[4,5,7] The neoplastic cells of this tumor containing estrogen and progesterone receptors, could suggest a hormonal role in pathogenesis. Women are commonly affected with a peak incidence in the fourth decade.[7,8]

However, this case report narrated above was that of a male patient, an exception to the general rule. The most common areas of occurrence are the pelvis and perineum. Occurrence in the head and neck region is exceedingly rare. Hence, a clinical occurrence in both the maxilla and mandible with invasion into surrounding structures conveyed through this manuscript is an instance of notably rarer propensity. Clinically, smaller lesions may bear a resemblance with traumatic fibroma, pyogenic granuloma and lipoma. Larger lesions may be misinterpreted for other neoplasms with myxoid features, chief among which is angiomyofibroblastoma. In this case, the gigantic size, occurrence in both maxilla and mandible and other clinical features made us think of odontogenic myxoma or hematogenous ameloblastoma as potential differential diagnoses. Distant metastasis is not a common occurrence with aggressive angiomyxoma, while local recurrence is a common finding. Radiographic features of this case revealed the aggressive nature of the tumor with bony destruction in both maxilla and mandible and concomitant invasion into surrounding structures, suggestive of its local infiltrative nature.[9] Histopathologically, angiomyxomas are characterized by a proliferation of spindle- or stellate-shaped cells and abundant myxoid stroma associated with a prominent vascular component. The vessels are represented by a large thick-walled muscle layer.[5-8] Similar histological findings were noted in this case as well.

Angiomyxomas are prone to local recurrences due to the lack of a capsule, its infiltrative growth pattern or may recur due to incomplete excision during surgery. Hence, wide excision with clear margins of at least 1 cm should be attempted with long-term follow-up. Recurrences may manifest many years after the initial excision. Due to the unfortunate demise of the patient, surgical excision and long-term follow-up was not possible in this case.

CONCLUSION

Aggressive angiomyxoma is a benign tumor with aggressive nature, and it is not well documented in the literature. To the best of our knowledge, this is the first report of a "gigantic aggressive angiomyxoma" affecting both the maxilla and the mandible. Clinicians should carefully evaluate the clinical, radiological and histological data 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 (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 initial s 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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