CASE REPORT

AN AGGRESSIVE ANGIOMYXOMA .... A 'RAREST OF RARE CASE' WITH CONFOUNDING CLINICAL ENTITY

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ABSTRACT

Angiomyxomas are a group of relatively uncommon myxoid mesenchymal tumours associated with a high risk of local recurrence without any metastatic potential. The angiomyxomas are rarely reported in the head and neck region. This paper entails a case of deep aggressive angiomyxoma presenting as a huge unusual growth in the left side of the face, originating from buccal mucosa, reported to be present for about 15 years, which was accurately identified but unfortunately the subject died before the surgical intervention. An attempt has been made to highlight the clinical and pathologic standout features of this tumour, so that it can be diagnosed and treated properly in future

KEY WORDS

Angiomyxoma, rare, benign

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INTRODUCTION

To establish an apt accurate diagnosis of a disease, growth or pathology in contemporary clinical practice requires attainment of ample amount of expertise and perfect clinical eye and experience, so as to harness the holistically acquired observations, clinical presentations investigatory findings into place, and intermingle them with the diagnostician's wit, wisdom and experience. Unravelling the pathologies of rare occurrence and conclude witha definite diagnosis is a furthermore herculean task, because of rareness of their prevalence, and their pathognomonic signs and symptoms relatively difficult to identify as a stand out feature. One such condition which put the skills of the clinician to test is myxoma. The term "myxoma" was bagged by Virchow in the first edition of Die Krankhaften geschwülste in 1863.[1] Soft-tissue myxomas are the benign tumours of primitive mesenchyme, closely mimicking the structure of mucoid connective tissue of umbilical cord.[1,2,3] The aggressive angiomyxoma is a dreadfully rare clinical entity, which is locally infiltrative in nature and of myxoid mesenchymal connective tissue origin with specific predilection for the perineal regions.[4] They show frequentlocal recurrences (36%–72%)[5] but lacks malignant potential.[4,5,6] Considering their locally aggressive nature, a comprehensive management plan, with a long-term follow-up is necessary.[4,5] These rareentities are infrequent in the head and neck region. This paper presents a case of aggressive angiomyxoma involving buccal mucosa presenting as a growth for 15 years, which was accurately diagnosed but unfortunately the patient expired before the surgery. An attempt has been made to highlight the clinical and pathological features of this tumour.

CASE REPORT

A 30-years-old, male patient, visited the oral and maxillofacial pathology & microbiology unit of Dr. R. Ahmed Dental College and Hospital, Kolkata, West Bengal, with chief complaint of swelling on the left side of his face which was hampering his day to day activity due to its gigantic nature along with

Figure 1: Extra Oral View Of The Growth

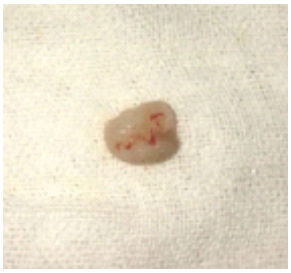

Figure 2: Biopsy Sample From The Lesion

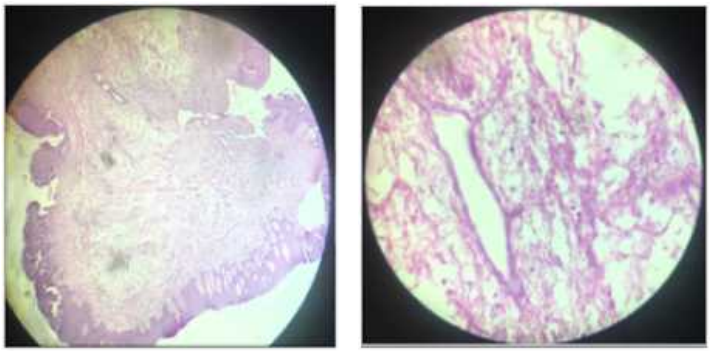

Figure 3: Microscopic Images Under Low And High Magnification Showing A Myxomatous Stroma Consisting Of Spindle Shaped Cells And Numerous Small To Medium Sized Blood Capillaries.

associated facial deformity. On history, patient revealed that the swelling first appeared 15 years back in 2004, which was gradually increasing in size. The extra oral swelling measured about 16 cm × 12 cm in size, firm in consistency and non-tender. The skin over the swelling was pinchable and depigmented and there was a deviation of the mouth in the opposite side. Due to the huge swelling and massive weight, the patient was unable to move his jaws and due to the deformity of the mouth, proper intraoral examination was also difficult. His medical history was non-contributory.

After clinical examination, to substantiate the clinical findings, an incisional biopsy was done from the lesion and the biopsy sample was processed for histopathological examination. Haematoxylin & Eosin stained section revealed presence of non-descript epithelium backed by myxoid connective tissue stroma. Numerous vessels of varying shapes and sizes were noted. Few of those vessels revealed perivascular hyalinization. Few areas of myeloid differentiation were also noted. These features were suggestive of deep aggressive angiomyxoma which was also correlated clinically. The patient was asked to visit the surgery department of any government medical college& hospital for excision of the lesion. Unfortunately, the patient died on the day before the surgery.

DISCUSSION

Steeper and Rosai in 1983 first described Aggressive Angiomyxoma (AA) as a slowly growing but non metastasizing neoplasm.[4,5,6,7] There are three types of it: aggressive, superficial and angiomyofibroblastoma.[5] The term 'aggressive' specifically denotes its propensity for local aggression deep invasion and recurrences after excision.4 The etiopathogenesis is still unclear and a matter of literary dispute.[1,4,6] It is thought to be idiopathic in nature. Women are more vulnerable than men [6:1] mostly found in the fourth decade.[1,7] However, in this case report we got a male patient of early third decade. AA are mostly found in the vulva, pelvic floor and perineum region 7 and is a rare entity in head and neck region. These lesions bear clinical resemblance with traumatic fibroma, pyogenic granuloma, lipoma. The main histologic differential diagnosis includes superficial angiomyxoma, soft tissue myxoma, angiomyolipoma, myxoid nerve sheath tumour, myxoid neurofibroma, odontogenic myxoma, oral focal mucinosis, and myxofibrosarcoma. In this case, the lesion presented in the form of a huge growth with a sessile base, firm consistency, mild tenderness and lymphadenopathy. The distant metastasis is not a common occurrence with aggressive angiomyxoma, while local recurrence is common finding. Microscopically, bulk of the tumour is composed of spindle- or satellite-shaped fibroblasts with poorly defined,

paleeosinophilic 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.[1,5,8] Similar histological findings were noted in this case report also. Steeper and Rosia showed ultra-structurally 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 distinguished it from other myxoid tumours.[5,10,11,12,13] 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 routine follow-up. The prognosis of this tumour is said to be good.

CONCLUSION

Aggressive angiomyxoma is an aggressive benign tumour and it is not well documented in the literature. In our case we were surprised by its gigantic size and deep-seated appearance with a probable poor prognostic result due to its hugeness. Awareness of potential diagnostic pitfalls and a very careful evaluation of clinical, radiological, histological and immunohistochemistry data are inevitable 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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Nil.

CONFLICTS OF INTEREST:

There are no conflicts of interest.

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