Soft tissue fibrosarcoma of pre maxillary region in an adult: report of a unique pathological entity

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Abstract

Fibrosarcoma of the oral and maxillofacial region is a rare entity with poor prognosis. Most common sites are the extremities, with only one percent of fibrosarcoma arising in the head and neck area. Oral fibrosarcoma are locally infiltrative and destructive, spreads by haematogenous dissemination. The positive immunostaining for vimentin, together with negativity for muscular immunomarkers help to diagnosis the fibrosarcoma. Surgical management of fibrosarcoma in maxillofacial region is far from satisfactory, because of lack of inadequate clearance. This paper describes an unusual case of soft tissue fibrosarcoma of premaxillary region in an adult aged 71 years.

Introduction

Fibrosarcoma is a neoplasm of mesenchymal cell origin that is composed of malignant fibroblasts in a collagenous background.[1] Kulander and Bolen[2] defined it as a neoplasm of fibroblasts capable of producing metastatic spread. It can occur in any location, bone extremities being the main affected site. Fibrosarcoma is a very rare malignancy with possible occurrence in the whole head and neck region. It accounts only for one percent of all tumours in this region. Occurrence of fibrosarcoma in the maxillofacial region is extremely rare.[3] If present, the mandibular region is more often involved than the maxillary one.[4] This report describes a patient with soft tissue fibrosarcoma of the anterior maxillary region with clinical, radiographic, pathological and management aspects.

Case Report

A 71-year-old male patient visited Pacific Dental College and Hospital, Udaipur, Rajasthan, with complaints of pain and swelling of upper front region of jaw for the past 12 days (Figure 1). He gave the history of dull, but continuous pain, which aggravated during night time and gradually increasing swelling. Clinically, the lesion was thought to have developed following extraction of an upper anterior tooth 15 days earlier. The tooth had been extracted due to mobility and intense pain. However, the pain persisted even after extraction. The patient noticed the growth of a mass arising from the site of extraction. The antibiotics and analgesics provided no relief and the tissue continued to grow, so rapidly in fact, that 10 days later he could no longer close his mouth without traumatizing the mass. The patient's medical history was non-contributory. He was a chronic alcoholic and used to clean his teeth using tobacco leaves. Patient was moderately built with asymmetrical face. Left submandibular lymph nodes were firm, hard and palpable. Clinically swelling extended from philtrum to outer canthous of eye on the left side of the face. Intraoral examination revealed generalized attrition and periodontitis. Most of the posterior teeth were either carious or grossly decayed. Submandibular lymph nodes were palpable. The orthopantomograph revealed well-defined unilocular radiolucency apical to the extraction socket of maxillary left lateral incisor (Figure 2). Plain and post contrast computed tomography scan of facial bone performed using 3 mm thickness cuts in axial planes. In addition sagittal and coronal reformats were assessed. It showed soft tissue density mass with well-defined margins in midline of face overlying maxillary alveolus causing mild erosions more involving left alar nobule of nose. Mass was extending along the left lateral surface of nose to reach up to medial canthus of left orbit (Figure 3). Right eyeball was small in size showing areas of calcification suggestive of pthisis bulbi. A linear defect was seen involving right nasal bone and frontal process of maxillary bone. A provisional diagnosis of aggressive fibromatosis was made. After the provisional diagnosis a representative portion of target lesion was biopsied by incisional technique under local anesthesia and sent for histopathological examination (Figure 4).

Histopathological examination revealed a fibrocellular stroma with spindle cells arranged in herring bone pattern (Figure 5). The fusiform cells showed hyperchromatic nuclei with dispersed chromatin. Mitoses ranged between 9 to 10 MF/ 10 HPF was evident. Focal areas of necrosis were also noted within the stroma. The tumour cells showed strong cytoplasmic positivity for Vimentin, focal positivity for S-100 protein (Figures 6 and 7) and negative for PanCytokeratin Epithelial Membrane antigen smooth muscle actin and desmin. Based on the clinical, radiographic and immuno-histo-pathological findings a diagnosis of fibrosarcoma, Grade 2/3 (Coindre grading system) was made. After the final diagnosis of fibrosarcoma the whole mass was excised (Figure 8) and along with that left side partial maxillectomy was done (Figure 9). In order to aid in feeding and deglutition an obturator was fabricated (Figure 10).

Radiation therapy and chemotherapy were used as adjuvant treatment. Chemotherapy with etoposide (75 mg/m^2), ifosfamide (1.2 g/m^2), and cisplatin (20 mg/m^2) was given intravenously daily for five days every four weeks. Two months later, the patient had a left partial maxillectomy. In addition, the patient was subjected to postoperative radiotherapy for a period of six weeks. A total of 6000cGy of radiation was given in divided doses. Due to the poor prognosis after one year and two months of the treatment the patient displayed multiple metastases. Unfortunately, the patient died two years after initial diagnosis.

Discussion

The fibrosarcoma is a relatively uncommon entity with unknown origin. They are believed to have arisen either from the periosteum, periodontal membrane, enclaved embryonic mesenchymal cells of developing teeth or cells of the connective tissues surrounding nerves and vessels within the jaw.[5]

Although fibrosarcomas may develop in any mesenchymal tissue in which fibroblasts are present, they are most frequently located in the extremities and the trunk. Fibrosarcoma can

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Key words: fibrosarcoma, maxillary neoplasm, soft tissue sarcoma.

Conflict of interests: the authors declare no potential conflict of interests.

Received for publication: 27 December 2011.
Revision received: 8 April 2012.
Accepted for publication: 7 May 2012.

Clinics and Practice 2012; 2:e58
doi:10.4081/cp.2012.e58

arise in soft tissues or within bone. In the head and neck region, soft tissue sarcomas are extremely rare, accounting for less than 1% of all neoplasms. Fibrosarcomas of soft tissues usually affects a wider age spectrum of patients with an age range of 35-55 years.[6] The present case is a soft tissue fibrosarcoma of premaxillary region in a 71-year-old adult male.

Typically, the tumour presents with swelling, pain, parasthesia, occasionally losing of teeth and ulceration of the overlying mucosa.[7] It may even mimic periodontal lesion with associated bone loss.[4] Facial asymmetry, and in advanced cases, considerable displacement of the eye with resultant diplopia often occurs. The growth rate varies. Local aggressiveness rather than metastasis is a prominent character of this lesion. Soft tissue fibrosarcomas

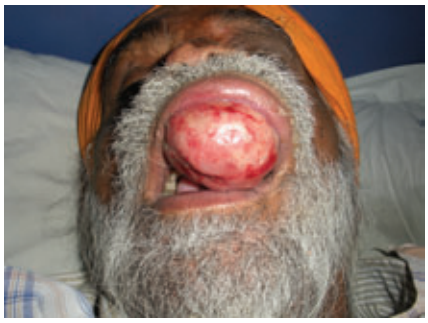

Figure 1. Clinical picture of the patient revealing the mass.

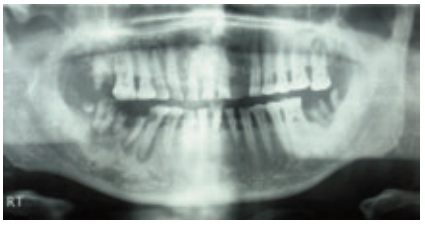

Figure 2. Orthopantomograph revealing well-defined unilocular radiolucency apical to the extraction socket of maxillary left lateral incisor generalized bone loss and retained root stumps.

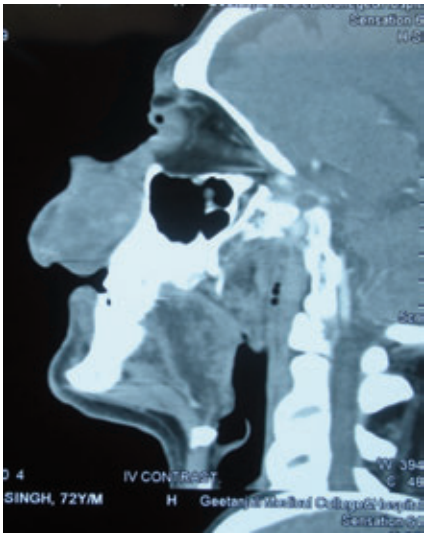

Figure 3. Plain and post contrast computed tomography scan pictures revealing soft tissue density mass with well defined margins in midline of face overlying maxillary alveolus causing mild erosions more involving left alar lobule of nose.

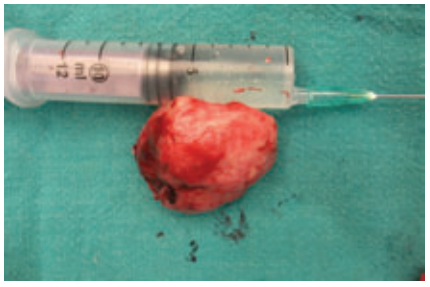

Figure 4. Representative portion of the target lesion was biopsied by incisional technique.

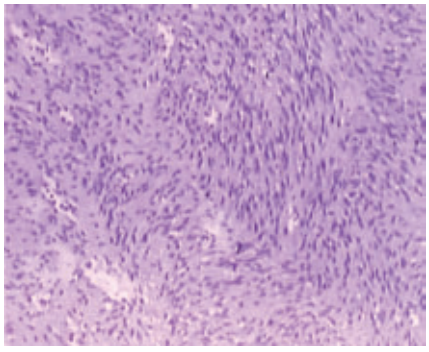

Figure 5. Photomicrograph showing fibro cellular stroma and have the herringbone pattern (H & E, 10x).

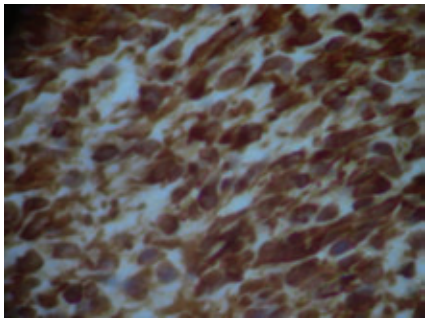

Figure 6. Photomicrograph showing strong cytoplasmic positivity for vimentin.

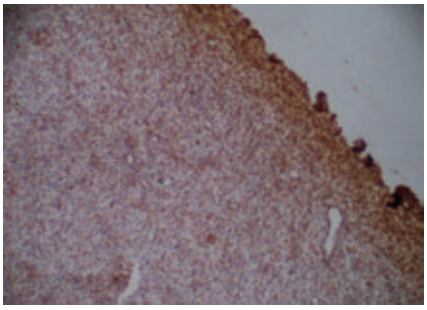

Figure 7. Photomicrograph showing focal positivity for S-100 protein.

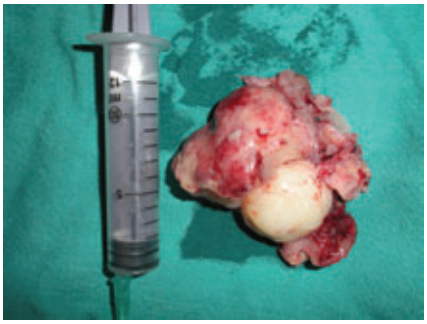

Figure 8. Showing excised mass.

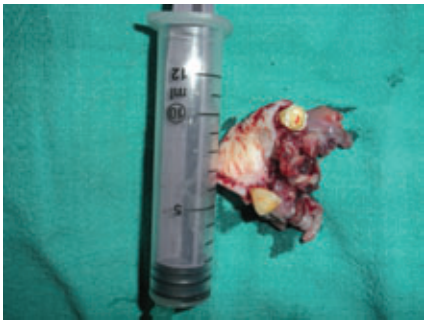

Figure 9. Left side partial maxillectomy.

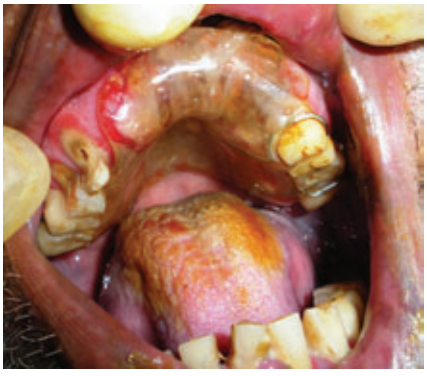

Figure 10. In order to aid in feeding and deglutition an obturator was fabricated.

most often present as painless masses. The duration however is often shorter than with lesions involving the bone. Because these lesions often arise deep in the muscular fascia, they may become extremely large tumors prior to diagnosis.[8] In our case, patient had an asymmetrical face and complained of a painful, gradually increasing swelling which continued to grow so rapidly in fact that, ten days later he could no longer close his mouth without traumatizing the mass. Radiological imaging of fibrosarcomas revealed radiolucent lesions with geographical, moth-eaten or permeative pattern of bone destruction.[7] The absence of tumoural calcification or ossification can be of importance in differentiating fibrosarcomas from other malignancies such as chondrosarcomas and osteosarcomas.[1] However the present case showed a well defined unilocular radiolucency without any calcifications. Histologically, the diagnosis of fibrosarcoma is made much less frequently today because of the recognition and separate classification of other spindle cell lesions that have similar microscopic features. The degree of differentiation is variable, being comparable to a benign fibroma or an anaplastic tumor, becomes a challenging diagnosis.[8] The present case was diagnosed as fibrosarcoma based on the immunohistochemical features. The histological similarity and positive immunostaining for vimentin and S100 protein, together with negativity for pancytokeratin and epithelial membrane antigen helped to confirm the diagnosis of high-grade fibrosarcoma.

The treatment choice to fibrosarcoma is radical surgery with 3 to 5 cm clearance margins. This is not possible in the oral cavity and Sinonasal region because of anatomy and mutilation effect. This restriction leads to failure of local control regardless of the histologic grade and is responsible for their high mortality.[9] The need for adjuvant radiotherapy and/or chemotherapy is still unclear but there is normally an indication in high grade tumours because these tumours may present with subclinical or microscopic metastases at the time of diagnosis. Prognosis is directly related to histological grade, tumor size and adequate surgery treatment with margins free.[3]

Fibrosarcoma is a rare malignancy in the oral cavity; local recurrence is frequent, but metastases are rare. Fibrosarcoma spreads by both local invasion and haematogenous dissemination. A latent period of 10 years before metastasis is quite possible and the commonest site for metastases is lungs.[10]

The present case reaffirms the importance of early investigation of symptoms that are chronic in nature. Any one complaining of pain, swelling, loosening of teeth, parasthesia, lesions changing in character, or progressively ill-fitting dentures should be scrutinized carefully. The dentist must be suspicious of an underlying disease process possibly responsible for these symptoms. Early detection often results in a less extensive and disfiguring surgical course.

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