

FIBROSARCOMA: A MALIGNANT MESENCHYMAL NEOPLASM: A CASE REPORT

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ABSTRACT:

Fibrosarcoma (FS) is a malignant mesenchymal neoplasm of the fibroblasts that is uncommon in the head and neck and constitutes less than 1% of malignancies and approximately 6% of the soft tissue sarcomas. In 1940 Ewing established the initial concept of primary intraosseous fibrosarcoma, and since then the discussion regarding its existence has been going on. Here we report a rare case of fibrosarcoma in 19 year old male patient with manifestation in the maxilla and the diagnosis and treatment plan.

Key words: Fibrosarcoma, herringbone pattern, malignant mesenchymal neoplasm



INTRODUCTION:

Fibrosarcoma is a malignant neoplasm of fibroblastic origin affecting both soft and hard tissue which can occur in any location, with bone extremities being the main affected sites. In 1940 Ewing established the initial concept of primary intraosseous fibrosarcoma, and since then the discussion regarding its existence has been going on.^[1] Intraosseous fibrosarcoma is an uncommon tumor accounting for approximately 5% of all malignant intraosseous tumors. Its occurrence rate is approximately 15% in the cranium, with the mandible being the most common site.^[1] Occurrence in the maxilla is rare, with an incidence ranging from 0% to 6.1%.^[2] here we report a rare case of fibrosarcoma in 19 year old male patient with manifestation in the maxilla.

CASE DETAIL:

A 19-year-old male patient reported to the department of Oral Medicine and Radiology with the chief complain of swelling in the upper jaw of anterior teeth region since 6 months. It started as small swelling in the anterior region of the maxilla on the right side, which has increased in size gradually to attain the present size.(fig1) Medical and family history were noncontributory. On physical examination all the vital sign were with in normal limits and on extra oral examination, a diffuse swelling was present on right side of face in the malar region extending from ala of the nose to middle of cheek anterioposterioly and from infraorbital margin to corner of mouth superioinferiorly measuring about 4X3cm. It is non tender and soft to firm in

consistency on palpation. Intra oral examination revealed a diffuse swelling in the maxillary anterior region extending from 11 to 16 anterioposterioly, the swelling was extended to palatal side causing elevation of palate on right side up to midpalatal raphe, obliteration of vestibular sulcus was present. On palpation Swelling was soft to firm in consistency and nontender. Upper lateral incisor is displaced distally causing spacing between right central incisor and lateral incisor. (fig2, fig3). On the bases of history and clinical features, Ameloblastoma was suspected with differential diagnosis of, dentigenous cyst, AOT, COT. On hematological investigation all the value were within normal limits except for raised eosinophils. Orthopantamograph showed radiolucency in the maxillary anterior region extending from 11 to 16 with patchy radiopacity dispersed in between. Right upper lateral incisor was displaced (fig: 4). Computer Tomography on axial section revealed a heterogeneously enhancing lesion measuring approx. 47x43x35mm involving the upper alveolar arch on the right side to causing destruction of underlying bone. Extension into lower nasal cavity with destruction of intervening nasal floor bilaterally was evident. (Fig 5) Incisional biopsy was performed and histopathological examination under 10x and 40x revealed highly cellular stroma with proliferating fibroblasts. Microscopically the cells showed a typical "herringbone pattern" with sheets of cells arranged in intertwining whorls. Tumor cells showed hyperchromatism, cellular pleomorphism,

and increased mitotic activity. In a few areas the cells were arranged in interlacing fascicles and a moderate amount of collagen was seen. (Fig 6, 7) Based on clinical and histopathological investigation, a low grade fibrosarcoma involving upper alveolar arch was given.

DISCUSSION:

Fibrosarcoma is a malignant neoplasm of the fibroblastic origin. It can occur at any location being the bone extremities the main affected site. In the maxilla occurrences are rare with an incidence ranging from 0 to 6.1% of all primary fibrosarcoma of the bone [3] during 1950 and 1975, it was believed to be the most common soft tissue malignancy. Fibrosarcoma may arise as a primary tumor in any part of the jaws and may be classified as of either peripheral (periosteal) or central (endosteal) type. It frequent effect in the fifth and sixth decades of age but cases in children and adolescents were also described in the literature [4] World Health organization in 2002 defined fibrosarcoma as a "malignant tumor, composed of fibroblast with variable collagen and in classical cases a herringbone architecture [5] clinically fibrosarcoma most often present as an innocuous, lobulated, sessile painless and nonhemorrhagic submucosal mass of normal coloration, fibrosarcoma of intraosseous origin are symptom free until it reaches a considerable size, symptom begin such as swelling paresthesia, pain loosening of the teeth and ulceration of the overlying mucosa [5]

Radiographically Fibrosarcomas have ill-defined borders that are best described as ragged. They are poorly demarcated and noncorticated and lack any remembrance of a capsule. These tumors are generally shaped in a fashion that suggests that they have grown along a bone; therefore they tend to be elongated through the marrow space. The radiographic border may underestimate the extent of the tumor because these lesions typically are infiltrative. If soft tissue lesions occur adjacent to bone, they may cause a saucer-like depression in the underlying bone or invade it as would a squamous cell carcinoma. Finally, sclerosis may occur in the adjacent normal bone whether the fibrosarcoma is peripheral to bone or central. Fibrosarcomas have little internal structure, in most cases the lesions are entirely radiolucent. If the lesions have been present for some time and are not overly aggressive, either residual jawbone or reactive osseous bone formation occurs the most common effect on adjacent structures and the destruction in the mandible, the alveolar process, cortices of the neurovascular canal and inferior border of the jaw, are lost. In the maxilla, the inferior floor of the maxillary sinus, posterior wall of the maxilla, and nasal floor can be destroyed. In either jaw, lamina dura and follicular cortices are obliterated. Destruction of the outer cortical plate is accompanied by a protruding soft tissue mass. Root resorption is uncommon and teeth are more likely to be grossly displaced and lose their support bone so that they appear to be floating in space. In addition, widening

of the periodontal membrane space occurs with this tumor, as in other malignancies. Periosteal reaction is uncommon; but, if the lesion disrupts the periosteum, a Codman's triangle or sunray spiculation may be evident.^[6]

Histopathologically Fibrosarcomas of bone are invasive tumors with no distinct margins. The cells are uniform and spindle shaped and arranged in fascicles, which often forming a herringbone pattern. Histologically, degree of differentiation of this neoplasm are variable, comparable to either a benign fibroma or an anaplastic tumor. Therefore it should be taken to distinguish it from other spindle cell neoplasm. Combined histological and immune-histochemical analysis aid in the definitive diagnosis of such spindle cell lesions. Histological grading of Fibrosarcoma of bone is based on the degree of cellularity, degree of cellular differentiation, mitotic activity, the amount of collagen produced by the tumor cells, and the extent of necrosis. Van Blarcom *et al.*, categorized this lesion into four grades (grade 1 being best differentiated to grade 4 being least well differentiated) on the basis of cellular differentiation by 'Broders' method.^[7] Using the following criteria of amount of fibers, nuclear atypia, and mitotic figures, Taconis *et al.*, graded Fibrosarcomas of jaws into three types. Grade I – well differentiated, less than 2 mitoses in 20 HPF, Grade II – moderately differentiated less than 10 mitoses in 20 HPF, and Grade III – high mitotic rate, 10 or more mitoses in 20 HPF.^[7] Depending on the number of mitotic figures, tumor differentiation, and

the presence of tumor-necrosis, French Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) grading system is currently, the most widely accepted.^[8,9] Most common complication of Fibrosarcoma is frequent local recurrences, which can lead to infiltration, local destruction, airway compression, esophageal compression, and extension into the mediastinum. Although incidence of local and distant metastasis is low, both lymphatic and hematogenous metastasis can occur.^[10,11]

Treatment of choice for Fibrosarcoma of bone is wide resection with clear margin. Prophylactic lymph node dissection is not required. In Fibrosarcomas that cannot be completely excised because of their location or extreme size, postoperative radiotherapy of 6000-7,000 cGy is appropriate. In grade III Fibrosarcomas, postoperative adjunctive chemotherapy is recommended, ostensibly to treat potential subclinical or microscopic metastasis. When chemotherapy is employed, agents used successfully for sarcomas are preferred, including adriamycin, actinomycin D, oncovin, cyclophosphamide, prednisone, and daunorubicin. Apart from the treatment modality, the prognosis of Fibrosarcoma is

significantly influenced by site of origin and histopathological grading of the neoplasm. Unlike soft tissue FS, FS of bone has a poorer prognosis with 5-year survival rate of 4.2-31.7%. A difference in clinical course between FS of jaw and its long bone counterpart has been reported by many authors. A consistent favorable prognosis of Fibrosarcoma of jaw has been observed during the 5, 10, and 20 year observation period as compared with long bone fibrosarcoma. Like most sarcomas, there is a strong correlation between the prognosis of FS and its histologic grade. Low grade differentiation influences the survival rates in a negative way.^[12, 13, 14 & 15]

CONCLUSION:

This is a distinct type of soft tissue sarcoma. Unfortunately, there are no distinct clinical characteristics of fibrosarcoma, and disease is poorly described in the context of all fibroblastic tumors. Fibrosarcoma rarely affects the jaws bone. It is a tumor of mesenchymal origin and the epidemiologic factors are still unknown, but many authors report radiation therapy history as a possible one.

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FIGURES:

Fig: 1



Fig: 2



Fig: 3



Fig: 4

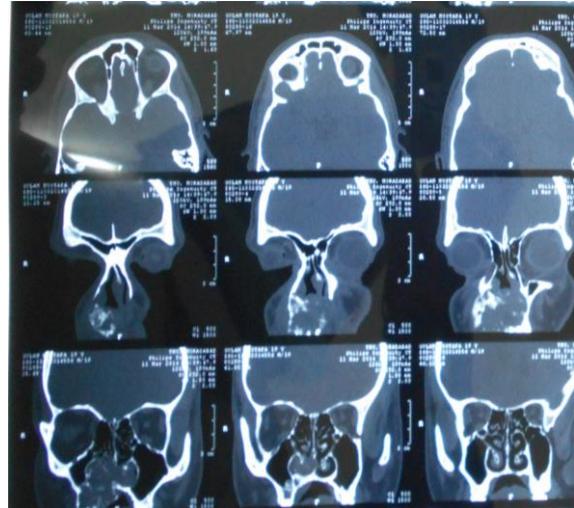


Fig: 5

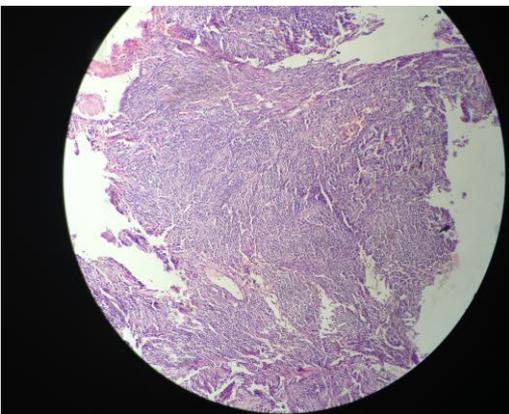


Fig: 6

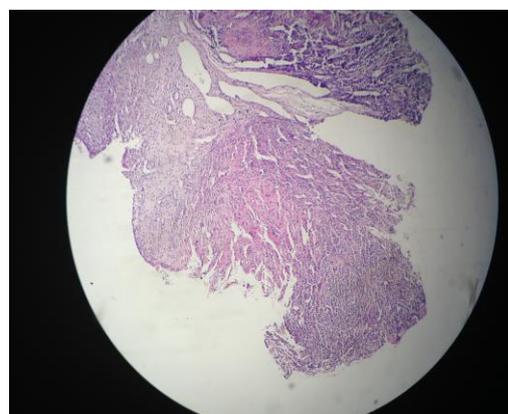


Fig: 7 & Fig: 8. Shows highly cellular stroma with proliferating fibroblasts and the cells showed a typical "herringbone pattern"