



## Fibrosarcoma of Maxilla with Extension into Maxillary Sinus: A Rare Case Report

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### ABSTRACT

Fibrosarcoma is a malignancy of mesenchymal tissue origin and is an unusual finding in oral cavity, moreover, the intraosseous fibrosarcoma is extremely rare to see in jaws. More often it is seen in the mandible as compared to maxilla and rarest to involve maxillary sinus. Whether low or high grade, the tumor is usually aggressive and destructive in biologic behavior, infiltrating the surrounding tissues and with infrequent metastasis. This paper will discuss a rare case of fibrosarcoma of maxilla perforating into the maxillary sinus.

**Keywords:** Fibrosarcoma, Malignancy, Maxilla.

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### INTRODUCTION

Malignancies of fibroblasts are decidedly rare in the oral and oropharyngeal region, but fibrosarcoma is, nevertheless, the most common mesenchymal cancer of the region, representing more than half of all sarcomas.<sup>1</sup> The etiology of fibrosarcoma is not known. There are no specific predisposing factors, although some arise in pre-existing connective tissue tumors and other bone pathologies such as fibrous dysplasia, chronic osteomyelitis, bone infarcts, and Paget's disease. Some recent studies have shown that

alteration in one or more genes in the 2q14-22 region might contribute to its pathogenesis.<sup>2,3</sup>

Twenty-three percent of head and neck fibrosarcomas occur within the oral cavity.<sup>4,5</sup> Intraosseous fibrosarcoma is an uncommon tumor accounting for approximately 5% of all malignant intraosseous tumors. Its rate of occurrence in the cranium is approximately 15%, with the mandible being the most common site.<sup>6</sup> However, occurrences in the maxilla are rare with an incidence ranging from 0 to 6.1%.<sup>7</sup> Interestingly, fibrosarcoma of the maxillary antrum is one of the rarest locations and only few cases have been reported in the literature.<sup>8</sup>

Intraosseous fibrosarcomas may develop endosteally or possibly periosteally, the latter affecting bone by spread from adjacent soft tissue. Occasionally, the lesion erodes the roots of involved teeth.<sup>7</sup>

The tumor affects men and women with equal frequency and can occur in patients of any age but are most common between the third and sixth decades of life.<sup>9</sup>

We report a case of fibrosarcoma of maxilla extending into maxillary sinus in a 63 years old male patient, which is a rare finding.

### CASE REPORT

A 63 years old male patient referred to the Outpatient Department of Kothiwal Dental College and Research Centre, Moradabad, Uttar Pradesh, India with chief complaint of growth in left upper back jaw region. Proper history taking revealed that the patient has undergone for the extraction of root stumps in left posterior maxillary region by the general dental practitioner 1 month back. After few days he noticed a growth in the same region. Initially the size of growth was small but increased rapidly till the date patient reported to the department. In the entire clinical course the growth was asymptomatic except discomfort while mastication and speech.

Extraoral examination revealed diffuse swelling present in relation to left side of face extending anteroposteriorly from ala of nose to the preauricular region and superoinferiorly from zygomatic prominence to superior to lower border of mandible, with no secondary changes of overlying skin (Fig. 1). On palpation the swelling was nontender. Regional lymph nodes were nonpalpable.

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Intraoral examination revealed solitary, sessile, well demarcated, nodular, mixed erythematous and creamy white colored growth left posterior maxillary alveolar ridge area, extending anteroposteriorly from 25 region to back upto left maxillary tuberosity area, mediolaterally from posterolateral aspect of hard palate to buccal vestibule and superoinferiorly measuring approximately 2.5 cm (Fig. 2). On palpation it was soft to firm and nontender. Provisional diagnosis of squamous cell carcinoma was made with salivary gland or vascular neoplasm kept as differentials.

Radiographic investigations were done with the aid of orthopantomograph and PNS projection, which revealed soft tissue shadow of tumor mass with diffuse, infiltrating and destructive border on left side of posterior maxilla extending from 24 region to maxillary tuberosity area. Extension of soft tissue shadow or cloudiness of tumor perforating the maxillary sinus floor is evident in PNS projection. 25, 26, 27 and 28 were missing (Figs 3 and 4).

After routine hematological investigations, incisional biopsy was taken from two different sites of growth and processed for histopathological assessment. Hematoxylin and eosin stained sections revealed the presence of fibrocellular connective tissue stroma composed of sheets of proliferating oval and spindle shaped fibroblasts. Cells were arranged in herring bone pattern with parallel sheets of cell arranged in intertwining whorls. Cells showed hyperchromatism, moderate cellular and nuclear pleomorphism, increased mitotic figures. Also showed presence of few collagen fibers, diffuse chronic inflammatory cell infiltrate and few areas of hemorrhage (Fig. 5). Histopathological diagnosis of intermediate grade fibrosarcoma was made.

Based on the clinicopathological diagnosis, surgical resection was planned and performed. The excised tissue was re-evaluated for the confirmation of diagnosis and revealed the similar histopathological features. So, the final diagnosis was intermediate grade fibrosarcoma.

## DISCUSSION

World Health Organization (WHO) in 2002 defined fibrosarcoma as a 'malignant tumor, composed of fibroblasts with variable collagen and, in classical cases, a herringbone architecture'.<sup>10</sup>

Clinically, fibrosarcoma most often presents as an innocuous, lobulated, sessile, painless and nonhemorrhagic submucosal mass of normal coloration, however, sometimes may be a rapidly enlarging, hemorrhagic mass similar in clinical appearance to an ulcerated pyogenic granuloma and peripheral giant cell granuloma.<sup>1</sup> Intraosseous fibrosarcoma is symptom-free until it reaches a considerable



Fig. 1: Clinical photograph showing extraoral diffuse swelling on left side of face



Fig. 2: Clinical photograph showing soft tissue growth in left posterior maxilla alveolar ridge area mixed erythematous and creamy white appearance

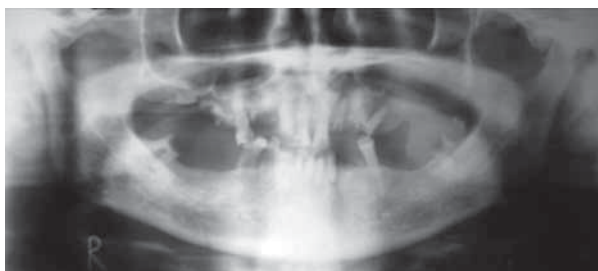
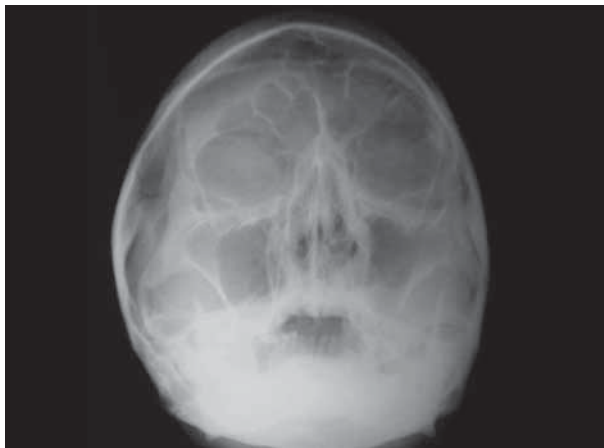


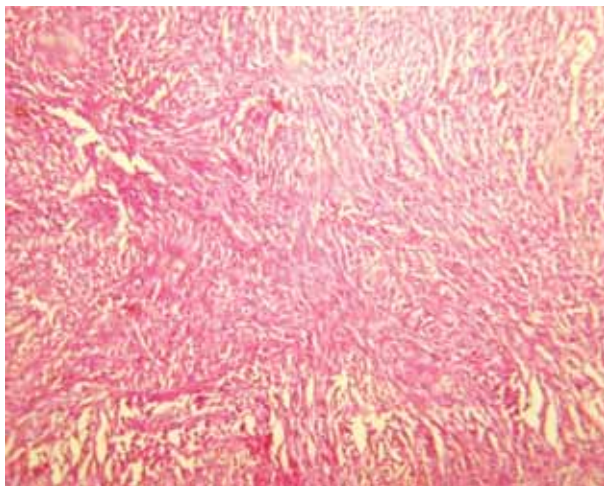
Fig. 3: Orthopantomograph showing diffuse soft tissue shadow of tumor mass extending from 24 region to left maxillary tuberosity area

size, symptoms begin such as pain, swelling, paresthesia, loose teeth, and ulceration of the overlying mucosa.<sup>6</sup> In present case the patient was asymptomatic with classical clinical findings.

Radiographically, the tumor appears like an osteolytic lesion, with ill-defined borders, thinning and disruption of the cortex, resulting in soft tissue invasion.<sup>6,7</sup> as we have seen in the present case.



**Fig. 4:** PNS projection showing cloudiness in left maxillary sinus due extension of tumor



**Fig. 5:** Photomicrograph showing typical herring bone pattern (H&E, 400x)

Microscopically, fibrosarcoma has got variable presentation due to variation in degree of cellularity, mitotic activity, pleomorphism, and giant-cell formation, which has led to histological grading of the tumor. Grade I (well-differentiated) refers to tumors of uniform nuclear appearance with an appreciable amount of collagenous intercellular substance. Mitotic figures are rare or absent and giant cells are not observed. In grade II (intermediate) tumors there is more cellularity and less intercellular substance. Nevertheless, the nuclei, though more closely packed, are still fairly uniform in size, shape, staining properties, and showing typical herring bone pattern. Occasional mitotic figures are seen. Grade III (high) tumors are anaplastic cellular growths often containing many giant cells and mitotic figures.<sup>3,11</sup> Histological grading is important to do, as it has direct correlation with the prognosis of tumor.

The diagnosis can usually be made by routine light microscopic examination of hematoxylin and eosin

(H&E) stained sections as it was in present case; however, occasionally it is difficult to distinguish fibrosarcoma from other malignant neoplasms such as malignant fibrous histiocytoma, fibroblastic osteosarcoma, leiomyosarcomas, lymphomas and metastatic disease. This cognitive state of diagnostic dilemma demands immunohistochemistry, that adds to the pathologist's diagnostic techniques when trying to distinguish morphologically similar tumors. Fibrosarcoma stains strongly positive for the intermediate filament vimentin. Markers for muscle (desmin, smooth muscle actin, HHF-35), human osteoblasts (osteocalcin), macrophages (CD-68), leukocyte common antigen (LCA), neural tissue (s100, neuron specific enolase), melanoma (HMB-45), neutrophils (CD-31), hematopoietic cells (CD-34), epithelial tissue (cytokeratin, epithelial membrane antigen), and CD-99 will be absent. Thus, fibrosarcoma is essentially a diagnosis of exclusion.<sup>2,12-15</sup>

Wide surgical excision is generally advocated for fibrosarcoma. Radiotherapy and chemotherapy can be used in inoperable cases or as a palliative treatment. Although recurrence is not uncommon, metastasis is infrequent. Intraosseous tumors are more likely to metastasize via bloodstream than are soft tissue lesions. The overall 5-year survival rate ranges between 30 and 50%.<sup>2,7</sup> To determine the prognosis it is important to do clinical staging and histological grading.

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