

Fibrosarcoma of Maxilla

¹Pradkhshana Vijay, ²Manas Bajpai, ³Nilesh Pardhe, ⁴Mohd Saleem

ABSTRACT

Fibrosarcoma is a rare malignant neoplasm of mesenchymal and fibroblastic origin rarely affecting head and neck region. It reveals anaplastic fibroblastic proliferation of spindle-shaped cells. The tumor is aggressive and causes infiltration of surrounding tissues with distant metastasis. Fibrosarcoma of oral cavity mostly involves mandible frequently and rarely maxilla. We report a case of fibrosarcoma of maxilla in a 45-year-old male patient who presented with a rapidly enlarging swelling of maxillary alveolus with ulceroproliferative growth causing maxillary arch constriction. The case report with a brief review of literature of the tumor has been discussed.

Keywords: Aggressiveness, Malignancy, Surgical excision, Ulceroproliferative growth.

How to cite this article: Vijay P, Bajpai M, Pardhe N, Saleem M. Fibrosarcoma of Maxilla. Oral Maxillofac Pathol J 2018; 9(2):101-103.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Fibrosarcoma is one of the most common soft tissue malignancies. It mostly affects the extremities and is rare in the head and neck region. It accounts for 1 to 3% of all sarcomas.¹ In the oral cavity, mandible is frequently involved. Involvement of maxilla is rare. No such predisposing factors are present and may arise from existing soft tissue pathology or any other bone pathology like Paget's disease or fibrous dysplasia.² Recent studies have shown genetic alteration in 2q14-22 region, contributing to its pathogenesis.²

Fibrosarcoma of maxilla occurred in any age with no specific age predilection. Common symptoms include pain, swelling, loosening of teeth, and sometimes ulceration over the mucosa. Tumor can invade the sinuses and cause obstructive symptoms.³

CASE REPORT

A 45-year-old male patient presented with a painless swelling in the right posterior maxillary region since 3 months. The swelling was rapidly enlarging and measured 3.5 × 4 cm in size. No significant contributory family or medical history was present. Extraoral inspection revealed swelling, which was extending superoinferiorly from infraorbital region to upper lip and anteroposteriorly from mid tragus region to commissure of right lip. Swelling was oval in shape with well-defined border. Skin over the swelling appeared stretched but was of normal color as of adjacent skin mucosa. Swelling was firm and indurated. Also, the lesion was involving right hemimandible (Fig. 1). Intraoral examination revealed an ulceroproliferative growth in the maxilla extending from maxillary second premolar buccolingually to tuberosity area posteriorly, leading to constriction of maxillary arch (Fig. 2). The second permanent molar was missing, which was extracted 1 year back as it was mobile. The buccal cortical plates were expanded and there was displacement of 16.

Radiographic findings using orthopantomogram revealed radiolucent lesion involving posterior right maxilla and maxillary sinus, which leads to erosion of bone and displacement of 16 and root resorption. A well-defined radiolucency was seen involving 31 to 34, causing resorption and displacement of these teeth. Also revealed was a well-defined radiolucency involving anterior



Fig. 1: Extraoral examination reveals swelling of right posterior maxillary extending superoinferiorly from infraorbital region to upper lip and anteroposteriorly from mid-tragus region to commissure of right lip

^{1,4}Senior Resident, ²Associate Professor, ³Professor and Head

^{1,4}Department of Oral Pathology and Microbiology, King George's Medical University, Lucknow, Uttar Pradesh, India

^{2,3}Department of Oral Pathology and Microbiology, NIMS Dental College, Jaipur, Rajasthan, India

Corresponding Author: Pradkhshana Vijay, Senior Resident Department of Oral Pathology and Microbiology, King George's Medical University, Lucknow, Uttar Pradesh, India, Phone: +919829493322, e-mail: vijaydhar38@yahoo.in



Fig. 2: Intraoral examination reveals ulceroproliferative growth in the maxilla extending from maxillary second premolar buccolingually to tuberosity area posteriorly leading to constriction of maxillary arch

mandible (Fig. 3). Other blood investigations and chest X-rays were normal. Incisional tissue biopsy was done and was sent for histopathological evaluation.

The histopathological examination revealed fibrocellular connective tissue stroma with spindle-shaped

cells varying in size and shape. The cells were arranged in bundles, fascicles, and few areas showing a classic "Herringbone pattern." These cells were dysplastic in nature and were showing cellular and nuclear pleomorphism, nuclear hyperchromatism, and increased nuclear cytoplasmic ratio. Bizarre mitotic figures about 2 to 4 per field were noted. Also the presence of few collagen fiber bundles was noted (Fig. 4). Based on the histopathological findings, a confirmative diagnosis of intermediate fibrosarcoma was made.

Following this, the patient was referred for surgical excision, but the patient refused the treatment.

DISCUSSION

The World Health Organization has defined fibrosarcoma in 2002 as malignant tumor comprising fibroblasts with variable collagen, and in classical case Herringbone architecture.¹ It is a malignant tumor of mesenchymal origin. The tumor is rare and accounts for 5% of malignant intraosseous neoplasms and mainly affects the long bones; 0.05% of cases occur in head and neck region, of which 23% are affecting the oral cavity.⁴ Mostly it occurs

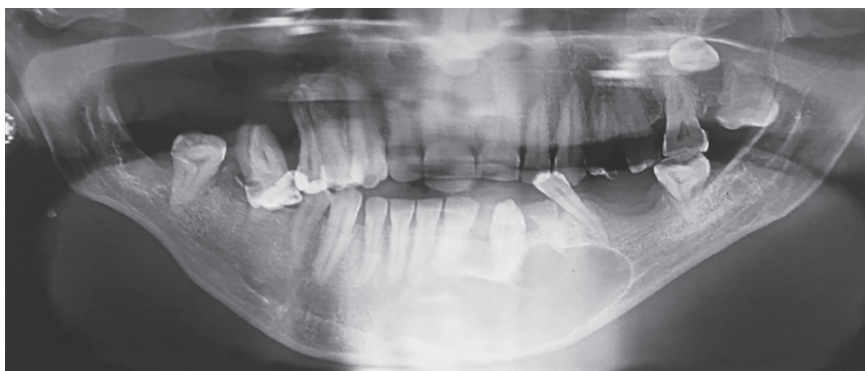
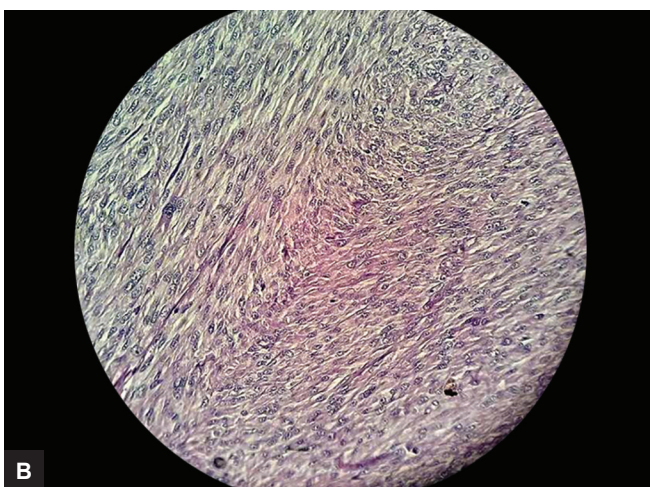
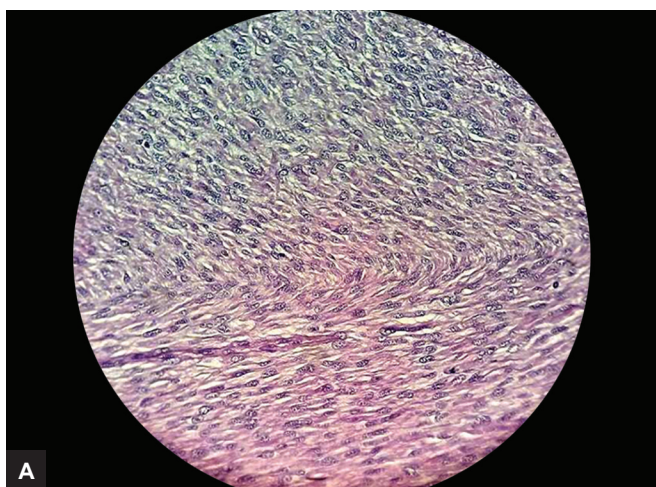


Fig. 3: Orthopantomogram reveals radiolucency involving posterior right maxilla involving maxillary sinus. Also a well-defined radiolucency involving anterior mandible is noted



Figs 4A and B: Histopathological examination reveals cells were arranged in bundles, fascicles, and few areas showing a classic Herringbone pattern. Few mitotic figures are noted

in the fifth to sixth decade of life with equal sex predilection. But few studies have shown male predominance, which is similar to present case occurring in 45-year-old male patient. Mandible appears to be a common site of occurrence compared with maxilla in the oral cavity. We are reporting a case of fibrosarcoma of maxilla, which is quite rare. Primary fibrosarcomas occur in jaws as central (endosteal) or peripheral (periosteal) lesions. Secondary fibrosarcomas occur as a complication of fibrous dysplasia, Paget's disease, osteomyelitis, or bone infarct. Also, it could be due to malignant transformation of giant cell tumor of bone or an outcome of prior irradiation,⁵ though no such contributory history was present in our case.

The cause of fibrosarcoma is not clearly understood, but may be associated with genetic alterations. Chromosomal alterations have been associated with some fibrosarcomas. Localized radiotherapy can contribute to increased risk of fibrosarcoma. Other factors could be tissue damage by heat and scarring. Paget's disease and osteomyelitis have also been implicated as one of the causative factors.^{6,7}

Clinically, the patients appear to be asymptomatic and only 30% cases present with symptoms like jaw swelling with or without associated pain, loose teeth, and paresthesia. Usually, it presents as a lobulated, sessile, nonhemorrhagic mass of normal mucosal coloration.⁴ But, aggressive fibrosarcomas present as rapidly enlarging, hemorrhagic mass that can clinically resemble pyogenic granuloma, peripheral ossifying fibromas, or peripheral giant cell granuloma.¹ Our case presented with a rapidly enlarging swelling, which was painless with ulceroproliferative surface. Radiographically, it presents as osteolytic lesion with ill-defined margins, thinning and destruction of cortex with soft tissue invasion.³ Our case presented with a well-defined radiolucency with displacement of associated tooth and root resorption.

Histopathologically, fibrosarcomas are invasive lesions with indistinct margins. The cells are uniform and spindle-shaped arranged as fascicles in a classic Herringbone pattern. The degree of differentiation varies and care should be taken to differentiate it from other spindle cell tumors.^{2,3}

Grade I (low) refers to tumor of uniform nuclear appearance with collagenous intercellular substance. Mitotic figures are rare. In grade II (intermediate) there is more cellularity and less intercellular substance. Nuclei are packed, but still have uniform shape and size with a typical Herringbone pattern. Mitotic figures are noted. Grade III (high) tumors are anaplastic with numerous mitotic figures.⁵

Fibrosarcoma shows a strong expression of vimentin and is a diagnosis of exclusion. Malignant tumors that can be considered in its differential diagnosis are

monomorphic synovial sarcoma, spindle cell carcinoma, malignant fibrous histiocytoma, leiomyosarcoma, ameloblastic fibrosarcoma, amelanotic melanoma, and malignant peripheral nerve sheath tumor.⁷

Monomorphic synovial sarcoma with spindle cell component shows expression of vimentin, cytokeratin, and cluster of differentiation 99, absence of which rules its diagnosis. Spindle cell carcinoma shows a biphasic pattern with strong positivity for pan-cytokeratin and epithelial membrane antigen. Malignant peripheral nerve sheath tumor shows spindle-shaped cells having high mitotic rate and serpentine nuclei with cells expressing S-100. Fascicles of spindle cells with blunt vesicular nuclei and strongly positive smooth muscle actin and vimentin are noted in leiomyosarcoma. Amelanotic melanoma poses a problem but expression of HMB-45 differentiates it. Spindle cell component with presence of odontogenic islands helps to differentiate ameloblastic fibrosarcoma.⁷

The treatment of choice is wide surgical excision. Radiotherapy with chemotherapy can be advocated as palliative treatment.² Prognosis depends on histologic grade, tumor side, and appropriate surgical treatment with tumor free margins. Survival rate ranges from 20 to 35%.³

CONCLUSION

Fibrosarcoma is malignant tumor of fibroblastic origin, rarely affecting maxillofacial region. Histopathology plays an important role in diagnosis, especially in cases that lack typical Herringbone pattern architecture in which immunochemistry helps to attain a confirmatory diagnosis and advocating an adequate treatment.

REFERENCES

1. Wadhwan V, Chaudhary MS, Gawande M. Fibrosarcoma of the oral cavity. *Indian J Dent Res* 2010 Apr-Jun;21(2):295-298.
2. Sood N, Sehrawat N. Central primary fibrosarcoma maxilla—a rare presentation. *Otolaryngology (Sunnyvale)* 2016 Jun;6(3):237.
3. Khanna S, Singh NN, Singh A, Purwar A. Fibrosarcoma of maxilla with extension into maxillary sinus: a rare case report. *Oral Maxillofac Pathol J* 2014 Jan-Jun;5(1):462-465.
4. Malati T. Tumour markers: an overview. *Indian J Clin Biochem* 2007 Sep;22(2):17-31.
5. Enzinger, FM.; Weiss, SW. Fibrosarcoma. In: *Soft tissue tumors*. 2nd ed. New Delhi: The C V Mosby Company; BI Publications Pvt Ltd; 1990. pp. 201-22.
6. Swain N, Kumar SV, Dhariwal R, Routray S. Primary fibrosarcoma of maxilla in an 8-year-old child: a rare entity. *J Oral Maxillofac Pathol* 2013 Sep;17(3):478.
7. Yamaguchi S, Nagasawa H, Suzuki T, Fujii E, Iwaki H, Takagi M, Amagasa T. Sarcomas of the oral and maxillofacial region: a review of 32 cases in 25 years. *Clin Oral Investig* 2004 Jun;8(2):52-55.