

Malignant Fibrous Histiocytoma of Maxilla

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ABSTRACT

Malignant fibrous histiocytoma (MFH) is an aggressive soft tissue sarcoma occurring in the late adulthood. It may rarely arise as a primary neoplasm in bone, but usually in the long bones and accounts for less than 1% of the malignant tumors of bone. Its presentation in the facial bones and skull is extremely rare. Malignant fibrous histiocytoma of bone is a highly malignant tumor that recurs, metastasizes, and commonly results in death despite aggressive surgical therapy. This article documents the clinical, radiographic and pathologic features of malignant fibrous histiocytoma of maxilla in a 45-year-old female patient and discusses the considerations related to the diagnosis.

Keywords: Malignant fibrous histiocytoma, Soft tissue sarcoma, Maxilla.

How to cite this article: Pazhani A, Jimsha VK, Khatri M, Ramesh V, Daniel MJ, Srinivasan SV. Malignant Fibrous Histiocytoma of Maxilla. *J Indian Acad Oral Med Radiol* 2013; 25(1):77-81.

Source of support: Nil

Conflict of interest: None declared

INTRODUCTION

Malignant fibrous histiocytoma (MFH) was initially described in 1964 by O'Brien and Stout and was considered it to have a histiocytic origin.¹ It is a common soft tissue sarcoma in adults occurring in the 5th to 7th decades of life and accounts for 10.5 to 21.6% of all soft tissue malignant neoplasms.² The tumor rarely occurs in the bone, commonly in the metaphysis of long bones, such as the femur and tibia and its occurrence in the maxillofacial region is very rare,^{3,4} accounting for only 3 to 10% of all cases.⁵ Males are affected more often than females (in the ratio of 9:4).⁵ Malignant fibrous histiocytoma of maxilla or mandible usually presents as a painful swelling with significant facial asymmetry. The etiology of this tumor is unknown but seems to be multifactorial. Genetic background, environmental factors such as trauma,⁶ radiotherapy^{1,7} and malignant transformation from benign lesions has been suggested to be involved in the etiology of this tumor.⁸

CASE REPORT

A 45-year-old, woman reported to the Department of Oral Medicine and Radiology, Mahatma Gandhi Postgraduate Institute of Dental Sciences, Puducherry, India with a chief complaint of swelling in relation to right side of face for past 2 months, associated with pain for approximately 1 month, over which time the swelling had progressively

enlarged. She also gave a history of nasal obstruction. The patient reported a past history of trauma 25 years back in relation to the right cheek and extraction of right anterior tooth 20 days back for complaint of persistent tooth ache and after extraction the swelling has rapidly increased in size. The patient had no harmful habits.

On extraoral examination, a swelling of approximately 4 × 4 cm was noted in relation to right cheek with loss of nasolabial fold and a significant asymmetry of right middle one-third of the face. The surface of the swelling was smooth and there was no sinus or pus discharge. On palpation the swelling was firm in consistency and tender (Fig. 1). One firm and tender right submandibular lymph node was also present.

Intraoral examination revealed a swelling in relation to right anterior maxilla approximately 4 × 3 cm, extending beyond the midline by 1 cm and on the mucosa overlying the swelling, an ulcer measuring approximately 1 × 1 cm covered by yellowish slough was seen (Fig. 2). On palpation, swelling was soft to firm in consistency, tender and also segmental mobility of anterior maxilla was elicited. The teeth in the vicinity of swelling were grade 2 mobile and depressible into their socket.

On the basis of history and clinical findings, a provisional diagnosis of benign odontogenic tumor probably ameloblastoma was given. The clinical differential diagnosis included benign conditions like central giant cell granuloma, Pagets disease of bone and fibrous dysplasia and malignant tumors like osteosarcoma, central malignant tumor of minor salivary gland, carcinoma of maxillary antrum and metastatic tumor of the jaw.



Fig. 1: Diffuse swelling in relation to right cheek with loss of nasolabial fold

Thermal vitality test was performed and the teeth 14, 15, 16 and 17 were found to be nonvital. Aspiration was performed with a sterile needle of 0.5 mm and was found to be nonproductive.

Intraoral periapical radiograph showed a radiolucency in relation to right maxilla with loss of lamina dura in relation to the teeth 15, 16 and 17 and there was no evidence of root resorption of the involved teeth (Fig. 3).

Panoramic radiograph revealed complete destruction of floor of the right maxillary sinus (Fig. 4) and on maxillary occlusal view a radiolucency was seen crossing the midline with deviation of nasal septum to the left side (Fig. 5). Also a significant expansion of the buccal and palatal cortical plates was observed. Paranasal sinus view revealed haziness in the right maxillary sinus and destruction of lateral wall and floor of right maxillary sinus (Fig. 6). Axial and coronal computed tomograms showed homogeneous mass in the right maxilla invading right maxillary sinus and thinning of right infraorbital margin (Figs 7 and 8). The chest radiograph revealed a normal study (Fig. 9).

An incisional biopsy was performed under local anesthesia and the lesional tissue was sent for histopathologic examination. Hematoxylin and eosin sections were highly cellular in nature basically showing two types of cells predominantly-spindle type and round clear cells. The cells were basically hyperchromatic, pleomorphic in nature having a storiform pattern of arrangement with fibrous structures in adjoining areas (Fig. 10). Active mitotic figures were also seen. Histopathology of the lesion was suggestive of malignant fibrous histiocytoma.

Complete resection of the tumor followed by post-operative radiotherapy and chemotherapy were recommended and patient was referred to higher center for management. Patient failed to undergo treatment and unfortunately died 5 months after the initial diagnosis.

DISCUSSION

Malignant fibrous histiocytoma (MFH) is a pleomorphic soft tissue sarcoma.^{9,10} It is considered to be a neoplasm with fibroblastic nature and facultative histiocytic differentiation. The tumor was first recognized in 1964 by O'Brien and Stout,¹ who considered it to have a histiocytic origin. However, it has remained a controversial



Fig. 3: Intraoral periapical radiograph showing radiolucency in relation to right maxilla with loss of lamina dura of teeth 15, 16 and 17



Fig. 4: OPG showing erosion of floor of right maxillary sinus



Fig. 2: Swelling of the right maxilla crossing the midline and with surface ulceration

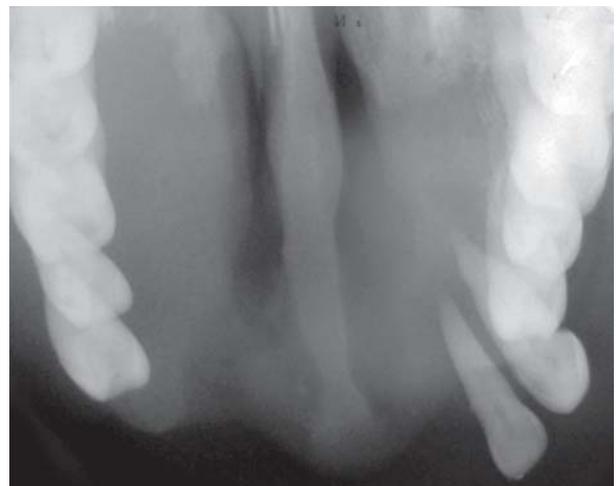


Fig. 5: Maxillary occlusal view revealing an ill defined radiolucency crossing the midline and deviation of nasal septum to the left side

entity because of its uncertain histogenesis. Weiss et al described this tumor with four types of histologic presentation; storiform pleomorphic, myxoid, giant cell and inflammatory.^{1,8} The storiform-pleomorphic type is the most common subtype of malignant fibrous histiocytoma occurring in head and neck, followed by the myxoid type. Feldman and Norman for the first time in the 1970 described primary malignant tumor of bone that satisfied the histopathologic criteria of malignant fibrous histiocytoma.

As a primary neoplasm in bone, it commonly occurs in the metaphysis of long bones of extremities, such as the femur and tibia and its occurrence in membranous bones is quite unusual. The head and neck region is an uncommon site for this malignant tumor.^{1,9,11} In the head and neck, malignant fibrous histiocytoma has been reported to involve the nasal cavity and paranasal sinuses most frequently, accounting for 30% of all cases. In the sinonasal tract, it occurs most commonly in the maxillary sinus, followed by

the ethmoidal sinus, nasal cavity, sphenoid sinus, and frontal sinus. Other reported sites in the head and neck include the craniofacial bones (15-25%), larynx (10-15%), soft tissue of the neck (10-15%), major salivary gland (5-15%), oral cavity (5-15%), pharynx, ear and eyelid.¹ In our case malignant fibrous histiocytoma has occurred as a primary neoplasm in the right maxilla and invaded the maxillary sinus.

The etiology of this tumor is unknown but seems to be multifactorial.⁸ Genetic background, environmental factors such as trauma,⁶ radiotherapy^{1,7} and malignant transformation from benign lesions has been suggested to be involved in the etiology of this tumor.⁸ A history of antecedent trauma has been reported in about 20% of the cases suggesting that some of these tumors may represent an initial proliferative response to trauma.⁶ So, trauma may be the etiologic factor in this case.

The tumor has got a male predilection¹² and commonly occurs in late adulthood.^{10,12,13} Maxilla is more commonly



Fig. 6: Paranasal sinus view showing haziness of right maxillary sinus

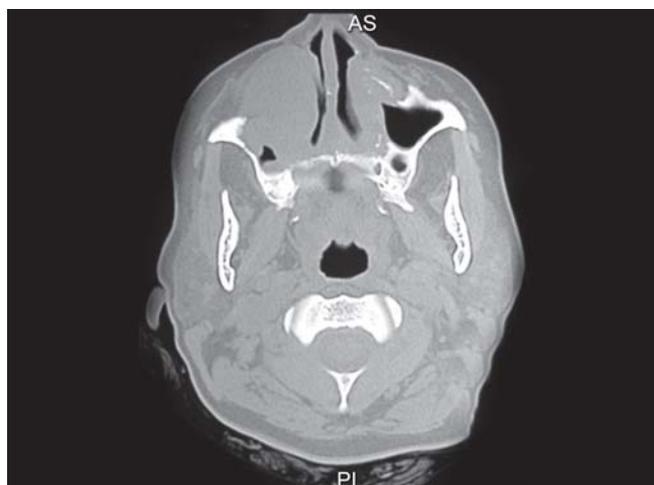


Fig. 8: Axial computed tomogram showing a homogeneous mass in relation to right maxilla invading the right maxillary sinus

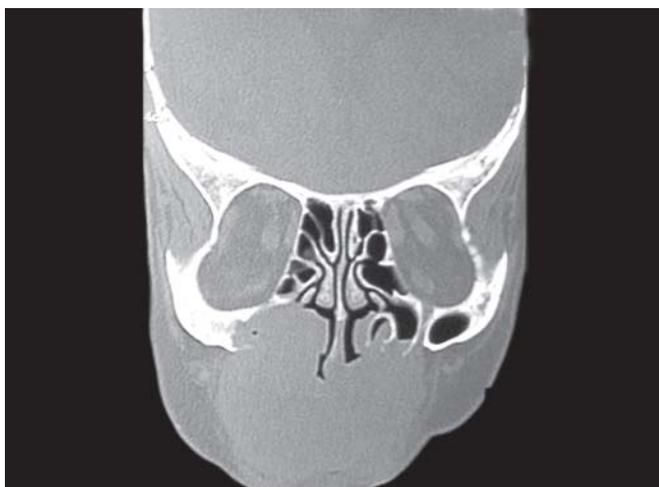


Fig. 7: Coronal computed tomogram showing a homogeneous mass in relation to right maxilla invading the right maxillary sinus

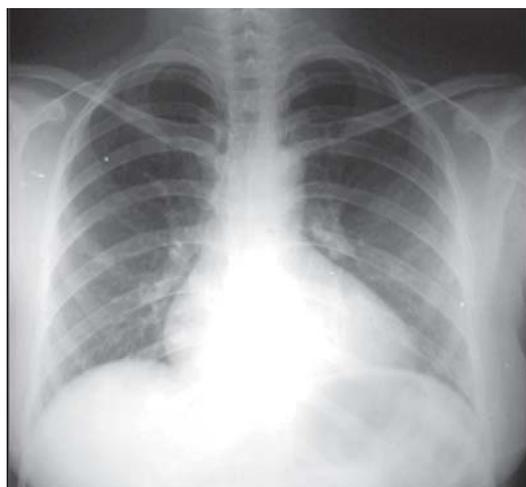


Fig. 9: Chest radiograph showing a normal study

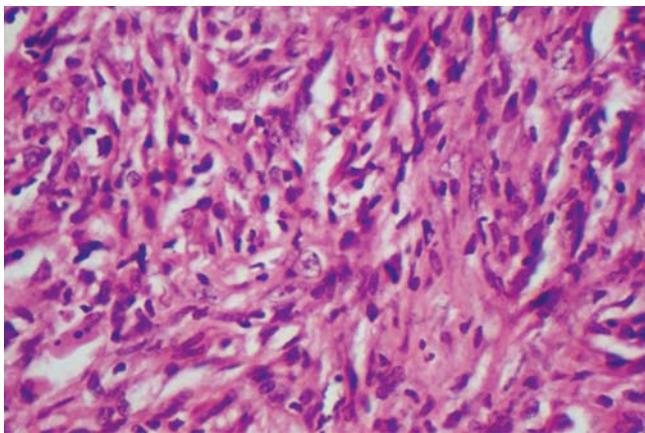


Fig. 10: Hematoxylin and eosin section showing spindle type and round clear cells with a storiform pattern of arrangement

involved than the mandible. Clinically, the common signs and symptoms of malignant fibrous histiocytoma occurring in maxillofacial region consist of a gradually progressing swelling that may be tender on palpation and the surface of the swelling may or may not be ulcerated.⁹ Fracture may be the first sign when the tumor occurs as a primary neoplasm in bone. In cases where the tumor is involving the maxillary sinus, there may be associated symptoms like nasal obstruction, nasal discharge and epistaxis associated with the onset of the swelling.¹⁴ Less frequently, the presenting symptom may be a persistent tooth ache or nonhealing extraction socket. According to Kanazawa et al (2003), clinical symptoms were usually present from 2 weeks to 6 months before diagnosis.³ In our case, the patient presented with a gradually progressive swelling in relation to right cheek which was painless initially. Pain was associated with the swelling later when there was ulceration of the surface of swelling involving the right maxilla. The patient had the associated symptom of nasal obstruction suggesting invasion of the tumor into the maxillary sinus. Radiographically, most of the reported lesions have presented as an extensive ill defined radiolucency without marginal sclerosis and periosteal reaction similar to the present case.

Malignant fibrous histiocytoma shows an aggressive biological behavior and exhibits a high incidence of local recurrence and metastasis. As a primary neoplasm of bone, it aggressively infiltrates adjacent tissues this can be responsible for a high recurrence rate. In our case, the computed tomogram has revealed the invasion of the tumor into the right maxillary sinus and also thinning of right infraorbital rim.

According to Huvos et al, metastatic spread in patients with primary tumor in bone occurs via hematogenous dissemination, predominantly to the lungs,¹⁵ rather than to regional lymph nodes. Metastases more commonly occur in the lungs (90% of metastasis) followed by lymph nodes (12%), bone (8%) and liver (1%).⁸ About 5% of cases already reveal metastasis when the primary tumor is

diagnosed. In our case, there was nodal involvement but no metastasis to lungs.

The management of malignant fibrous histiocytoma of bone consists of surgical resection with wide margins followed by radiotherapy and chemotherapy.² The treatment modalities vary depending on the size of the tumor and the preference of the surgeon.¹⁶ The prognosis of this tumor is often unfavorable and recurrence rate is approximately 44 to 48%. The 5-year survival rate of the tumor is reported to be 36.5 to 53%. Close follow-up and monitoring after treatment is also important since recurrence is a frequent problem and early metastasis to the lungs is common. Malignant fibrous histiocytoma of jaws commonly result in death despite aggressive therapy.⁸

CONCLUSION

This case report has presented the clinical, radiographic and histopathological features of malignant fibrous histiocytoma of maxilla, as well as a review of diagnostic and management considerations. Although the tumor is rare in the oral cavity, this lesion should always be considered in the differential diagnosis of tumors of jaw bones because of its aggressive biologic behavior, distant metastasis and a comprehensive treatment required for its management.

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