

Malignant Fibrous Histiocytoma of the Mandible: A Rare Case Report with Review of Literature

¹T Dinesh Kumar, ²M Rekha, ³RS Neelakandan, ⁴TR Sarawathy

ABSTRACT

Introduction: Malignant fibrous histiocytoma (MFH) is the most frequent soft-tissue sarcoma of adulthood. The occurrence of MFH in membranous bones, including the mandible, is quite unusual and involvement of mandible accounts for only 3% of all MFH bone lesions. Around only 33 cases have been reported in the world literature (period of literature search: 1985–2016) prior to the present case.

Case presentation: A 19-year-old male presented with a complaint of diffuse swelling in the left mandibular angle and ramus region. Radiographs and computed tomography (CT) scan showed extensive osteolytic areas. Segmental resection of the mandible was done and was reported as MFH.

Treatment and prognosis: Recurrences were noted first in the postauricular region, then in mandibular chin, and then in maxilla, which were also surgically treated but the prognosis was poor.

Clinical implications: The behavior of MFH is more aggressive than that of fibrosarcoma and osteosarcoma of mandible with a high rate of local recurrences and frequent metastasis.

Conclusion: We report a rare case of MFH occurring in the mandible of a 19-year-old male with emphasis on the need for aggressive management of these lesions.

Keywords: High tumour recurrence, Malignant fibrous histiocytoma, Mandible, Metastasis.

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INTRODUCTION

Malignant fibrous histiocytoma (MFH) was first described as a new malignant tumor by O' Brian and Stout in 1964. The name MFH is proposed for a variant of fibrous histiocytoma in which mono or multinucleated cells of bizarre malignant appearance are added to histiocytic and fibroblastic background. It is relatively rare in the head and neck region, where the most commonly affected sites are the sinonasal tract, craniofacial bones, larynx, and soft tissues of the neck. Involvement of mandible accounts for only 3% of MFH bone lesions,^{1,2} and lesions occurring in the larynx, maxillary sinus, and mandible have the worst prognosis.⁷ Around only 33 cases have been reported in the world literature³⁻⁶ prior to the present case. The biological behavior is highly aggressive with a high local recurrence rate and metastasis most common to the lungs and carries a poor prognosis.

CASE REPORT

A 19-year-old male presented with a complaint of swelling in the left mandibular angle and ramus region since 1 month and pain since 2 days. On general examination, the patient was found to have kyphoscoliosis (present from birth). There was diffuse swelling measuring 5 × 8 cm involving the left mandibular body and angle extending onto the ramus region (Fig. 1). The overlying skin was stretched and shiny and the lesion was firm in consistency with a few fluctuant areas. Intraorally, there was a diffuse intrabony swelling in relation to 36, 37, 38 involving both



Fig. 1: Extraoral swelling measuring 5 × 8 cm involving the left mandibular body and angle extending onto the ramus region

^{1,2,4}Professor, ³Principal, Professor and Head

¹Department of Oral and Maxillofacial Pathology, RVS Dental College & Hospital, Coimbatore, Tamil Nadu, India

²Department of Oral and Maxillofacial Surgery, Sri Venkateswara dental college & Hospital, Pondicherry, Tamil Nadu, India

³Department of Oral and Maxillofacial Surgery, Meenakshi Ammal Dental College & Hospital, Chennai, Tamil Nadu, India

⁴Department of Oral and Maxillofacial Pathology, Chennai, Tamil Nadu, India

Corresponding Author: T Dinesh Kumar, Professor Department of Oral and Maxillofacial Pathology, Trichy, Tamil Nadu, India, e-mail: dr_dineshmds2007@yahoo.co.in; and dineshrekhakhavya2008@gmail.com

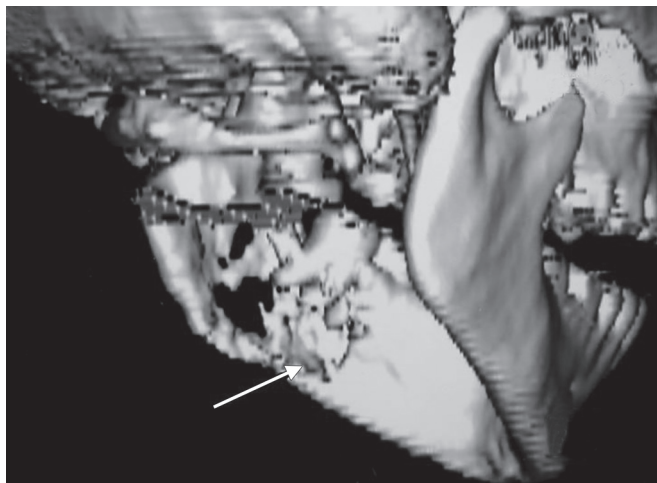


Fig. 2: Three-dimensional (3D) CT scan showed extensive osteolytic area in the left angle and ramus of the mandible measuring 3.54×2.34 cm

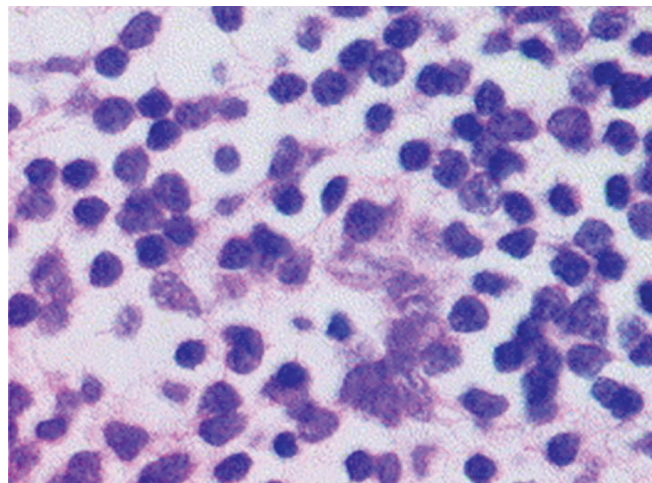


Fig. 3: Photomicrograph, high power showing tumor cells with varying degree of pleomorphism, hyperchromatism, mitotic activity resembling histiocytes-like cells, and spindle cell elements

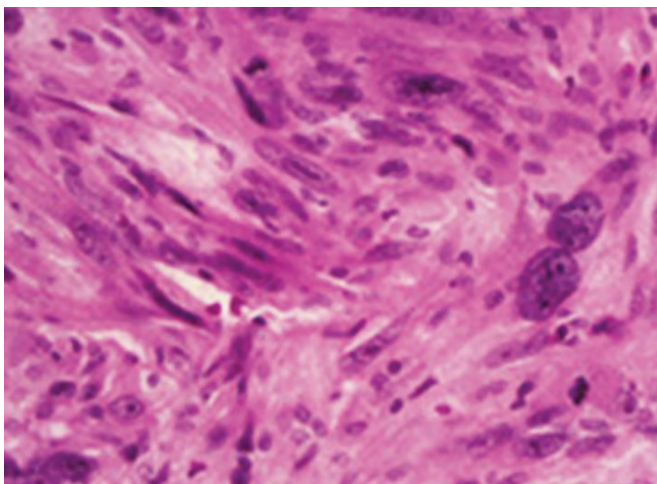


Fig. 4: Histopathological appearance of MFH showing storiform pattern



Fig. 5: Similar recurrent swelling in the left postauricular sub-mastoid region

cortices. Orthopantomogram and computed tomography (CT) scans were done, which showed extensive osteolytic areas in the left ramus and angle of the mandible measuring 3.54×2.34 cm and multifocal cortical destruction (Fig. 2). Aspiration cytology was done, which yielded a white jelly-like material. Based on the cytological findings, the lesion was diagnosed as osteosarcoma. There was no evidence of metastasis.

Since the lesion was diagnosed as osteosarcoma, segmental resection of the posterior part of the mandible was done under general anesthesia. Histopathologically, the lesion was diagnosed as MFH-pleomorphic storiform variant (Figs 3 and 4). The patient was sent for chemotherapy, which the patient discontinued in the middle of therapy.

Two months later, the patient presented again with a similar swelling in the left postauricular sub-mastoid region (Fig. 5). The lesion was surgically excised and the defect was covered with split thickness skin



Fig. 6: Recurrent ulceroproliferative lesion in the chin region

graft. Within a month, the patient again developed an ulceroproliferative lesion in the chin region (Fig. 6). The recurrent lesion was also surgically excised with resection of anterior part of the mandible, and the

resultant defect was reconstructed with Narayanan's bipedicle flap.

The patient again reported with recurrence 2 months later, in the left maxillary region involving the infraorbital rim. The patient was referred for postoperative chemotherapy, which was unsuccessful and the patient died within 1 month of the last recurrence.

DISCUSSION

Malignant fibrous histiocytoma is the most common soft-tissue sarcoma in adults, accounting for 20 to 30% of all soft-tissue sarcomas. It remains relatively uncommon in the head and neck region, accounting for 3 to 10% of all cases.¹ In addition to soft tissue, the tumor has been reported to occur in virtually every part of the body, including bone, viscera, and skin. In the head and neck area, the more common sites involved are the craniofacial bones (15–25%), larynx (10–15%), soft tissue of the neck (10–15%), major salivary glands (5–15%), and oral cavity (5–15%).⁸

Malignant fibrous histiocytoma of the mandible is very rare. To our knowledge, only 33 cases have been reported in the world literature^{3–6} prior to this case. The disease shows a distinct male predilection.⁸ The age distribution at diagnosis varied from 1.5 to 69 years but was more common in the latter half of life. The case presented here is a young male of 19 years.

The disease has a stronger tendency to localize in the posterior part of the mandibular body and the ascending ramus.^{9,10} The etiology of these tumors is unknown but cases have been described where MFH lesions have arisen in previously irradiated tissues and in areas of previous bone infarcts.¹⁰ A history of antecedent trauma in about 20% of the cases suggests that some of the tumors may represent an initial proliferative response to trauma.¹¹

The exact histogenesis of the tumor also remains controversial. It was initially proposed that the histiocytes that characterize the disease were acting as facultative fibroblasts, and this explained the apparent bimodal cell population found histologically. According to another theory, ultrastructural and tissue culture experiments showed small numbers of undifferentiated mesenchymal cells, suggesting that the histiocytic and fibrocystic cell lines are derived from these progenitor cells.¹² More recent studies, however, suggest that myofibroblasts may be the cell of origin.

Clinically, these lesions present themselves as swelling, pain, paresthesia, and mobility of the teeth, which were usually present from 2 weeks to 6 months before diagnosis and the mean tumor size at presentation was about 6.6 cm in diameter.

Radiologically, the reported findings were irregular bone margins, moth eaten appearance, erosion of cortex, pathological fracture, and tooth root resorption. Most of

the mandibular lesions presented as extensive ill-defined osteolytic lesion radiographically, which is not specific to MFH.⁵

Histologically, there are five variants of MFH: Storiform-pleomorphic (50–60%), myxoid (25%), giant cell (5–10%), inflammatory (5%), and angiomatoid.^{1,6} The storiform-pleomorphic type is the most common and is a highly cellular tumor, which can range from well differentiated to anaplastic. Histologically, low-grade (well-differentiated) MFH bone tumors are much less frequent than high-grade tumors (poorly differentiated) and have been associated with a better prognosis.^{13,14} Larynx, maxillary sinus, and mandible have the worst prognosis in the head and neck lesion.⁴

The biological behavior of MFH shows a high incidence of local recurrence and metastasis and carries poor prognosis. Malignant fibrous histiocytoma of bone aggressively infiltrates along facial planes or between muscle fibers, accounting for a high local recurrence rate.^{11,15} According to Huvos,¹³ metastasis occurs via hematogenous route to the lungs. No case of proven mandibular MFH has metastasized to lymph nodes; this is consistent with MFH of soft tissues, where only 12% had nodal secondaries.¹⁶ The prognosis of this tumor is often unfavorable and the recurrence rate is approximately 44 to 48%. Metastases more commonly occur in the lung (90%), followed by lymph nodes (12%), bone (8%), and liver (1%). About 5% of cases already reveal metastasis when the primary tumor is diagnosed.⁶

The course of the disease seems to correlate best with the size and depth of the tumor and the presence or absence of acute and chronic inflammation.¹ Small, superficially located tumors with a large number of surrounding acute and chronic inflammatory cells have a lower incidence of metastasis and they have a longer survival rate. Tumors with a myxoid component are also less likely to metastasize. The inflammatory variant seems to have the worst prognosis.

The management of mandibular MFH requires early radical surgery with removal of adjacent normal tissues (3 cm tumor-free margins),¹⁰ followed by radiotherapy and chemotherapy, when needed. The decision to submit a patient for radiation therapy depends on the size, site, grade, and the width of the surgical markings.¹² The administration of anticancer drugs, such as methotrexate, cisplatin, doxorubicin, cyclophosphamide, vincristine, has been reported in patients, but the efficacy of these drugs is yet to be established. It is important to select multiagent combination therapy, considering the interaction of these agents. Whatever be the mode of therapy, the overall 5-year survival rate is 48% with a

2-year survival rate of 60%.¹ Close follow-up after treatment is mandatory as local recurrence as in our case and early metastasis to lung are common.

CONCLUSION

A case of histologically proven MFH of mandible is presented. The case is presented for its rare occurrence, highly aggressive nature, high local recurrence rate, and tendency for early metastasis to lungs. Therefore, such mandibular lesions require early radical surgery with minimum of 3 cm tumor margins, followed by radiotherapy and chemotherapy, when indicated. A very close follow-up is also mandatory.

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