MALIGNANT FIBROUS HISTIOCYTOMA OF HAND - A CASE REPORT

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ABSTRACT

Malignant Fibrous Histiocytoma (MFH) is one of the most common soft-tissue sarcoma of late adult life. The common sites of involvement include the extremities especially the lower extremities, retroperitoneum and head and neck. Hand is an uncommon site of presentation. These tumors follow an aggressive course and recur unless widely excised. We present a case of an 18-year old female with MFH of second metacarpal of left hand.

***Key Words**: Malignant Fibrous Histiocytoma (MFH), Soft tissue tumour, Hand.*

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INTRODUCTION

Malignant fibrous histiocytoma (MFH) was first described by O'Brien and Stout in 1964[1]. Initially, MFH was defined as a neoplasm showing both fibroblastic and histiocytic differentiation; but according to latest WHO nomenclature, in which significant changes in nomenclature of lipomatous and fibrous malignancies were made; there is no evidence of true histiocytic differentiation and this morphological pattern is shared by a variety of poorly differentiated malignant neoplasms[2]. The different variants of MFH include storiform, pleomorphic, myxoid, inflammatory, giant cell and angiomatoid. MFH is the most common soft tissue sarcoma of late adult life[3]. The peak incidence is in the fifth and sixth decade of life but an age range of 10 to 90 years has been reported. The male to female ratio is 2:1. The common sites of involvement include the extremities (70-75% with lower extremities accounting for about 60%), retroperitoneum (10-15%), head and neck (5-10%) and sites within the abdomen and pelvis. There are various factors that predispose to developing MFH; which include previous radiotherapy, chromosomal abnormalities, RAS mutations, beta-catenin mutation and ultraviolet radiations. Hand is an unusual location and presentation in the hand is often mistaken for a ganglion or a hemangioma.

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CASE REPORT

An 18-year old female presented with a painless, slowly enlarging mass in left hand for five months (Figure 1). The swelling involved second metacarpal area with gradual loss of function of index finger and grip of hand. The swelling gradually increased in size. There was no history of trauma, previous surgery or radiotherapy. Her medical history was not contributory and she denied recent body weight loss. Clinical examination revealed that this mass was a fixed, firm, non tender swelling involving the dorsal and volar aspect of second metacarpal, involving the first web-space. Neurovascular bundle of the index finger were intact. Plain radiography showed soft tissue mass with compression of second metacarpal (Figure 2 and 3). Chest radiograph and ultrasound of abdomen demonstrated no evidence of metastatic disease. MR images revealed a mass with well defined margins in dorsal area of second metacarpal of right hand (Figure 4). A biopsy was taken which suggested it to be a low-grade malignant fibrous histiocytoma on histological appearance. Ray amputation of second metacarpal was done and mass was totally resected (Figure 5, 6 and 7).

Microscopically, the mass revealed neoplastic mesenchymal cells having markedly pleomorphic and hyperchromatic spindle nuclei and moderate to abundant amount of cytoplasm. Mitotic figures were frequently seen. The findings

were compatible with MFH. Post-operative adjuvant radiotherapy was given. The patient is free of symptoms and there is no evidence of recurrence after a period of eighteen months.

DISCUSSION

Malignant fibrous histiocytoma is one of the most common soft-tissue sarcomas occurring in late adult life. There are different variants of this

Figure 1: Appearance of hand before surgery

Figure 2: Anteroposterior view - X-ray left hand

Figure 3: Oblique view – X-ray left hand

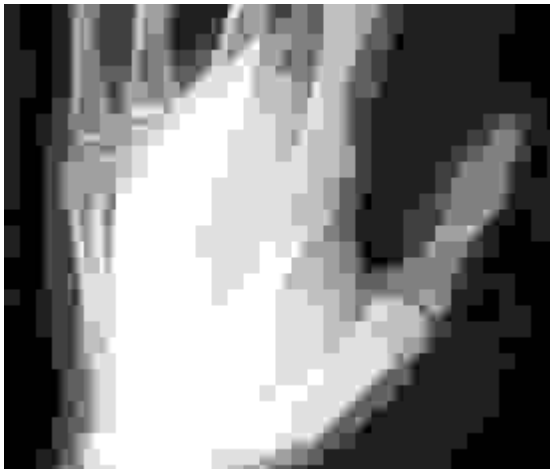

Figure 4: MRI image of index finger

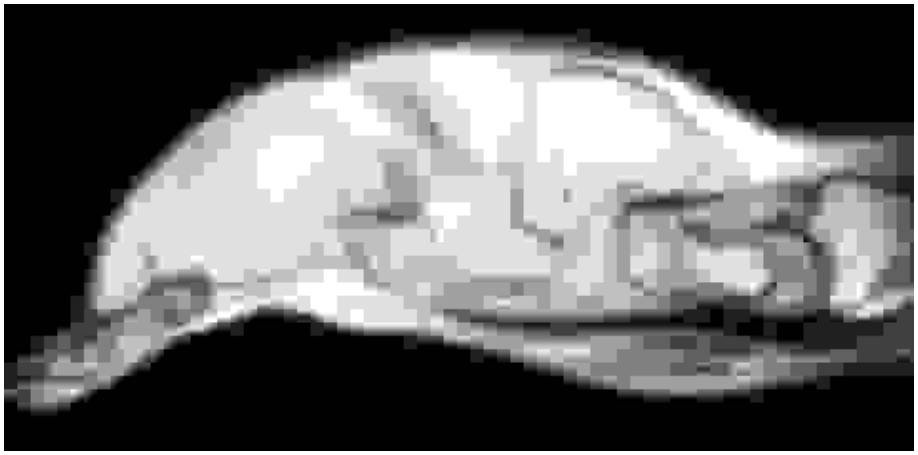

Figure 5: Ray amputation

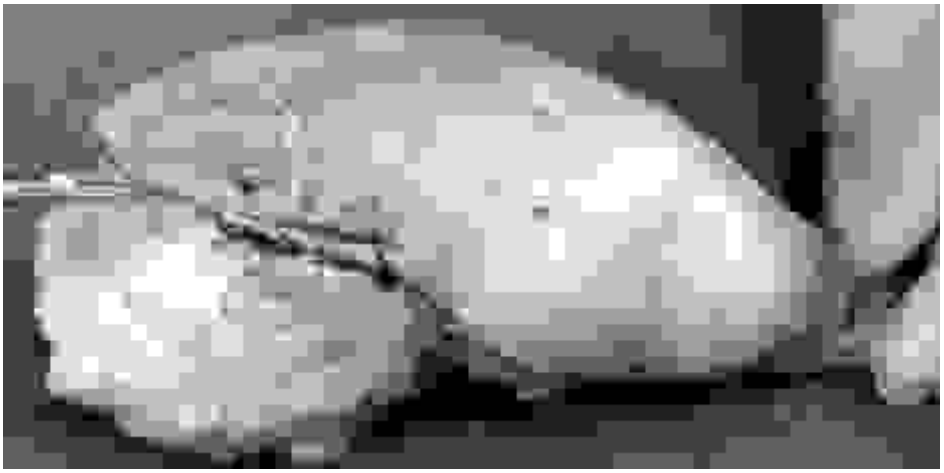

Figure 6: Appearance of hand after surgery

Figure 7: X-ray left hand – Post-operative

disease, which are storiform, myxoid, giant-cell, inflammatory and angiomatoid. This distinction is important as all have relatively different pathogenesis, treatment and prognosis; for example, myxoid variety is most amenable to chemotherapy while angiomatoid type is more common in children and adults. In our case, the patient had pleomorphic histological type and lesion on hand. Rooser et al in his study reported pleomorphic subtype, proximal location and increased histological grade to be factors which result in decreased survival rates[4]. But, Kearney et al in a series of 167 cases did not found the histological subtype as a significant factor affecting the survival rates[5].

Malignant Fibrous Histiocytoma mostly involves the extremities, retroperitoneum, head and neck, abdomen and pelvis. There have been reports of these sites in literature[6]. Involvement of hand, whether primary or secondary, is rare. There have been very few cases that have been reported[7]. Buecker et al in his study reported that patients presenting with primary malignancies of hand have a better chance of survival then similar tumors at other locations because of early detection and easy accessibility for therapeutic interventions[8]. In a study of 78 cases, Bertoni et al concluded that MFH tumors which are distal to elbow and knee have a better prognosis compared with distal tumors[9].

Commonly, MFH appears as a single mass which is painless and gradually grows overtime and is frequently accompanied with areas of hemorrhage and necrosis and in majority of cases, tumors are intramuscular. This is important as small tumors are analogous to early disease and large tumor size represent advance disease with increased chances of metastasis. Rooser et al and Bertoni et al in their studies have identified large tumor size to be independent factors associated with poor survival rates[4,9]. In contrast, Kearney et al did not found tumor size to be a significant risk factor but in his study majority of patients has small size tumors[5].

In our case, the exact diagnosis of MFH was made on the basis of histopatholical examination of the excisional biopsy specimen. Imaging was initially done using MR scans but they were only helpful in delineating the margins of the tumor. Literature shows that CT scans are best suited for those patients who cannot undergo MR scans and are also helpful as calcifications, seen in about 5 – 20% of cases is best demonstrated on CT scans. There have been a number of studies, which suggest that MR images provide detailed information of anatomical location and tumor involvement[10]. Only myxoid subtype gives a clear picture on MR images due to its specific signal intensity and enhancement pattern[11]. For distinction of MFH into its various sub-classifications, MR images are not helpful and only histopathological examination can reveal the histological subtype.

Treatment of malignant fibrous histiocytoma, like other tumors, is chemotherapy and preferably excision, if possible. MFH is a very aggressive tumor, so for better prognosis, it needs to be excised after diagnosis as soon as possible. There is data which supports use of adjuvant chemotherapy in improving the patients' condition and prevention of metastasis of the tumor[12,13]. In a retrospective study by Hsu et al on 76 patients with MFH, they concluded that postoperative radiotherapy was significantly associated with improved local control rates[14]. In another study by Belal et al on 92 patients, they concluded that adjuvant radiotherapy results in decreased local recurrence but has no effect on mortality[14]. In our case, we excised the metacarpal along with the metacarpo-phalangeal joint and phalanges in order to ensure resection of the tumor with clear margins and to allow wound closure with healthy tissue. The amputation was well tolerated, the wound healed well and the patient suffered little functional impairment.

CONCLUSION

Malignant fibrous histiocytoma (MFH) is one of the most common soft tissue sarcoma in adults but is rare in hand. The prognosis is largely dependent on the grading of tumor. Wide resection of the tumor mass with adjuvant therapy gives excellent results for low grade MFH tumors, achieving aim of limb-salvage procedure.

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