Malignant fibrous histiocytoma of chest wall

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Abstract
Malignant fibrous histiocytoma (MFH) is a pleomorphic sarcoma generally affecting extremities and retroperitoneum. Chest wall is rarely involved in this disease. A 52-year-old female was seen with MFH affecting left posterolateral chest wall and is being reported due to the rarity of its occurrence.

Key words
Malignant fibrous histiocytoma, pleomorphic, recurrence, radical excision, radiation therapy.

Introduction
Chest wall tumors are generally metastases from cancers of the breast, lung, kidney and thyroid, or else, multiple myeloma arising from ribs and malignant fibrous histiocytoma (MFH) is rarely encountered in this location. MFH is otherwise the most common soft tissue sarcoma in adults (20-30%) and generally occurs in the extremities, abdominal cavity, and retroperitoneum. This tumor involves the deep fascia, skeletal muscles and subcutaneous tissue. Proper imaging to define the extent of the tumor and complete surgical excision is the cornerstone of management. This report describes the clinical and radiologic features of a case of malignant fibrous histiocytoma of the posterolateral chest wall affecting a 52-year-old lady.

Case presentation
A 52-year-old postmenopausal lady was seen with a mass over the left side of posterior chest. The mass had been noticed 10 months back and had gradually increased in size. There was no history of pain or trauma nor was there any past medical or surgical history of significance. There were no symptoms suggestive of involvement of any other system. On examination, the patient was of average build and there was a firm lump about 18 cm x 15 cm over the posterolateral aspect of left hemothorax (Figure 1). It was not tender and the skin over it had areas of necrosis and superficial ulcerations with no active discharge or foul odor. The lump was mobile over the underlying muscles. There was no lymphadenopathy. There was no other visible or palpable lump. Routine laboratory blood works were within normal limits.

MRI studies revealed a soft tissue mass measuring 15 cm x 10 cm x 16 cm in size arising in the posterior back in subcutaneous tissue with low signal intensity on T1 weighted images and intermediate signal intensity on T2 weighted images and bright in signal intensity on the short tau inversion-recovery (STIR) weighted sequences and associated with central areas of necrosis (Figure 2). The mass lesion was associated with peripheral enhancement. There was no definite involvement of the latissimus dorsi muscle and no edema seen within it.

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Figure 1 Malignant fibrous histiocytoma of left posterolateral chest wall.

Figure 2 MRI of the patient shown in Figure 1 defining the chest wall tumor.

Furthermore, no neuromuscular bundle or lymph node involvement was seen. Following gadolinium administration, avid enhancement of the lesion peripherally with central areas of necrosis was seen. No distant metastases were detected. The diagnosis of malignant fibrous histiocytoma was made provisionally and the patient was attached to services of a regional cancer center where the diagnosis was confirmed and patient underwent multimodality management including excision of tumor, chest wall reconstruction and postoperative radiation therapy. The patient was seen after 15 months of management (for a different reason) and had no features of recurrence.

Discussion

Malignant fibrous histiocytoma (MFH) is a deep-seated pleomorphic sarcoma of uncertain origin, occurring in middle aged adults.\(^1^,^2\) MFH was initially described in 1964 by O’Brien and Stout\(^3\) who considered it to have a histiocytic origin. It has since been increasingly recognized to be a discrete entity. Irrespective of its precise histogenesis, the tumor contains both fibroblast-like and histiocyte-like cells in varying proportions, accounting in part for its broad morphologic spectrum and the diagnosis must be made by exclusion of other definitive sarcomas, especially myogenic and neurogenic sarcoma. However, in recent decades, evidences from literature\(^4^-^6\) tended to show that MFH represents a final common pathway in soft tissue tumors that undergo progression towards undifferentiation and in 2002; the World Health Organization (WHO) declassified MFH as a formal diagnostic entity and renamed it as an undifferentiated pleomorphic sarcoma not otherwise specified.\(^7\)

It occurs most frequently in the deep fascia and skeletal muscles of the extremities, abdominal cavity, and retroperitoneum.\(^2^,^8\) The lower extremity is the most common site of involvement. Chest wall is rarely involved in this disease.\(^2\) MFH can occur de novo though there are reports in literature that indicate the onset of MFH in the chest wall after old pleurisy or pyothorax, radiotherapy, burns, or after thoracotomies.\(^9^,^10\) The usual presentation is that of a slowly growing painless mass. The differential diagnoses include metastases from cancers of the breast, lung, kidney and thyroid followed by multiple myeloma arising from ribs.\(^2\)

The usual presentation is of a lump that has arisen over a short period of time ranging from
weeks to months. Often some mild trauma draws attention to the lump. The mass does not usually cause any pain unless it is compressing a nearby nerve. Symptoms such as weight loss and fatigue are not typical but can present in patients with advanced disease. Retroperitoneal tumors can acquire large dimensions before their detection as patients do not feel a mass per se but rather associated constitutional symptoms such as anorexia or increased abdominal pressure. Some unusual presentations of tumor are also reported in literature. Falidas et al. reported a case of malignant fibrous histiocytoma masquerading as gluteal abscess.

MFH is differentiated from fibrosarcoma by the presence of giant cells. There are many subtypes of MFH which include storiform ("spoke-like")/pleomorphic, myxoid, giant cell, inflammatory, and angiomatoid. Storiform/pleomorphic MFH is the most frequent histologic type and accounts for 50% to 60% of lesions and has spindle cells arranged in cartwheel pattern around blood vessels. The myxoid variant is the second most common type accounting for approximately 20% of cases and is least aggressive.

Imaging has an important role in management of this disease. Plain-films are usually nonspecific, showing only focal soft-tissue density. Calcifications may be seen in 5% to 20% of cases. Erosion or invasion into the bone can be seen and is highly suggestive of this diagnosis. Other soft-tissue sarcomas (with the exception of synovial sarcoma) do not have the tendency for cortical involvement. CT scans illustrate these tumors as large, lobulated soft-tissue masses of attenuation similar to muscle. Hypo-dense areas corresponding to myxoid regions, hemorrhage, or necrosis are frequently present centrally. Erosion of bone, periosteal reaction and pathologic fracture can be defined better with CT images. MR imaging of MFH show a lobulated (and often large) intramuscular mass having relatively well defined margins with intermediate intensity on T1-weighting and high signal intensity on T2-weighting. Heterogeneous signal is often seen on all pulse sequences reflecting the presence of various histologic elements including high signal for hemorrhage, and low signal for fibrous tissue and calcifications. Positron emission tomography/computed tomography (PET/CT) is also reported in recent literature as an effective imaging modality for MFH defining primary tumor and metastases are seen as hypermetabolic masses.

Once the diagnosis of MFH has been confirmed, a multimodality approach needs to be adopted and an individual treatment plan needs to be tailored for each patient. Surgery is the cornerstone of management and consists of radical excision with wide safety margins and dissection of the loco-regional lymph nodes. Post- or sometimes even preoperative radiotherapy may be a supplement of treatment. Chemotherapy is usually less effective and reserved for patients with recurrent disease or at the highest risk of disease recurrence. Local recurrences are reported in approximately 20-30% of all patients. Recurrences have been found lowest in extremities and highest in retroperitoneal and head and neck sarcomas and this variation is related to the inability to completely resect the tumor and achieve in anatomic locations outside of the extremity.

Prognostic factors that are known to correlate with survival in patients with MFH include tumor grade, depth, size, metastatic status, patient’s age, and histologic subtype. Oda et al. has shown that tumor location in the abdominal cavity, the retroperitoneum or the head and neck, tumor size of 5 cm or more, deep tumor location,
high histological grade (grade 3) based on the French Federation of Cancer Centers' grading system and high stage (stage III or IV) based on the American Joint Committee on Cancer (AJCC) staging system were significantly worse prognostic factors. Gibbs et al.\textsuperscript{16} has reported 10-year survival of 90%, 60%, and 20% for low, intermediate and high-grade tumors, respectively.

References