## CASE 18

**Patient:** A 32-year-old Thai woman from Samutprakarn

**Chief Complaint**: Solitary erythematous mass at right arm for 3 month

**Present Illness**: The patient noticed asymptomatic erythematous mass at right arm that gradually increased in size for 3 month. This mass was accidentally hit by a solid material causing it to ulcerated and bleed. She lacks other systemic symptom.

**Past History:** She was diagnosed with overlapping rheumatoid arthritis with systemic lupus erythematous since 2007. The disease is currently inactive.

**Dermatological Examination** (Figure 18.1): Solitary well-circumscribed reddish ulcerated nodule, firm consistency with contact bleeding at right arm

**Figure 18.1**

**Histopathology (S10-5291)** (Figure 18.2-3):

- Dense diffuse infiltrate of lymphocytes, histiocytes, fibroblasts admixed with atypical spindle and polygonal cells in the dermis and extended into subcutis
- Atypical cells showing large pleomorphic nuclei, many in mitosis and some are atypical mitotic figures

**Immunohistochemistry** (Figure 18.4-5)**:** Atypical cell positive with vimentin, focal positive with CD68 but negative with S100

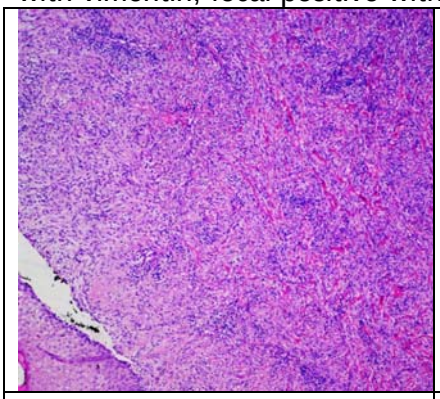

**Figure 18.2, H&E 40x**

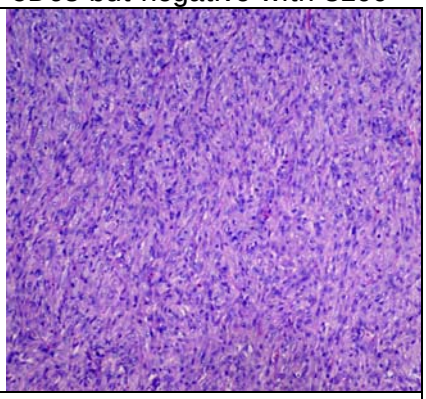

**Figure 18.3, H&E 200x**

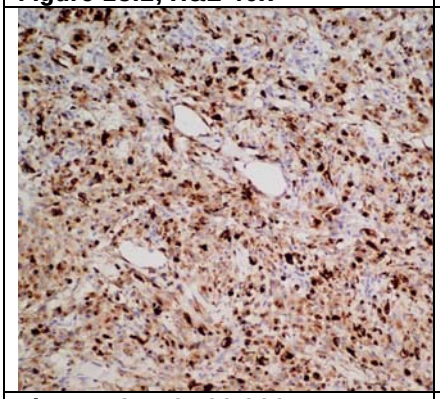

**Figure 18.4, CD68 200x**

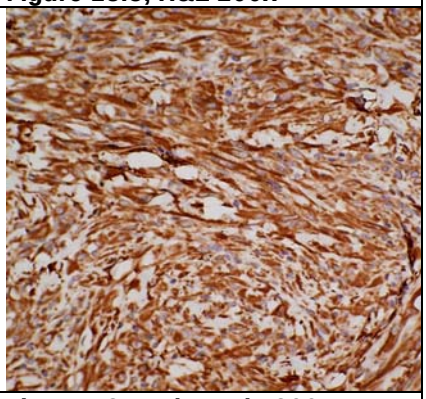

**Figure 18.5, Vimentin 200x**

**Investigation:** Normal Chest X-Ray

**Diagnosis:** Malignant fibrous histiocytoma

**Treatment:** Wide excision

**Presenter:** Chayada Kokpol

**Consultant:** Kumutnart Chanprapaph

**Discussion:**

Malignant fibrous histiocytoma (MFH) is defined as a high-grade pleomorphic sarcoma. It is a heterogeneous group of soft tissue sarcomas, composed of fibroblastic, histiocytic, and bizarre cells. Malignant fibrous xanthoma and fibroxanthosarcoma are other terms used for this tumor.[1]

The pathogenesis of soft tissue sarcoma has not been well defined. Depending on the cell of origin, various genetic, physical, chemical and viral etiologies have been suggested.[2]

Demographic data demonstrate that MFH most commonly occur in men (two third of the cases), age groups 50 to 70 years old. White persons are affected more often than those who are black or Asian.[1] The majority of MFHs (>90%) are deep tumors located beneath the fascia; however, the tumor occasionally occurs in the subcutaneous tissue.[1]

Cutaneous presentation of MFH is usually solitary skin-colored subcutaneous nodules, varying from elastic to firm in texture. It is progressively enlarging. The extremities are the most common site of involvement, with lower extremities affected more commonly than the upper extremities. Approximately 10-15% of MFH tumors are on the head and neck region.[2]

Clinical finding is not specific, therefore diagnosis relies on histopathology. Histologic finding of MFH composes of spindle cell in a storiform pattern. The stroma may be finely fibrillary, myxoid or densely collagenous. Bizarre epithelioid and giant cells may be present and may contain small amounts of lipid. Many mitotic figures, bizarre giant cells and necrosis are common.[2]

Immunohistochemistry using a broad antibody is required to rule out metastatic carcinoma, lymphoma, leiomyosarcoma and melanoma.

MFH is classified in four subtypes: storiform-pleomorphic, myxoid, giant cell and inflammatory (Table 1).[3]

However, the concept of MFH has generally fallen into disrepute. This tumor show no definable line of differentiation, and these are probably best classified for the most part as undifferentiated pleomorphic sarcomas (former pleomorphic and storiform MFH), myxofibrosarcoma (former myxoid MFH).[2,5]

MFH is an aggressive sarcoma; tumor location, size, and histologic grading directly influence prognosis.[6] Superficial tumors are generally smaller than deep tumors and have a better prognosis.[1]

MFH metastasize in 43% of cases. Metastases occur most commonly to the lungs (90%), lymph nodes (12%), bone, and liver (1%).[1] The 5-year survival rate varies with tumor size. Five-year survival for tumors less than 5 cm is 82%. This figure falls to 68% for 5- to l0-cm tumors and 51% for tumors greater than 10 cm.[1] Considering all MFH subtypes as a group, the 5-year metastasis-free survival rate is roughly 60%.[7]

Surgical excision is the primary mode of treatment for MFH. Because this tumor can spread a considerable distance beyond the gross tumor margins; aggressive wide and deep local excision has been recommended even for the superficial lesions. Recurrence usually occurs within the first 2 years after excision of the primary tumor. So in our case we decided to perform wide excision. Furthermore, we remained closely periodic follow up this patient. [1] Brown and Swanson retrospectively studied 17

patients with a total of 20 MFH tumors and reported that for the superficially located tumors, Mohs micrographic surgery seems to be an excellent treatment modality.[8]

Because of the high recurrence and metastatic rate of these soft tissue sarcomas, early consideration should be given to the use of adjuvant radiation therapy and/or chemotherapy for high risk MFH tumors. Soft tissue sarcomas generally respond to radiation therapy, which is usually given postoperatively. The value of adjuvant chemotherapy for soft tissue sarcomas remains controversial.[9]

## References


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**Table I** Summarizes the characteristics of the subtypes of MFH.[4]

| Subtype | Percentage(%) | Location | Histology | Prognosis |
|---|---|---|---|---|
| Storiform-pleomorphic | 66% | Deep-seated tumor, skeletal muscle of extremities, retroperitoneum | Variable appearance spindle, ovoid, giant cells in whorls, frequent and abnormal mitoses | Recurrence rate 44% Metastatic rate 42% |
| Myxoid | 25% | Skeletal muscle of extremities, retroperitoneum | >50% myxoid material arcs of blood vessel | Recurrence rate 66% Metastatic rate 23% |
| Giant cell | <10% | Multinodular tumor, skeletal muscle of extremities | Osteoclast-like giant cells, foci of osteoid or mature bone | Recurrence rate 50% Metastatic rate 42% |
| Inflammatory | <10% | Retroperitoneal tumor with constitutional symptoms | Xanthoma cells, inflammatory neutrophilic infiltrate | Recurrence rate 50% Metastatic rate 30% |