Malignant fibrous histiocytoma

Definition
Primary malignant neoplasm of bone and or soft tissue which contains fibrous and histiocytic elements.

Epidemiology
Rare. According to Stalley 1 per million or one per 2 million
Affects older age group 40-70. More common in males.

Site
Metaphysis of long bones, more so lower limbs than upper limbs.

Aetiology
20% arise secondary to other lesions such as fibrous dysplasia, bone infarction, Paget's disease and irradiation.

Pathology
Buzzword is "storiform pattern" – there is an appearance of wheel spokes radiating from a slit like vessel, in a fibrous background. A storiform pattern is not however diagnostic of MFH because it is also seen in osteosarcomas, leiomyosarcomas, neurogenic sarcomas, haemangiopericytomas and non ossifying fibromas.

Other typical histological features are bizarre histiocytic cells, numerous mitotic figures, and scattering of chronic inflammatory cells.

Histological variants:
1. MFH – myxoid subtype
    a. Sheets of myxomatous tissue with large histiocytes and abnormal mitoses
2. MFH – Inflammatory subtype
    a. Field of malignant histiocytes peppered with acute inflammatory cells, which may obscure the fibrous elements
3. MFH – Giant cell type
    a. Large number of multinucleated giant cells

Stout's hypothesis – theory of histiocytic and fibrous origin: histiocytes are derived from bone marrow monocytes - "tissue macrophages". Histiocytes have the potential to transform to spindle cells which produce collagen, and are indistinguishable from fibrocytes.

MFH is a diagnosis of exclusion; if the tumour resembles MFH but produces regions resembling another malignant tumour it is diagnosed as that tumour.

Clinical
Pain, swelling, pathological fracture
Relatively high metastatic potential, usually to lungs and other bones

Investigations
Laboratory
No characteristic alterations

Radiography
Lesions usually lytic or permeative; may expand cortex without breeching it.

Frequent periosteal reaction.

High index of suspicion if permeative radiolucency adjacent to bone infarct, fibrous dysplasia or Paget's disease.

MRI: T1 - low or intermediate intensity
T2 - high intensity

Immunohistochemistry
Vimentin stain positive
More specific is A1-AT antibody which also binds normal histiocytes

Treatment
Induction chemotherapy
Limb sparing surgery

Prognosis
Poor – around 60% 5ysr, but probably has a better prognosis than other sarcomatous lesions.

Prognosis worse for secondary lesions. 40% alive at 4 years