Eosinophilic granuloma

Definition
Condition at the benign end of the spectrum of histiocytosis X associated with unifocal or multifocal granulomas containing Langerhans cells and eosinophils.

Terminology
Histiocytosis X comprises a spectrum of disorders.

EG is most benign. Next is Hand-Schuller-Christian disease (exophthalmos, diabetes insipidus, and bone destruction). Most severe and potentially fatal is Letterer-Siwe syndrome, which is characterised by hepatosplenomegaly, lymphadenopathy, anaemia, acute infections and a downhill course.

Epidemiology
First two decades of life.
Slightly more common in males.

Pathology
The cell of origin is the Langerhans cell, a dendritic antigen presenting cell that is found all throughout the body, particularly in the skin and bones.

The Langerhans cell contains Birbeck granules, and has a nucleus with a deep cleft.

Other cells found in the granulomas include eosinophils, PMNLs, giant cells, and mononuclear cells. The Langerhans cells are arranged in clusters. Necrosis is very commonly seen.

Grossly the lesion is soft, tan to reddish and occasionally greenish.

Distribution
Skull, spine, pelvis and long bones, particularly the femur.
Most commonly unifocal but may be multifocal – up to 50%.
Has a preference for the skull and femur in children (sites of haematopoietic tissue) and the pelvis, ribs and skull in adults.

Clinical
Commonest symptom is pain. The pain is often worse at night.
May be accompanied by a palpable mass, fever and a limited range of motion.
Skin lesions are often seen.

Investigations
Laboratory
Rarely helpful. Occasional leukocytosis and elevated ESR. Usually no eosinophilia.

Imaging
XR typically shows lucent lesion sometimes with sclerosis. The lesion is typically in the medullary canal of the diaphysis of a long bone. It can occur in the epiphysis, however, and cross open growth plates.

It may cause cortical destruction.

It is the great mimicker and is in the differential diagnosis of any skeletal lesion. There may be expansion or periosteal reaction.

In the spine causes vertebra plana.

MRI – typically low T1 signal and high T2 signal. Surrounding oedema.

Differential diagnosis
Osteomyelitis
Ewing's sarcoma

Treatment
For EG, treatment is usually observation and the tendency is for resolution. Healing usually takes at least 5 months.

If there is ongoing pain or a lesion in a weight bearing bone then intralesional corticosteroids or curettage and bone grafting is used.

Radiotherapy can be used for inaccessible lesions