Eosinophilic Granuloma of the Sternum

Case 1159

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Section: Paediatric radiology
Technique: Ultrasound
Technique: CT
Case Type: Clinical Cases
Authors: C Brenner, R Hayes
Patient: 3 years, male

Clinical History:

A three year old male with a slightly tender sternal lump presented for investigation. Radiographs, US and CT showed a single lytic, expansile bony lesion, with reduced uptake on radionuclide scanning.

Imaging Findings:

A three year old male child was referred to the surgical team for evaluation of a rapidly growing, slightly tender mass in the manubrium sterni. The child was otherwise well with no significant past medical history. Clinical examination revealed a hard, non-mobile slightly tender mass at the manubrium. Plain radiographs showed an expansile lytic lesion involving the sternum (Figure 1). Ultrasound (US) showed the mass to be arising from the sternum (Figure 2). A technetium 99m radionuclide scan showed reduced uptake. A CT of thorax (Siemens Somatom Plus spiral scan, post IV contrast, slice thickness 5mm, pitch=1) showed a destructive 2.5-3cm lesion involving the sternum with a significant soft tissue component both anterior and posterior to the sternum (Figure 3). Abdominal ultrasound was normal. Possible differential diagnoses at this point included a primary or secondary bone tumour. Histiocytosis was also considered, with an infective process felt to be least likely. A biopsy revealed the presence of multiple eosinophils and immunohistochemical staining for S100 and CD1A were strongly positive, indicating the diagnosis of Langerhan’s Cell Histiocytosis. Curettage at biopsy was the only intervention at this point. A subsequent radiographic skeletal survey showed no further lesions. The child remained stable on follow up, with the mass still present but unchanged. Nine months later, the child complained of knee pain. A radiographic skeletal survey showed destructive lesions in the skull and left humerus. (Figure 4a,b). He was commenced on steroids. The lesions showed evidence of early healing on plain film, however the child has developed mild Cushing’s disease as a side-effect of treatment.

Discussion:

Eosinophilic granuloma represents part of the spectrum of Langerhan’s Cell Histiocytosis. This multisystem disorder can affect many sites including bone, brain, lung and the reticuloendothelial system. Eosinophilic granuloma tends to affect male children between three and twelve years of age. The bones usually affected are the long bones (diaphyseal), skull and pelvis. The sternum is an uncommon site of involvement. The lesions of eosinophilic granuloma are typically destructive without sclerotic margins. If multiple, the lesions can be found in various stages of evolution. They tend to resolve either spontaneously or following curettage. If the lesions are unresponsive clinically or radiologically, steroids or further chemotherapy may be used. In our institute, when there is progressive disease or organ dysfunction, the oncologists treat with methyl-prednisolone or vinblastine, with 6-mercaptopurine, methotrexate and etoposide being second-line agents. Radiotherapy is not used in our institute. Current published literature supports the fact that isolated bone lesions in the absence of organ dysfunction, can regress.
spontaneously or post biopsy and curettage. More extensive disease with organ involvement usually is managed with steroids and a variety of chemotherapeutic agent including vinblastine and etoposide. A recent randomised control trial showed these latter two agents to be equally effective in treatment of multisystem Langheran's cell histiocytosis although if there was no response within six weeks, patients were at increased risk of treatment failure and should have further chemotherapy with alternative agents. In general, patients with isolated bone disease have the best prognosis. Those with multisystem disease, in particular those under two years of age with multisystem disease and organ involvement have a more guarded prognosis. Disease recurrence in those who have shown disease regression or resolution is difficult to predict; it usually involves the skeleton or skin. Recurrence rates are reported to be in the order of 22-35%.

**Differential Diagnosis List:** Eosinophilic Granuloma of the Sternum

**Final Diagnosis:** Eosinophilic Granuloma of the Sternum

**References:**


Figure 1

Description: Plain radiograph of sternum showing a lytic expansile lesion with associated soft tissue mass. Origin:
Description: Ultrasound documenting lesion in sternum with large soft tissue component. Origin:
Description: CT showing destructive lesion with soft tissue mass, involving manubrium. Origin:
Description: Lateral skull radiograph demonstrating lytic lesions in skull vault Origin:
**Description:** Frontal radiograph of left humerus showing lytic lesions in both metaphysis and epiphysis. This latter site is rare but recognised. **Origin:**