EOSINOPHILIC GRANULOMA OF THE ILIUM

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ABSTRACT

A case of eosinophilic granuloma of the ilium presented as Pyrexia of Unknown Origin and pain in the hip on the same side of the lesion. Investigations led to diagnosis and treatment.

INTRODUCTION

Eosinophilic granuloma is one of a complex of syndromes which are grouped together under the heading Histiocytosis X. The syndrome complex includes solitary eosinophilic granuloma, multiple eosinophilic granuloma, Hand-Schuller-Christian disease and Lettere-Siwe disease, in order of increasing risk to life. The underlying process appears to be profileration of non-neoplastic histiocytes accompanied by varying proportions of eosinophilic and chronicinflammatory cells.

CASE REPORT

A 14 year old boy, of previous good health, presented with a two week history of pyrexia ranging from 38-39°C. The boy also complained of pain in the left hip, left sacro-iliac joint region and was noted to have a limp on the same side. There were no respiratory, urinary or bowel symptoms and no relevant past history.

On initial examination there was

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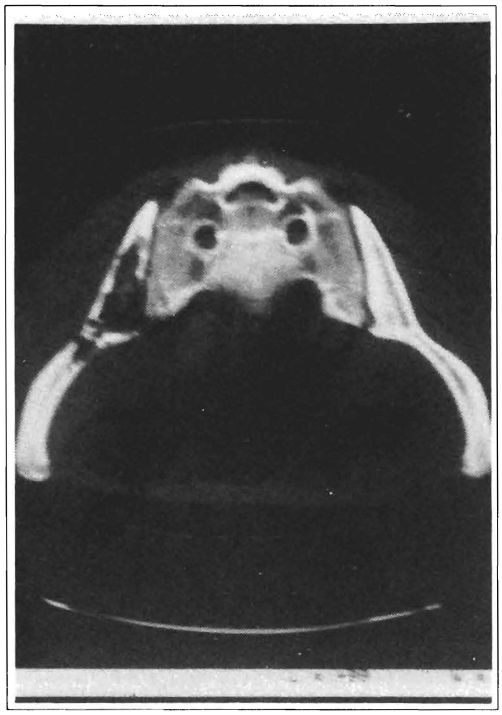

Fig. 1 - CT scan of Pelvis. Lytic lesion eroding the modullary carity of the left ilium.

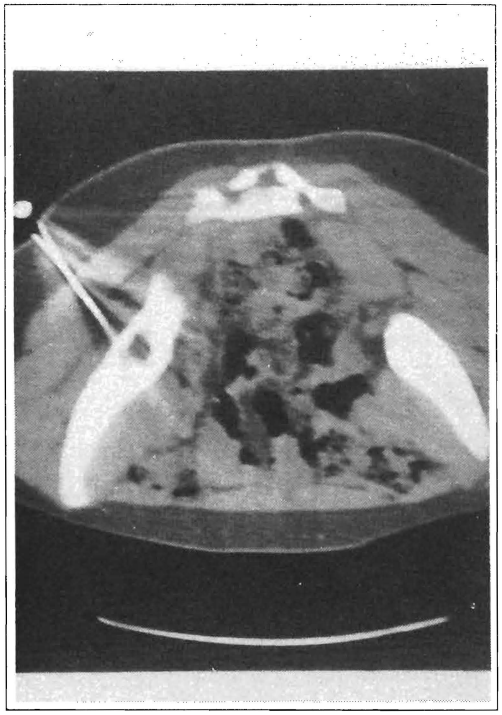

Fig. 2 - CT scan. Infiltration of lesion with steroid.

tenderness in the left sacroiliac region. There was full range of movement in the left hip, with positive Trendelenburg sign on the left. The boy also had severe acne vulgaris.

Full haematological investigations carried out soon after admission were all within normal limits and the E.S.R. was 6mm. Chest and pelvic X-rays were also reported as being within normal limits.

The following haematological investigations were all negative; BSR, Paul-Bunnell, HBsAG, LFT, ASOT and RA test. An MSU and throat swab were also negative.

There was no response to antibiotic therapy and the patient remained feverish. A bone marrow aspiration, fifteen days after admission was also negative. At the time the E.S.R. had gone up to 35 mm.

A C.T. scan was carried out in order to rule out the possibility of retro-peritoneal nodes. This showed a large lytic lesion eroding the medullary cavity of the left ilium (Fig.1). An orthopaedic opinion was sought and needle biopsy under C.T. scan imaging was performed, the samples being sent for histology and culture and sensitivity. Bacteriology showed Gram positive cocci in chains but no bacteria was cultivated. Histology was negative.

It was therefore decided to perform an open biopsy of the lesion. The latter consisted of necrotic bone with a paper-thin cortex and there was yellow gelatinous material surrounding the lesion. The biopsy was reported as showing an osteolytic process with sheets of eosinophils, reticu loendothelial cells, neutrophil lymphocytes and plasma cells and with many multinucleate giant cells. This was consistent with eosinophilic granuloma of the left ilium.

A skeletal survey showed no secondary lesions. The lesion was infiltrated, under C.T. scan imaging, with 80mg of Methyl Prednisolone (Fig. 2). Systemic Steriods were avoided because of the severe acne vulgaris. The pain subsided over the following two weeks, the pyrexia had already settled soon after open biopsy was done.

Follow-up two months after discharge showed the boy to be completely asymptomatic and X-rays of the lesion showed it to be completely healed (Fig.3).

DISCUSSION

Eosinophilic granuloma is a proliferation of nonneoplastic elements with eosinophils, plasma cells, lymphocytes and multinucleated giant cells (Langhans cells) with abundant pale-staining cytoplasm. The Langhans cells on electron microscopy show peculiar racket-shaped inclusion bodies in the cytoplasm (Birbeck granules) (1). Eosinophilic granuloma of bone is a self-limiting disorder, slightly more common in boys than in girls and usually seen in patients aged 5 - 15 years (2). Patients may complain of pain or local tenderness. The most commonly affected parts of the skeleton are the proximal femoral metaphysis, the skull, mandible, ribs and vertebral column where it can be associated with nerve root or cord compressions (1). When the lesion involves a long bone, it is usually diaphyseal and associated with a marked periosteal reaction, while when the lesion is in a flat bone, it is radiolucent without reaction, a so-called punched-out lesion (2).

An eosinophilic granuloma may affect soft tissue, including the oral mucosa, skin, lymph nodes and lungs. When the latter are involved, patients may

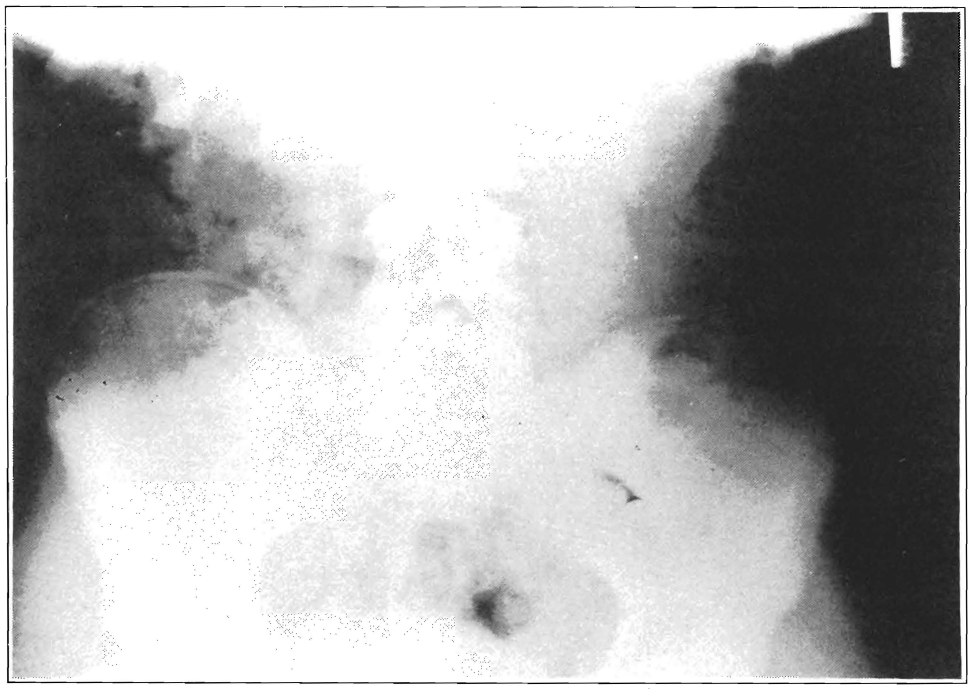

Fig. 3 - Pelvic X-ray. Complete resolution of lesion.

soft tissue, including the oral mucosa, skin, lymph nodes and lungs. When the latter are involved, patients may develop progressive fibrosis with impaired pulmonary functions (1).

In a child, eosinophilic granuloma may be confused with Ewing's sarcoma, osteolytic osteosarcom or osteomyelitis. In the adult it may be confused with lymphoma of bone,malignant fibrous histiocytoma, myeloma or metastatic disease. Destruction of bone may be so severe that it may lead to pathological fracture (2).

Patients with unifocal lesions may show spontaneous regression, thus treatment beyond biopsy is probably unnecessary in the solitary form, but curettage with or without bone grafting, low-dose chemotherapy, and recently corticosteroid injections all have been used successfully (3).

In general, if a second lesion does not appear within one year, the prognosis is good. In those patients who present with a more systemic illness characterised by fever and organomegaly, as well as multiple osseous lesions, the course of the disease is likely to be protracted (1). The disseminated form has been treated with radiotherapy, steroids, chemotherapeutic agents (such as vinblastine) and recently thymic extract, all varying in success (3).

The clinical course and the histopathology of this disease indicate that it is nonneoplastic in nature. The presence of eosinophils and the occasional occurence of skin lesions suggest the possibility that this is a peculiar immunoallergic phenomenon (1).

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