LESSON OF THE MONTH

Erdheim-Chester disease: typical radiological bone features for a rare xanthogranulomatosis

V Breuil, O Brocq, C Pellegrino, A Grimaud, L Euller-Ziegler

CLINICAL HISTORY
A 55 year old man was admitted to the rheumatology department owing to inflammatory back pain (occurring at 4 00 am, not aggravated by movement) without sciatica, increasingly severe over two years and associated with asthenia and weight loss (10 kg). No abnormalities were found on physical examination, except for tenderness on palpation of the spinous processes of L5 and S1. There was no significant past medical history. Laboratory tests showed a mild inflammatory picture: erythrocyte sedimentation rate 25 mm/1st h (normal 5–15), C reactive protein 12 mg/l (normal 0–10), fibrinogen 5.1 g/l (normal 2–4). Protein electrophoresis, red and white blood cell count, glucose, renal, and liver function tests were normal. Chemical bone markers (urinary pyridinoline, alkaline phosphatase, osteocalcin, parathormone, 25-hydroxyvitamin D, serum calcium and phosphorus, and urinary calcium and phosphorus) were normal.

RADIOLOGICAL FINDINGS
Standard lumbar spine and pelvic radiographs were normal. Technetium-99m bone scintigraphy showed no abnormal uptake in the spine or skull but disclosed an increased peripheral uptake.

A skeletal survey showed multiple mixed bone lesions with sclerotic areas surrounding smaller lytic foci. These roughly bilateral, symmetrical lesions involved the metaphyses and diaphyses of some long bones but spared the epiphyses. Bones affected were the femora, tibiae, radii, wrist, and tarsal bones.

With magnetic resonance imaging (MRI), these lesions appeared in low signal on T1 weighted sequences (fig 2) and in high heterogeneous signal on T2 weighted sequences with fat annulation. The computed tomography (CT) scan showed central osteosclerosis of the medullary canal with no evidence of osteolysis or periostosis.

An abdominal CT scan disclosed tissue infiltration surrounding the kidneys.

Biopsy of the tibial area displayed foamy lipid laden histiocytes, confirming the diagnosis of Erdheim-Chester disease.

TREATMENT
Because of the systemic involvement, prednisone (20 mg daily) treatment was started and, after one month, significant clinical and laboratory improvement was seen. At nine months the patient’s general condition had returned to normal, his back pain had completely disappeared, and all laboratory values had returned to normal. Moreover, the...
treatment have been used. 

In this case, we obtained a good clinical and laboratory response with a partial resolution of the retroperitoneal infiltration; the patient’s disease was stabilised with a small dose of steroids, but he remains dependent on it. The prognosis depends to a large part on the extent and distribution of extraosseous disease. In a review of 59 patients, Veyssier-Belot et al reported death related to the disease in 59% of cases, including 36% in less than six months: the most commonly reported causes of death are respiratory and heart failure. The efficacy of the various treatments is difficult to evaluate because of the rarity of the disease.

THE LESSONS

- Erdheim-Chester is a rare non-Langerhans histiocytosis showing characteristic radiological and histological features.
- Extraskeletal manifestations can occur in almost all organs, leading to a poor prognosis.
- As bone pain is the first revealing symptom, rheumatologists should be able to recognise the disease.

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Accepted 23 July 2001

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Ann Rheum Dis 2002 61: 199-200
doi: 10.1136/ard.61.3.199

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