Figure 1 Detail from chest radiograph showing upper humerus

Fewer than a hundred cases of Erdheim–Chester disease have been reported.³ Average age at presentation is around 53 years but the youngest patient was 7. The condition can affect not only the bones and retroperitoneum, as in this case, but also the hypothalamic/pituitary axis (leading to diabetes insipidus), the pericardium, the lungs and the skin (xanthomas).^{3–5} The most common presenting complaint is bone pain along with general symptoms such as fever,

weight loss and weakness.⁴ Hydronephrosis is due to infiltration of retroperitoneal fat.⁷

The prognosis depends largely on the extent and distribution of extraosseous disease. Systemic corticosteroids, chemotherapy and radiation treatment have all been used but formal trials are lacking.

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Fibrinous peritonitis in a patient with rheumatoid arthritis

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Ascites in rheumatoid arthritis has been linked to drug-induced liver damage but not previously to peritoneal disease.

CASE HISTORY

A man of 76, diagnosed 15 years ago with seropositive rheumatoid arthritis, had for 5 years been taking methotrexate as a disease-modifying agent (currently 7.5 mg per week). When he sought advice because of

Figure 2 Scintigrams

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Figure 1 Peritoneal biopsy specimen

shortness of breath and abdominal distension the methotrexate was immediately stopped. On examination he had gross ascites. Constrictive pericarditis secondary to rheumatoid arthritis was excluded by cardiac MRI. Diagnostic peritoneal tap revealed an exudate and so the possibility of peritoneal disease was further investigated by laparoscopy, at which peritoneal and liver biopsies were obtained. The liver biopsy was normal but the peritoneal biopsy showed a fibrinous peritonitis with a mild chronic inflammatory infiltrate (Figure 1) similar to that seen in fibrinous pericarditis associated with rheumatoid arthritis.

Repeated drainage of the ascites was necessary after the patient's discharge from hospital, but after institution of prednisolone 15 mg daily there was no recurrence. The methotrexate was not reintroduced because the rheumatoid arthritis was well controlled.

COMMENT

An extensive search of the published work has yielded two reported cases of ascites related to rheumatoid arthritis.^{1,2} In these instances the aetiology was judged to be methotrexate-induced liver damage and the ascites resolved on withdrawal of the drug. The only other documented cause of fibrinous peritonitis is practolol, an extinct beta-blocker that our patient had never received.³⁻⁶ The fibrinous peritonitis in this patient was histologically very reminiscent of the constrictive pericarditis seen in rheumatological arthritis.

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Two cases of hypothalamic-pituitary sarcoidosis

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Sarcoidosis is a multisystem granulomatous disorder that can affect the hypothalamus and pituitary glands.¹ Three decades ago the mean survival in hypothalamic-pituitary sarcoidosis, in a review of 19 cases, was 45 months.² We report on two patients still alive after diagnosis 20 years ago.

CASE HISTORIES

Case 1

A man of 34 sought advice in 1981 after 10 months of nausea, weight loss, depression and reduced libido. Severe depression was diagnosed and he was admitted to a psychiatric hospital. On examination, he was noted to have a nasal erythematous rash, sausage shaped fingers, reduced secondary sexual hair and small testes. Plasma sodium was 112 mmol/L. Anterior pituitary hormone stimulation tests (Table 1) and a water deprivation test demonstrated panhypopituitarism. A biopsy specimen of the nasal rash was reported as showing granulomatous inflammation consistent with lupus pernio, and in radiographs of the hands there were translucent cystic lesions likewise consistent with sarcoidosis. On CT the pituitary gland appeared normal. A Kveim test was positive. Cerebrospinal fluid had a raised protein (0.48 g/L) and white cell count (12/ μ L, all lymphocytes) but angiotensin converting enzyme (ACE) activity was normal. A whole-body gallium scan was normal.

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