Netherlands
The Journal of Medicine

PHOTO QUIZ

An X-ray that helps to solve the puzzle

A.P. Bech*, L.J. Reichert

Department of Internal Medicine, Rijnstate Hospital, Arnhem, the Netherlands,
*corresponding author: e-mail: annekebech@hotmail.com

CASE REPORT

A 70-year-old man, who used to be a metalworker, was admitted for analysis of pleural effusion. His medical history revealed longstanding hypertension, atrial fibrillation, transurethral resection of the bladder, heart failure and renal failure for which he was treated with haemodialysis. He did not mention any complaints besides some migrating pain of the knee and the wrist after activity. He did not have morning stiffness and could sleep well. On physical examination a raised right lung border was found. No lymph nodes were palpable. The left knee showed minimal hydrops without other signs of arthritis. Laboratory investigations showed a raised erythrocyte sedimentation rate (83 mm/h), leucocytes of 9.4/mm^2, a monocytopenia (0.09/mm^2), a lactate dehydrogenase of 352 U/l and a C-reactive protein of 1.45 mg/dl. Serology was negative for anti-CCP, complement factors, antinuclear antibodies, antineutrophil cytoplasmic antibodies and rheumatic factor. A puncture of the pleural fluid showed an exsudate. The culture of the pleural fluid showed no growth of bacteria and interferon gamma release assay testing was negative. A PET-CT scan showed the radiological picture of pleural thickening with pleuritis and perirenal inflammation. A pleural biopsy was performed which showed focal accumulation of histiocytes with positive staining for CD68 and negative staining for S100, CD1a and markers for a mesothelial origin. A bone marrow puncture showed no abnormalities. Bone radiographs of the legs and arms were taken, which confirmed the suspected diagnosis (*figures 1* and *2*).

WHAT IS YOUR DIAGNOSIS?

See page 134 for the answer to this photo quiz.

Figure 1.

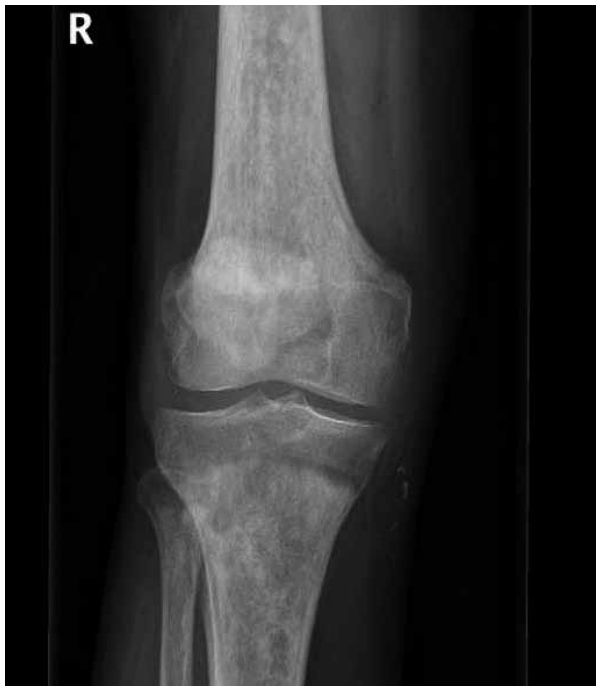
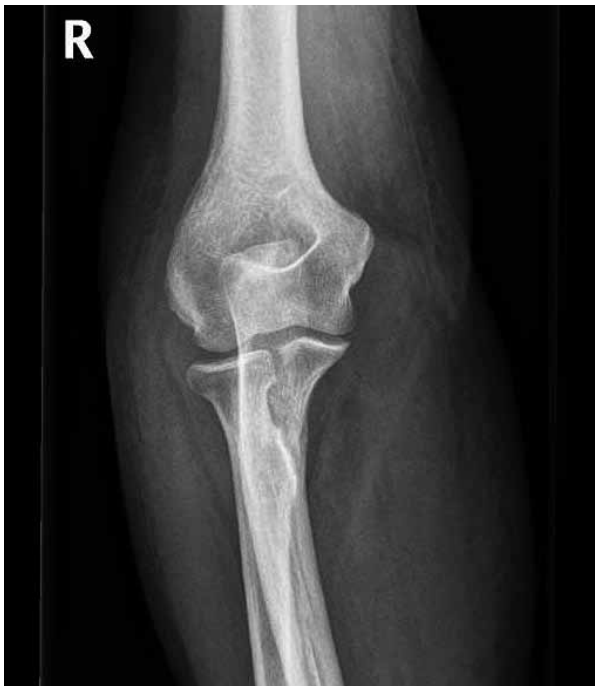


Figure 2.



ANSWER TO PHOTO QUIZ (PAGE 130)

AN X-RAY THAT HELPS TO SOLVE THE PUZZLE

DIAGNOSIS

The radiographs showed sclerosis of the diaphyseal and metaphyseal regions. The combination of the radiological signs, histological abnormalities, the pleural thickening, pleural effusion and perirenal inflammation confirmed the diagnosis of Erdheim-Chester disease. Erdheim-Chester disease is a non-Langerhans cell histiocytosis of unknown aetiology with a broad clinical spectrum. The diagnosis is based on the typical radiographic signs of symmetrical sclerotic or mixed sclerotic and lytic lesions of the metaphyseal and diaphysial regions combined with specific histological features of histiocytic infiltration with positive staining for CD68 and negative staining for S-100 and CD1a.[1] Bone pain is a typical complaint but approximately half of the patients exhibit extraskeletal manifestations as well.[2] These manifestations can be present in multiple organs such as the lung and kidney, as illustrated in this report. In total, 20 to 35% of the patients exhibit pulmonary disease, which can consist of interlobular septal thickening, centrilobular nodular opacities, fissural thickening, pleural thickening and pleural effusions.[3] Renal involvement of Erdheim-Chester disease is described as well. Perirenal involvement in Erdheim-Chester disease is characteristically manifested as a rindlike soft-tissue (inflammation) surrounding the kidneys and ureters.[4]

The prognosis of the disease depends on the extent and distribution of the extraskeletal manifestations. In general, one third of the patients with pulmonary involvement die within six months. Treatment is still controversial. Corticosteroids, interferon alpha, cyclophosphamide and surgery all have been described, but there is no consensus concerning the optimal treatment modality and the influence on the prognosis.

REFERENCES

1. Valentini D, Cappelli C, Mizzoni F, et al. Erdheim-Chester disease: a non-Langerhans cell histiocytosis. A clinical-case and review of the literature. Clin Ter. 2004;155(5):205-8.
2. Allen TC. Pulmonary Langerhans cell gistiocytosis and other pulmonary histiocytic diseases. Arch Pathol Lab Med. 2008;132:1171-81.
3. Wang CW, Colby TV. Histiocytic lesions and proliferations in the lung. Semin Diagn Pathol. 2007;24(3):162-82.
4. Surabhi VR, Menias C, Prasad SR, Patel AH, Nagar A, Dalrymple NC. Neoplastic and non-neoplastic proliferative disorders of the perirenal space: cross-sectional imaging findings. Radiograph. 2008;18:1005-17.