Case report 402

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Radiological studies

Fig. 1. AP and lateral roentgenograms of the right elbow show a number of well-delineated, osteolytic lesions affecting the distal end of the humerus and the proximal portions of the radius and ulna. The lytic areas tend to be confluent.

Fig. 2. An AP view of both wrists, including most of the metacarpals, shows similar, multiple, well-delineated osteolytic foci, affecting nearly all the bones included in the study (radius, ulna, metacarpals). The cortical surfaces show thinning.

Clinical information

This 41-year-old man was referred to the Department of Internal Medicine for evaluation of anemia. His complaints consisted of weakness, pain in the limbs, nocturnal sweats and difficulty in sleeping, persisting for 5 weeks prior to admission in 1983.

On physical examination, pallor and splenomegaly were evident. Laboratory tests included BSR 52/82 (Westergreen), LDH 694 U/l, gamma GT 38 U/l, hemoglobin 10.3 g%, platelets 84,000, leukocytes 11,900. The differential white blood count showed 20 stab cells, 15 segmented granulo-
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Fig. 3. A CT scan of both legs shows diffuse sclerosis of these limb bones, particularly affecting the right tibia. The findings are bilateral but not symmetrical.

Fig. 4. A lateral roentgenogram of the skull shows numerous osteolytic foci which are markedly confluent and appear to be distributed over the entire vertex.

tures of the medullary cavity and spongiosa was beginning to disappear. Some segments of the skeleton showed severe thinning of the cortices but no disruption. These changes were evident near the elbow (Fig. 1), the distal portions of the radius and ulna (Fig. 2) and in the bones of the hands and feet where the appearance resembled a "honeycomb" pattern (Fig. 2). Several bones showed a translucent, "glassy" appearance, particularly at the distal end of the humerus and additional segments of the radius and ulna. Sclerosing changes were beginning to appear with characteristic obliteration of the bony trabeculae. Such findings of increasing density of the medulla with sclerosis of the diaphyseal segments were documented in both femora. A CT study of the bones of the legs showed not only the destruction of the cortex and medullary cavity but also a striking central sclerosis (see Fig. 3). A roentgenogram of the skull showed many osteolytic lesions which were confluent and involved the entire vertex, particularly marked in the parietal area (Fig. 4).

Radiological studies of the trunk also showed marked increase in density of the spine, but no evidence of bony destruction. On the other hand, the innominate bones and ribs showed numerous osteolytic foci, some poorly delineated, in addition to the predominate sclerosing changes.

Skeletal scintigraphy with radionuclides demonstrated an abnormal increase in metabolic activity near multiple joints.

A month later the value of the hemoglobin fell to 8.3 g%. Therapy with Oximetholon evoked no response and the hemoglobin decreased to 7.4 g%. The alkaline phosphatase index reached 190.3. The patient was transfused with 3 units of washed erythrocytes and a bone marrow transplant was considered; however, problems existed in finding a suitable donor. As the transaminase values increased to 300 U, and the gamma GT rose to 100 U/l, therapy with Oximetholon was discontinued. Two months later a biopsy was obtained from an osteolytic lesion in the left humerus (Fig. 5A and B).

Over the following 8 months the patient received several transfusions of erythrocytes because of severe anemia. In the beginning of September 1984 the spleen increased in size, extending 9 cm below the costal arc and the LDH reached 1,600 U from a previous determination of 694. The leucocyte count was 3,500, the platelets 40,000 and the reticulocyte index was 1%. A hemolysis test was negative. The radiological findings remained unchanged.

Another biopsy from the bone marrow was obtained in January 1985.

Septic fever and uremia occurred after a few days and the patient died with signs of disseminated sepsis. At autopsy the cause of death was reported as terminal bronchopneumonia.