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Myelofibrosis With Massive Hepatosplenomegaly and Osteolytic Bone Lesions

Ginger M. Merry, MD, MPH
Paul B. Aronowitz, MD

Department of Medicine, California Pacific Medical Center, San Francisco, California.

An 83-year-old man with a 7-year history of myelofibrosis presented to the hospital with progressive weakness and fatigue, which resulted in him tripping and falling onto his left hand and arm 1 day prior to admission. His past medical history was significant for transfusion-dependent anemia and hypertension. His current treatment regimen for myelofibrosis included thalidomide and darbopoetin alfa.

Physical examination revealed a pale and edematous man who was holding his injured arm to his chest, but in no distress. He had massive hepatosplenomegaly (Figure 1) and pitting edema of the lower extremities that extended to his abdomen.

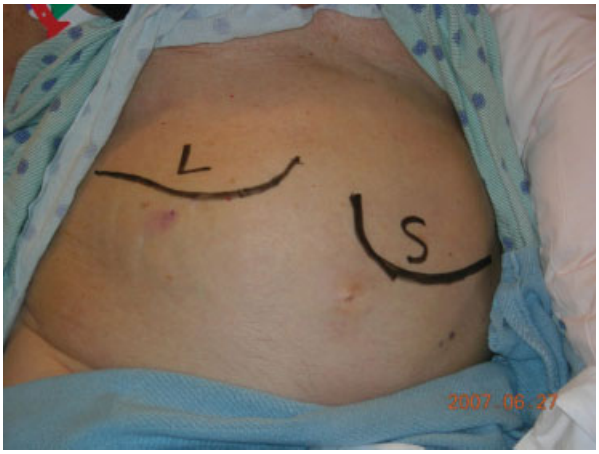

FIGURE 1. Massive hepatosplenomegaly.

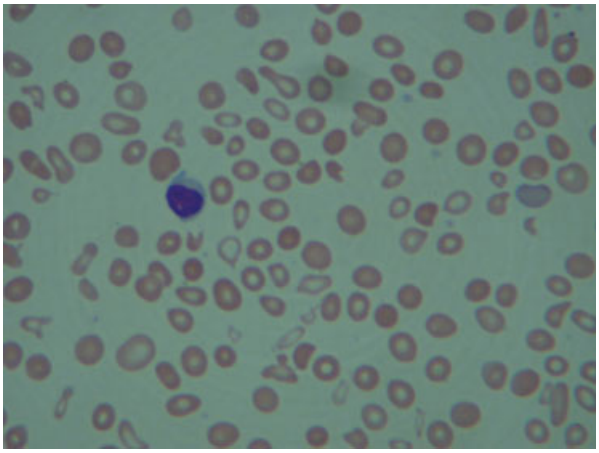

FIGURE 2. Blood smear.

Laboratory studies showed a white blood count of 5000, hematocrit of 29%, and platelets of 218,000. The peripheral blood smear (Figure 2) showed marked anisocytosis, poikilocytosis, and teardrop cells (Figure 2; arrow).

Imaging of the left arm and hand was significant for a third metacarpal fracture and first phalanx fracture. Of note, these x-rays also revealed numerous round lucencies within the osseous structures of the left hand, wrist, and forearm (Figure 3; arrow).

The patient's hospital course was uncomplicated and included casting of the left arm, treatment of his lower extremity edema, and transfusion for a slowly declining hematocrit. He was discharged home after several days but died 1 month later.

Primary myelofibrosis is a myeloproliferative disease that consists of 2 phases. The first phase is the growth and proliferation of abnormal bone marrow stem cells, which leads to ineffective erythropoiesis. This is followed by reactive myelofibrosis and extramedullary hematopoiesis.[1] These 2 phases of the disease can lead to a constellation of findings, as illustrated in these images. The median length of survival from diagnosis is 3 to 5 years, with the main causes of death

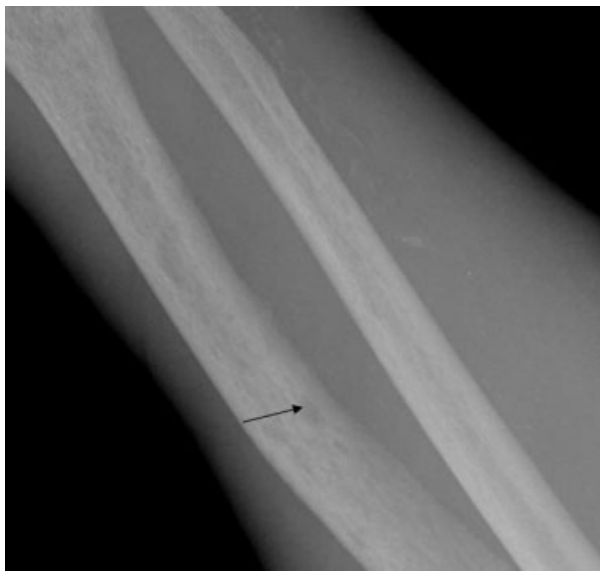

FIGURE 3. X-ray of left arm.

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being infection, hemorrhage, cardiac failure, and leukemic transformation.[1] Presenting signs, symptoms, and laboratory results may include cachexia, splenomegaly, anemia, an increased or decreased white blood cell count and/or platelet count, and an increase in lactate dehydrogenase. Radiographically, the most common findings are marked splenomegaly and osteosclerosis.[2]

Osteosclerotic lesions are found in 30% to 70% of patients with myelofibrosis and are a result of marrow fibrosis, which leads to the appearance of diffuse, patchy increases in bone density.[2] Osteolytic lesions, as seen in this case, are much less common. They appear in the literature in case reports, but are not considered to be a typical finding. They are usually painful and have been reported as a poor prognostic indicator.[3,4]

Address for correspondence and reprint requests:
Paul B. Aronowitz, MD, 2351 Clay St., Suite 360, San Francisco, CA 94115; Telephone: 415-600-1133; Fax: 415-775-7437;
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