MARTIN QUAN, MD
Department Editor

A young man with sickle cell crises

James D. Collins, MD

This 24-year-old black Hispanic man has had episodes of pain in his bones and abdomen since he was 2-years-old and was diagnosed with sickle cell anemia at age 10. He has eleven siblings with no evidence of sickle cell disease (SCD), and both parents are alive and well.

When he was 21, he experienced a severe sickle cell crisis characterized by bone pain that required multiple transfusions. In the following 3 months, he had three significant crises requiring short hospitalizations but no blood transfusions. Since then he has had several admissions to our medical center, one of which was complicated by possible pulmonary emboli.

Two years ago, he had a severe crisis with abdominal, back, and hip pain with a filling defect shown on a lung scan. He was treated with standard treatment of analgesic agents, nasal oxygen, and hydration. He also required 5 units of blood to increase his hematocrit level from 25% to 35% and his oxygen concentration by pulse oximetry from 62% to 75%.

During his most recent hospitalization, he came in with complaints of chest, shoulder, and abdominal pain, which did not subside by 24 hours with standard treatment. A transfusion of 3 units of packed red cells did not relieve the pain, but a limited exchange transfusion finally produced relief.

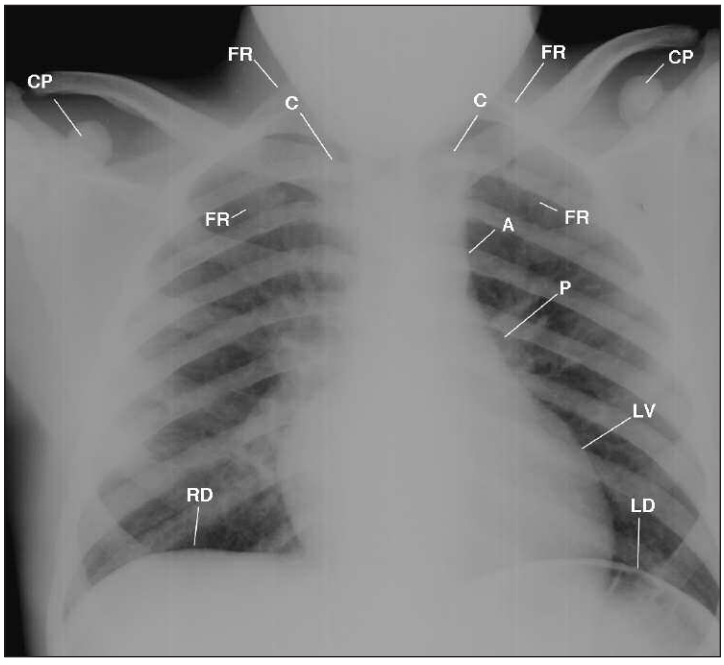

Figure 1 This posteroanterior (PA) chest radiograph displays hunched-up, rounded shoulders (signifying weakness due to anemia); forward shift of the cervicothoracic spine; sloping of the dense posterior ribs; absence of the spleen, and an enlarged heart. A = aorta; C = clavicle; CP = coracoid process; FR = first rib; LD = left hemidiaphragm; LV = left ventricle; P = pulmonary artery; RD = right hemidiaphragm.

Discussion

Sickle cell disease (SCD) is an autosomal recessive genetic disease caused by a single amino acid displacement (valine substituted for glutamic acid) on the β-globin gene. If each parent carries one defective gene (*Hb S*), a child has a 25% chance of inheriting two defective genes, a 25% chance of inheriting two normal genes, and a 50% chance of being an unaffected carrier of the trait like the parents. About 8%-10% of African-Americans carry the trait, and about 1.5%-2% have SCD. Prevalence is also high in people whose ancestors come from Spanish-speaking regions such as Central or South America and Cuba,

Dr. Collins is professor, department of radiology, David Geffen School of Medicine at UCLA. Dr. Quan is professor of family medicine, department of family medicine, David Geffen School of Medicine at UCLA. He is also editor in chief of *Family Practice Recertification*.

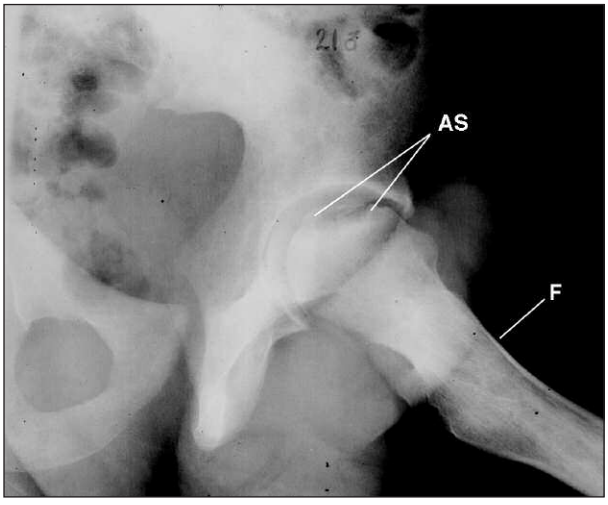

Figure 2 This left posterior oblique view of the left hip displays irregular demineralization of the superior portion of the left femoral head suggesting aseptic necrosis (AS). Observe the indistinct cortical margins of the dense pelvic bones and the thin cortical margins of the left Femur (F).

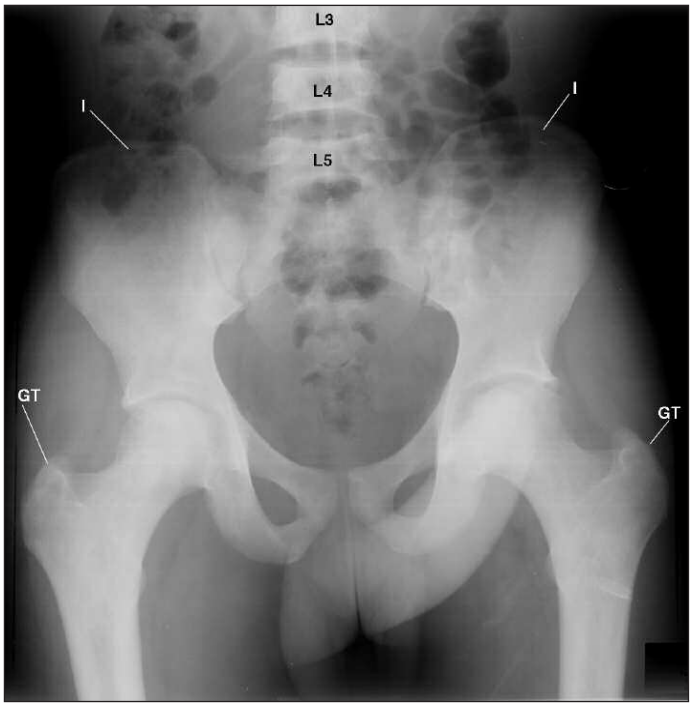

Figure 3 This anteroposterior (AP) view of the pelvis displays the dense bones of the lumbar vertebrae (L3-L5), iliac bones (I), and the greater trochanter (GT). Note the biconcave (fish-mouth) appearance of the vertebrae, which is characteristic of sickle cell disease.

Saudi Arabia, India, and Mediterranean countries such as Turkey, Greece, and Italy.[1-4]

The genetic abnormality produces dense, rigid, sickle-shaped erythrocytes that are sticky, adhere to endothelium, and clog capillaries and arterioles.[1] This vaso-occlusion, the predominant clinical manifestation of the disease, precipitates the painful crises and premature destruction of red blood cells or anemia (Figure 1). The pain is often severe and may be diffuse or localized and last from hours to 2 or more weeks. Serious related effects may include involvement (and eventual destruction) of the spleen, susceptibility to infection (including *Salmonella* osteomyelytis), occlusion of retinal vessels, bone and joint ischemia leading to aseptic necrosis (especially of the humeral or femoral head [Figure 2]), a hand-and-foot syndrome (in younger patients) caused by painful infarcts and dactylitis, priapism, and an acute, potentially catastrophic chest syndrome.[1,3,5]

At least 47 states now provide universal newborn screening for SCD,[4] and a number of guidelines have been developed for health maintenance of affected children, adolescents, and adults. Ongoing care is important in preventing crises and includes immunizations, prophylactic antibiotics, monitoring clinical and psychosocial status, patient education, avoiding strenuous physical activity and extremes of heat and cold, and maintaining adequate oral hydration at all times.

Treatment includes nonsteroidal anti-inflammatory drugs for less severe bone pain and narcotic analgesics for severe crises. Transfusion, antibiotics, oxygen, and hydration are standard care during crises. Hydroxyurea has become a therapeutic mainstay in severe SCD. This agent increases synthesis of fetal hemoglobin levels, which are not affected by sickling, has proven effective in treating patients with three or more painful crises a year, reduces incidence of the severe chest syndrome, and may improve long-term survival.[6,7] Oxygen therapy, adrenergic bronchodilators, and transfusions are effective for acute lung disease in sickle cell patients. The National Institutes of Health monograph, *The Management of Sickle Cell Disease*, is an excellent resource for those who want to explore the diagnosis, management, and other aspects of this disorder in detail.[8]

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Radiographic findings

Radiographic findings in sickle cell anemia are related to deossification due to marrow hyperplasia, thrombosis and infarction secondary to osteomyelitis, and growth defects. The marrow hyperplasia crowds and thins the trabeculae of osseous structures with resultant osteoporosis.

Findings in the vertebral column include softening of the vertebral bodies and multiple protrusions of the nucleus pulposus secondary to infarction of the vertebral body endplates, or biconcave compression (Figure 3). Trabeculae are sparce, and remaining vertical trabeculations show increased density.

In the long bones, widening of the medullary space with thinning of the long bones tends to disappear in adolescence, which presumably relates to the conversion of red marrow to yellow marrow in the extremities. The diploic spaces of the skull may thicken in the posterior parietal area (Figure 4) with a tendency toward radial trabeculation (hair-on-end appearance).

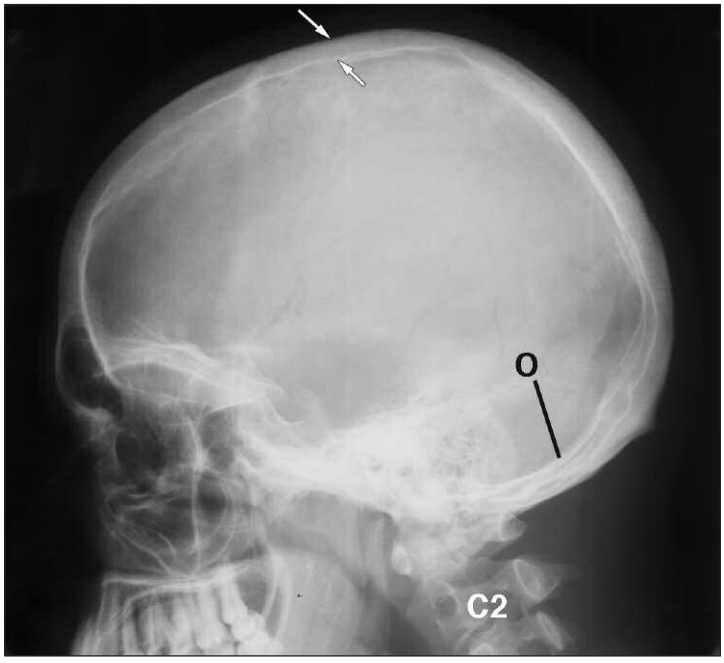

Figure 4 This lateral view of the skull displays the widening of the diploic space of the calvaria (arrows). Note the sparing of the occipital bone (O), which contains no marrow. C2 = second cervical vertebra.

Take-home message

Sickle cell anemia is a chronic, incurable disease that requires ongoing management with immunizations, prophylactic antibiotics, analgesic medications, and behavioral/coping therapies. Some crises can be managed at home with pain medicines, rest, and extra fluids. In especially intense crises, hospitalization with intravenous fluids, strong pain medications, and transfusions may be necessary. **FPR**

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