Severe Hypoxemia Secondary to Acute Sternal Infarction in Sickle Cell Anemia

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This article describes a 28-yr-old black man with sickle cell anemia who presented with severe chest pain secondary to acute infarction of the body of the sternum, hypoventilation, and hypoxemia with no evidence of acute chest syndrome. A bone scan performed 5 days after admission revealed increased uptake in the sternum, suggesting sternal infarction. Repeat bone scan performed 2 mo later demonstrated normal concentration in the sternum.

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Painful acute sickle cell crises most commonly affect the musculoskeletal system. Avascular necrosis, osteomyelitis, vertebral body collapse, and infarction of the long bones and ribs are the usual skeletal manifestations of sickle cell disease (*1–4*). The facial bones, especially the orbits, are uncommon sites of infarction (*5*). Localized infarction of bony segments of the sternum has been reported in children (*6*). In this report, we describe an adult patient with sickle cell anemia who presented with severe chest pain secondary to acute infarction of the body of the sternum which led to hypoventilation and hypoxemia with no evidence of acute pneumonia or infarction (no new infiltrate on chest radiography).

CASE REPORT

JE is a 28-yr-old black man known to have sickle cell anemia that was diagnosed in early childhood. Known complications of his disease, besides recurrent painful episodes, included a history of cerebrovascular accident secondary to subarachnoid hemorrhage with residual left hemiparesis and a history of seizure disorder controlled with Dilantin therapy.

On the day of admission he presented to the Emergency Room with severe pain involving his chest, back, abdomen, and legs. He gave no history of fever, cough, sputum production, hemoptysis or recent upper respiratory tract infection.

Pertinent findings on physical exam showed moderate distress secondary to pain, scleral icterus, oral temperature of 98.7°F, respiratory rate of 22/min with shallow and slightly labored breathing, regular heart rate of 86/min, and blood pressure of 110/70 mmHg. There was exquisite and severe tenderness over the body of the sternum and less severe tenderness over the anterior ribs and low back.

Because of his shallow and labored breathing arterial blood gases were determined. The pH was = 7.39 (n = 7.35–7.45), PO_2 = 40 mmHg (n = 83–108), PCO_2 = 45 mmHg (N = 34–45), bicarbonate = 28 MEq/liter (n = 22–27), CO_2 content = 29 (N = 22–27 MEq/liter), and O_2 saturation = 75% (n = 95–99). After the administration of 6 liters of oxygen via nasal cannula his PO_2 increased to 95 mmHg, PCO_2 decreased to 40 mmHg, CO_2 content decreased to 25 MEq/liter and O_2 saturation increased to 97%.

Hemoglobin on admission was 6.9 g%, hematocrit 20.1%, reticulocyte count 13.8%, and white blood cell count 18,200/μl with normal differential. Chest radiography showed mild congestive heart failure and no evidence of pulmonary infiltrates.

A ventilation/perfusion lung scan showed a matched defect at the right upper lobe with a low probability of pulmonary embolism. Whole-body bone scintigraphy, following an intravenous administration of 20 mCi of ^{99m}Tc-methylene diphosphonate (MDP), was performed 5 days after admission. There was decreased activity within the body of the sternum consistent with acute bone infarction (Fig. 1A). In addition, there was increased activity within the third left anterior rib and the sixth right anterior rib, indicating healing infarctions in these areas. Activity within both femoral shafts was nonuniformly increased consistent with healed infarctions. Moreover, there was relatively increased periarticular activity involving the shoulders, knees, and ankles, as is frequently seen in patients with sickle cell anemia. A follow-up whole-body bone scintigraphy with 20 mCi of ^{99m}Tc-MDP, was performed 2 mo later and revealed a normal looking sternum and increased tracer uptake within several midthoracic ribs posterolaterally consistent with healing infarcts (Fig. 1B).

Treatment consisted of bed rest, intravenous hydration, intranasal oxygen and narcotic analgesics. Hospital course showed fever up to 100.2°F during the first 2 days of admission and tachypnea up to 28/min on the first day of admission. Hemoglobin decreased to 4.4 g% on the sixth day of admission with a reticulocyte count of 16.8%. He received two units of RBC transfusion on the 7th day of admission. His painful crisis resolved gradually and he was discharged after 13 days of hospitalization. His arterial blood gases on room air before discharge were within normal limits.

DISCUSSION

Acute chest syndrome in sickle cell anemia is characterized by chest pain, fever, new pulmonary infiltrate, hypox-

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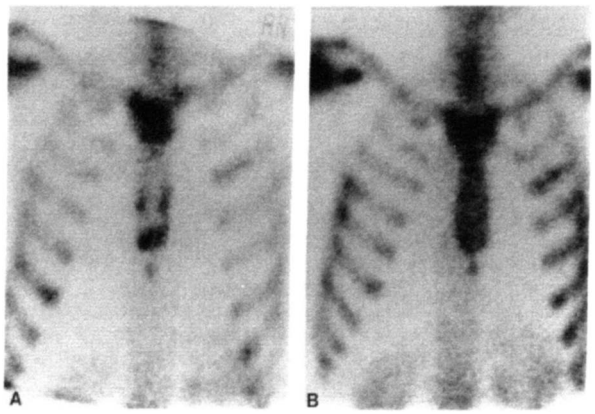

FIGURE 1. Following the intravenous administration of 20 mCi of ^{99m}Tc-MDP, a whole-body bone scan was obtained. Spot view of the chest (A) shows decreased activity within the body of the sternum which is compatible with acute bone infarction. A repeat scan (B) 2 mo later depicts healing of the sternal infarct. Also, notice healing infarction of the ribs.

emia, and decreased hemoglobin level (*7,8*). The sine qua non of the diagnosis of acute chest syndrome is the presence of pulmonary infiltrates on chest radiography, which may be due to pneumonia or pulmonary infarction (in situ thrombosis secondary to sickling). In the absence of pulmonary infiltrates the most likely diagnosis is acute painful crisis involving the musculoskeletal components of the chest. Our patient fits this latter category since he had no pulmonary infiltrates and had low grade fever only during the first two days of hospitalization which subsided without antibiotic therapy. The most likely cause of his hypoxemia is conscious hypoventilation due to severe chest pain in the sternal area upon inspiration. This is similar to the hypoxemia seen in patients with neuromuscular disease of the chest wall who hypoventilate because of weakness of the intercostal musculature.

It is well established that acute infarction in bone and bone marrow in patients with sickle cell anemia appears as areas of decreased accumulation of tracers such as ^{99m}Tc-MDP, ^{99m}Tc-sulfur colloid, and ^{67}Ga-citrate (*1,10,11*). Harcke et al. (*6*) described sternal infarction in children with sickle cell anemia and they have suggested how scintigraphy could play an important role in pointing out the sternal origin of the chest pain in these children. As the infarct heals, focal areas of decreased uptake return to normal after the hyperemic healing phase (*6,10*).

Another important finding in this patient is the severe anemia which developed during his hospitalization where the hemoglobin dropped to 4.4 g% thus requiring blood transfusion. Although aplastic crisis causing the anemia cannot be ruled out; decreased RBC production caused by massive sternal infarction is one possibility. In adults the sternum is a major site of active erythropoiesis and its infarction may accelerate the progression of anemia in a patient with sickle cell anemia whose red cell survival is already comprised.

SUMMARY

This case indicates that acute sternal infarction should be considered in patients who present with the clinical picture of acute chest syndrome without pulmonary infiltrates. Severe chest pain, hypoxemia, and worsening anemia are the major manifestations of acute massive sternal infarction.

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