Scintigraphic Manifestations of "Sternal Cupping"

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Sternal abnormalities in sickle-cell disease have been documented by bone scintigraphy and radiography in patients with homozygous sickle-cell anemia, but not in patients with sickle-thalassemia. We present here two unusual cases of sternal abnormalities in complicated sickle-cell disease. One is an infant with radiographic findings of "sternal cupping" and transient hypo-ossification of the sternum and sickle-thalassemia. The other patient is also a male infant with unusual, persistent under-ossification of bone, in association with radiographic findings of "sternal cupping." The second patient also had a 4P-chromosomal defect (Wolf-Hirschhorn syndrome) in which sternal hypo-ossification was described.

Key Words: sternal cupping; sickle-cell thalassemia disease; hypo-ossification; sternal infarction

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The classical clinical features and radiographic findings in sickle-cell disease have been well documented since the first description of the disease by Herrick in 1910 (*1*). The literature is replete with reviews of the sickle-cell disease process and resultant systemic changes. Involvement of nearly every bone in the body has been reported in radiological literature, but only rarely do articles appear concerning roentgenographic findings of sternal involvement. We present here two unusual cases with radiographic findings of sternal cupping; one in a patient with sickle-cell thalassemia, and the other in a patient with sickle-cell disease and Wolf-Hirschorn syndrome. There is only one report to date that describes nuclear bone scan manifestations of this abnormality (*2*).

CASE REPORTS

Patient One

A 14-mo-old male infant presented to the emergency room at Ben Taub General Hospital with a 2-day history of fever up to 102°F, lethargy and irritability. Review of systems was otherwise essentially negative. Family history included the child's mother, who has thalassemia. The child has had no previous unexplained fevers or crises. On examination, the child's temperature was 100.7°F rectally, and the only other abnormal finding was a palpable spleen.

The initial laboratory data included HGB 8.5 g/dl, HCT 25.2%, MCV 78.6 u^3/RBC, WBC 9700/mm^3. Hemoglobin electrophoresis showed the presence of hemoglobin F 48% and hemoglobin S 50%. The initial chest x-ray was normal. The patient was empirically started on the antibiotic cefoxitin intravenously after appropriate cultures were obtained. Due to persistent fever spikes, a bone scan was obtained to rule out osteomyelitis. The scan demonstrated an intense focal increase in activity in the midsternum (Fig. 1A), and focally increased activity in the right lower ribs anteriorly. A repeat chest x-ray demonstrated a right middle lobe infiltrate and the lateral view of the chest and sternum demonstrated sternal cupping of the second segment of the sternal body (Fig. 1B). Because of poor response, the child was switched to erythromycin and chloramphenicol and became afebrile on the third day of therapy. A repeat sternal radiograph 5 mo later revealed a more normal appearance (Fig. 1C) of the mid-sternal segment.

Patient Two

This patient, also a 14-mo-old male, presented with a 2-day history of fever up to 102°F. He was previously diagnosed as having sickle-cell disease and had six previous admissions for fever and/or painful crises. At 5 mo of age he was investigated genetically for an intrauterine growth retardation and microcephaly and was found to have Wolf-Hirschhorn syndrome (chromosome 4P defect), in which the short arm of the chromosome 4 is missing. In addition to multiple reported radiographic findings, anomalies of sternal ossification have been specifically noted (*3*). The patient underwent bone scanning to evaluate bone pain in the distal legs. Focal increased activity in the second sternal segment junction was noted in addition to decreased activity in the first segment (Fig. 2A). The CXR demonstrated absence of ossification of the first sternal segment (Fig. 2B). A follow-up x-ray 4 mo later demonstrated persistent absence of ossification in the first sternal segment, but widening and cupping of the second and third sternal segments (Fig. 2C). This finding and the decreased activity of this segment on the bone scan can be attributed to sternal ossification abnormalities described in Wolf-Hirschhorn syndrome.

DISCUSSION

The sternum is composed of four segments during development, which eventually fuse to form the body of the sternum between puberty and 25 yr of age. A fifth and sixth segment form the xiphoid and manubrium, respectively. Bone infarction in sickle-cell disease is presumed to be secondary to sludging in the small blood vessels supplying the

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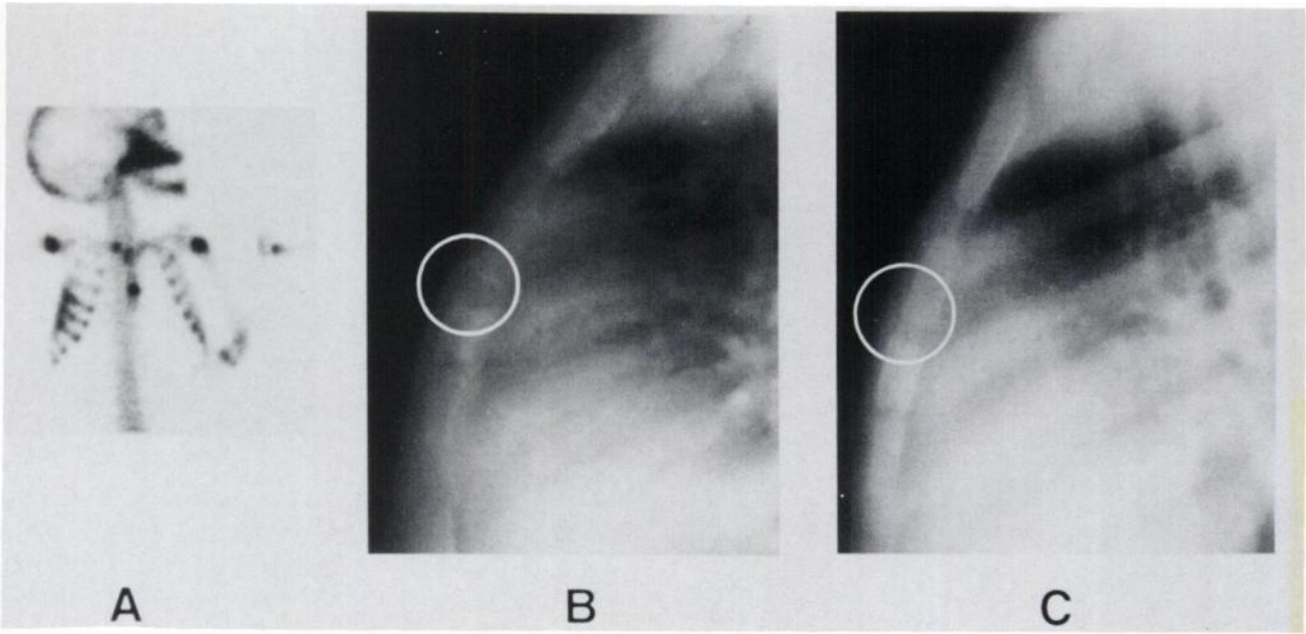

FIGURE 1. (A) Delayed bone scan demonstrates a focal area of increased uptake in the region of the second sternal segment. There is also increased activity in the ribs overlying a right lower lobe pneumonia. (B) Sternal cupping is apparent with widening, sclerosis and irregularity of the second sternal segment (circle). Some irregularity is also noted at the inferior margin of the second sternal segment. (C) Sternal cupping has almost resolved 5 mo later. The sclerosis of the second sternal segment is still present, suggesting infarction.

sternum, particularly during crisis. The cupping is thought to represent reactive bone formation following infarction (*4*). Radiographically, sternal cupping is seen as well-demarcated cupping and is noted to occur most frequently along the inferior margin of the first sternal segment. Cupping was not observed below the second sternal segment and multiple sternal segment involvement was noted in about half of the patients (*4*). Osteomyelitis or infarction of the sternum in patients with sickle-cell disease has rarely been reported. Bone scanning in these patients should not be used solely to evaluate radiographic manifestations of sternal cupping, but may be beneficial in patients with significant soft-tissue swelling and pain, or other areas of concern.

Levine et al. reviewed a total of 200 patients with either homozygous sickle-cell anemia or heterozygous sickle-cell variants for sternal cupping. This abnormality was found in 8% of patients with sickle-cell anemia and 10.6% of those with sickle-cell-hemoglobin C disease. It was not noted in patients with sickle-cell thalassemia, sickle-cell trait or normal children. The age range in which this abnormality was noted was 1.25 to 13 yr.

The cause of sternal segment cupping in children with sickle-cell disease is uncertain. One possibility is demineralization and weakened body structures secondary to marrow hyperplasia. This mechanism is the one postulated to explain central depression in the vertebral body end plates. However, the sternum is not a weight-bearing structure. Also, unlike the vertebral disorder, sternal cupping is a temporary phenomenon, a finding that does not support the foregoing hypothesis. A more reasonable explanation is that sludging of the sickled cells in the terminal arterioles leads to stasis and thrombosis, causing ischemia of the sternum. The actual cupping noted may be secondary to subclinical collapse of the sternal segments. Apparent resolution on follow-up x-rays may be secondary to collateral circulation (*4*). The explanation of the nuclear findings is equally puzzling. Increased localization of the bone tracers in the bone scan is most probably in the border zone of the cupping, where reparative osteoblastic changes take place, rather than in the cupping. This explanation fits nicely with the radiographic observation of a "blastic" process.

Although the finding of focal increased activity in the sternum most probably represents the manifestations of a benign process such as infarction, mention of this abnormality in the nuclear report is important. A lack of symptoms relating to the sternum would suggest an old infarction. However, in the acute stage, while differentiation from osteomyelitis can be difficult, the bone scan, when abnormal, can be useful in explaining sternal abnormalities as a cause for chest pain in patients with painful crises.

The effects of abnormal hemoglobin are noted in nearly every system of the body. Skeletal changes are usually due to chronic anemia and the resultant hyperplasia of the marrow, in addition to abnormalities caused by the sickling of the red blood cells, including infarction, osteomyelitis, marrow expansion, joint space narrowing and growth retardation (*5,6*). Nuclear medicine has been used to evaluate skeletal, marrow, hepatic and splenic abnormalities related to sickle-cell disease (*7–9*). The efficacy of the nuclear bone scan in differentiating infarction from

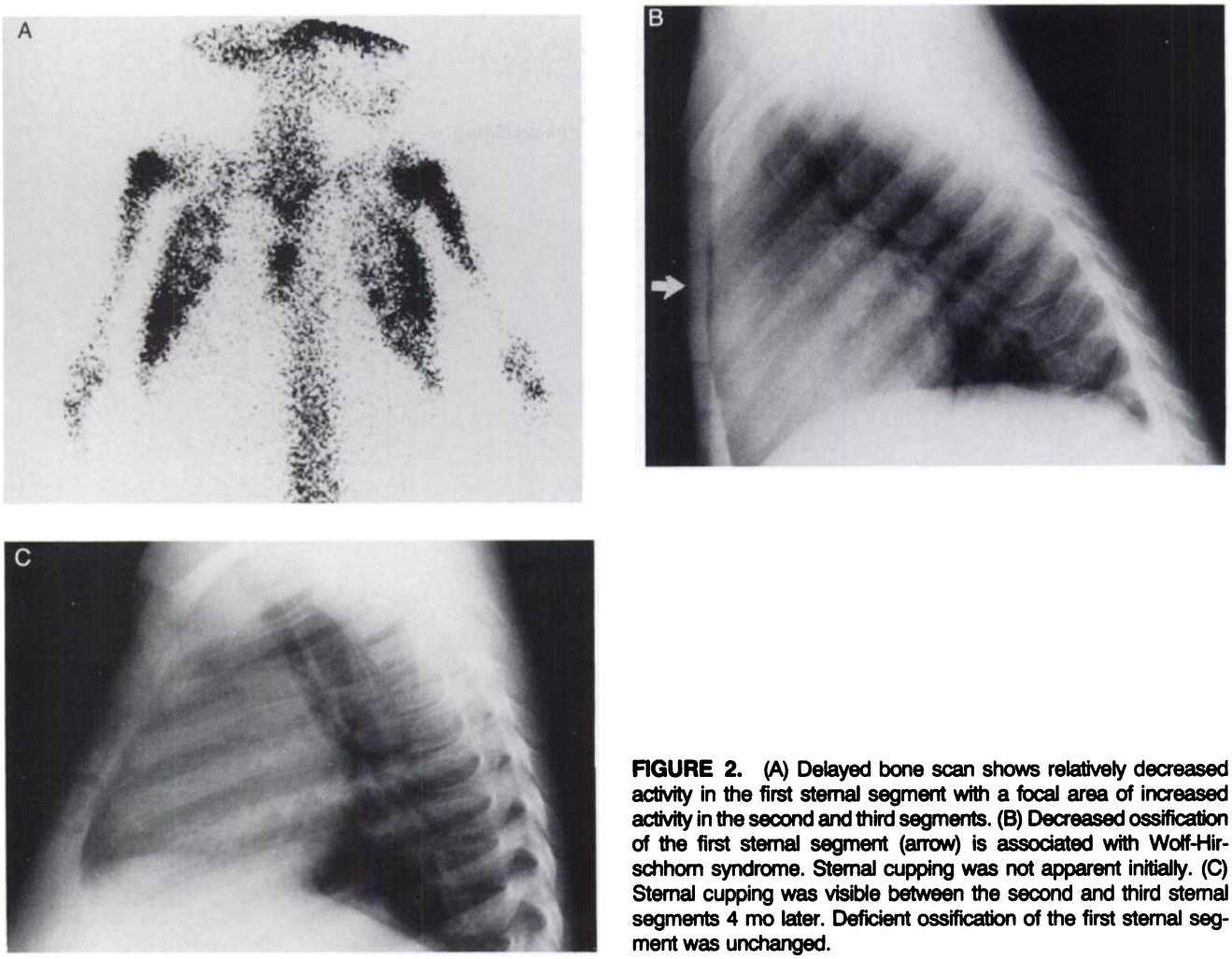

FIGURE 2. (A) Delayed bone scan shows relatively decreased activity in the first sternal segment with a focal area of increased activity in the second and third segments. (B) Decreased ossification of the first sternal segment (arrow) is associated with Wolf-Hirschhorn syndrome. Sternal cupping was not apparent initially. (C) Sternal cupping was visible between the second and third sternal segments 4 mo later. Deficient ossification of the first sternal segment was unchanged.

osteomyelitis in the acute phase has been questioned, as old infarctions could simulate infection. With the concurrent use of gallium scans, specificity has improved (*9*). Skeletal manifestations of sickle-cell disease on bone scans are also related to bone marrow expansion, infection or infarction. Diffusely increased activity in somewhat widened metaphyses is commonly noted in children and young adults with marrow expansion due to severe anemia or malignancy. There may be a significant peripheral extension of the marrow even in adults. The bone scan findings in osteomyelitis, consisting of increased perfusion blood pool and increased osteoblastic metabolic activity in the delayed (third phase) images in a focal region, have been discussed in numerous reviews (*7,10,11*). The spectrum of manifestations of bone infarctions varies from decreased activity in the case of early avascular necrosis or infarction, to increased activity in a healing infarction commonly manifested by sclerosis on radiographs. Infarctions in the bones of the hands and feet giving rise to dactylitis, also yields a focus of increased or even decreased activity on scintigraphy. Bone scan findings in the sternum have been reported in six patients with homozygous sickle-cell anemia (*2*).

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