

SICKLE CELL DISORDERS AND THE SKELETAL SYSTEM

To the Editor:

I read with keen interest the well-written and comprehensive review article on sickle cell disorders (SCD) by Behrens and Cymet in the September 2000 issue of *Hospital Physician*.¹ Under the subheading, "Other Chronic Manifestations," there was a brief mention of chronic pain in bones, joints, and extremities; osteosclerosis; osteoarthritis; and skeletal malformations secondary to bone infarction at a growing epiphyseal plate.

My colleagues and I have reported on a case of a 15-year-old Nigerian boy with the hemoglobin SS genotype, multiple faceted gallstones visible on plain radiographs, and the first documented occurrence of a "vanishing vertebra," in which there was virtual radiological disappearance of the L3 vertebral body.² I had coined the term "vanishing vertebra syndrome" at the time, to depict the radiologic absence of an L3 vertebral body in a plain radiograph of the spine. Sickle cell hemoglobinopathy is common in Nigeria, affecting about 2% to 3% of children.³

Clearly, repeated cycles of bone infarction following sickling-induced anoxia, with or without secondary osteomyelitis, and associated marrow hyperplasia induced by chronic hemolysis all contribute to chronic bone pathology in SCD. The "vanishing of vertebrae," spondylitis, pseudo-Schmorl's nodes, narrowed intervertebral disc spaces, and fish-mouth deformities of vertebral bodies seen in SCD can be explained on the basis of vaso-occlusive infarction, secondary infection, compressive forces along the spine, and reactive bone processes. Although it is called the "vanishing vertebra," this phenomenon is only an apparent radiologic disappearance, as the radiolucent osteoid of the "vanished" vertebra will necessarily still be present in the spine.

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In reply:

We appreciate Dr. Onuigbo's comments regarding our recent article.¹ The radiographs shown in his article are fascinating.² There truly are more findings in sickle

cell disorders than can be summarized in one review article. We agree that the most likely etiology for the "vanishing vertebra" is recurrent vascular occlusion.

The more common "fish-mouth" vertebrae are also caused by vaso-occlusion. Fish-mouthed vertebrae are formed because short, perforating branches of the periosteal vessels that supply the periphery are not as prone to vaso-occlusion as the long branches of the vertebral nutrient arteries that supply the center of the end plate.^{3,4} As we discussed in our review,¹ sickling and vaso-occlusion are more likely to occur the longer erythrocytes spend deoxygenating in the cramped, acidic environment of the capillaries. Therefore, the edges of the vertebrae extend normally because their vascular supply is less frequently compromised; whereas, the centers of the vertebrae are depressed and introverted.

Another common bone change in patients with sickle cell disorders is bony enlargement caused by marrow hypertrophy. This is frequently seen as the gnathopathy (prominent maxillary overbite) caused by maxillary bone overgrowth, and also as tower skull caused by prominent frontal bones.³

Serjeant, Ennis, and Middlemiss have published a series of articles with an excellent summary and pictures of common skeletal abnormalities accompanying sickle cell disorders.^{5,6,7}

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