Images in Clinical Tropical Medicine

Hair-on-End

Bridget Mnapo, Michael Shields, and Klaas Koop*

Thyolo District Hospital, Thyolo, Malawi; Queen Elisabeth Central Hospital, Blantyre, Malawi

FIGURE 1. (A) Skull x-ray with a hair-on-end pattern, reflecting bone marrow hyperplasia. (B) Peripheral blood slide with a Howell-Jolly body in an erythrocyte (arrow). (C) X-ray of pelvis showing left femoral head necrosis.

A 25-year-old man was admitted with a long history of weakness and wasting. In fact, he said he “was born—and fell ill.” He claimed to be still unmarried as a result of his illness. One year earlier he had been admitted with pain in his left hip. Despite several hospital admissions, an explanation for his illness had not been found.

He was wasted, pale, and slightly jaundiced. His chest, abdomen, and extremities appeared normal. His forehead and maxillae were prominent, and this prompted us to take an x-ray of his skull.

The x-ray (Figure 1A) showed a hair-on-end pattern compatible with a hemoglobinopathy—the skull changes reflect bone marrow hyperplasia. A peripheral blood slide revealed sickle cell disease. Several complications were noted: anemia, hyposplenism (Howell-Jolly bodies, Figure 1B), nephropathy (protein 2+ on dipstick). And, in hindsight, the pain in his left hip was caused by femoral head necrosis (Figure 1C) during a sickle cell crisis.

Sickle cell disease is a relatively common condition in Malawi and many other African countries: in one study 2.5%

of infants in southern Malawi were found to have the HbSS genotype.¹ The mortality among children with sickle cell disease is very high, primarily because the disease is under-recognized.² Indeed, in our patient sickle cell disease was diagnosed some 20 years too late, after the onset of several severe complications. Early recognition coupled to relatively simple interventions such as vaccinations, prophylactic antibiotics, and disease education has proven to reduce the morbidity and mortality of sickle cell disease patients.

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Authors' addresses: Bridget Mnapo, Thyolo District Hospital, Thyolo, Malawi, E-mail: bchisomo@gmail.com. Michael Shields, Queen Elisabeth Central Hospital, Blantyre, Malawi, E-mail: shields_m_l@hotmail.com. Klaas Koop, Thyolo District Hospital, Thyolo, Malawi; current address: Meander Medisch Centrum, Department of Pediatrics, Amersfoort, The Netherlands, E-mail: klaaskoop@gmail.com.

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*Address correspondence to Klaas Koop, Meander Medisch Centrum, Department of Pediatrics, PO Box 1502, 3800 BM Amersfoort, The Netherlands. E-mail: klaaskoop@gmail.com