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Multiple pathologic fractures in a 19-month-old boy with sickle cell disease

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Abstract

Multiple pathologic fractures in a patient with sickle cell disease is rare; bone involvement is well documented in this disease, however multiple pathologic fractures as a complication are uncommon. We present a case involving a 19-month-old boy known to have sickle cell disease who developed multiple pathologic fractures as a complication of his disease. The patient was treated conservatively with excellent outcome.

Keywords

Sickle cell; pathologic fractures.

Background

Sickle cell disease is an inherited autosomal recessive disorder of the β-globin gene caused by mutation in position 6 with replacement of glutamate by valine[1]. Among other complications, bone involvement is a well-documented feature in this disease[2]. Different anatomic sites are affected with variable manifestations and outcomes[3–6].

One of the most common forms of bone involvement is osteomyelitis[7]. patients with sickle cell disease have an increased incidence of osteomyelitis compared with the general population[8]. Osteomyelitis is documented as a complication of sickle cell disease in studies from different parts of the world[7,9–12]; the prevalence is as high as18% in a study by Buison et al.[9] from Cameroon, and as low as10 in 2000 in one study from the United States[7]. The common etiologic factor is *Salmonella*[7,13] although some studies have reported that *Staphylococcus aureus* is the predominant microorganism[14]. Osteomyelitis and bone infarction commonly occur concomitantly and differentiation between them is difficult using radiographs and bone scanning[7,15].

Other sequelae of sickle cell disease include vascular necrosis of the bone[16]. The prevalence of this morbidity was reported to be 26.6% in one study[16]; another study documented 464 sites of bone and bone marrow infarctions among the patients studied. Those authors concluded that knowledge of the distribution of the sites of bone infraction is of considerable clinical and

diagnostic importance in the ongoing evaluation and treatment of sickle cell hemoglobinopathy[17]. Vascular necrosis of the bone was associated with a higher number of hospital admissions and a higher rate of painful crises[18,19].

Osteopenia and osteoporosis are associated with sickle cell disease[20–24]. Iron overload has been correlated with this complication[20,21,25] and the explanation for this might be the inhibition of osteoblast proliferation and differentiation by iron[26]. Other correlations include low body mass index[21,27], low serum level of zinc[22] and low hemoglobin[28]. However, no association has been found between low bone mineral density and vitamin D status[24,27], which suggests abnormal bone formation as an underlying mechanism. In agreement with this is the fact that treatment of patients with sickle cell disease with vitamin D and calcium restores 25-hydroxyvitamin D levels to normal but markers of bone resorption remain unclear[29].

Osteomyelitis, a vascular necrosis of the bone, and ostopenia are associated with increased risk of pathologic fractures in sickle cell disease[4,14,30].

Case report

A 19-month-old boy, diagnosed with sickle cell disease at the age of 7 months, was homozygous for the hemoglobinopathy. He had a history of repeated hospital admissions because of anemia and he had undergone 4 transfusions since birth. He was brought to the emergency department because he was ill and inactive.

The parents noticed that the baby was ill and cried especially when they tried to hold him or make him setting up. They gave no history of trauma or falling; they mentioned that he does not go to daycare and is always at home with the mother; she denied any falls or trauma during bathing, playing or falling from the bed. There was no history of a similar condition.

On first assessment, he was conscious, ill, in pain and adopted an adductive position for the upper and lower limbs. Respiratory rate was 30/min. Pulse rate was 132/min. Temperature was 38.7°C. There was no bruises, scratches, lacerations or any other marks on the skin and no swelling or wounds on the skull. He was slightly jaundiced and pale. His chest was clear. The liver was palpable 2 cm below the costal margin, soft and not tender. The spleen was also palpable 2 cm below the costal margin. His limbs were a bit swollen, warm, very tender and he avoided movement and touch. His developmental status was normal for his age.

His laboratory findings were as follows: Hb was 4 g/dl, total white cell count was 12×10^9/l, and the platelet count was 414×10^9/l and the reticulocyte count was 3%. The peripheral blood film showed a dimorphic picture with sickle cells. Liver biochemistry, creatinine, electrolytes and urine analysis were normal.

Abdominal ultrasonography showed an enlarged liver with normal texture and no local lesions, splenomegaly 13 cm, no gallstones and normal kidneys. Plain radiographs of the lower limbs and pelvis showed multiple fractures at different anatomic sites including the right and left upper femur (Fig. 1), right and left lower femur (Fig. 2) and right proximal tibia and fibula (Fig. 2). Fractures at all sites were simple fractures.

The differential diagnosis of multiple fractures includes trauma, child abuse, pathologic fractures. No history of trauma was given and there was no previous history of trauma. Regarding the possibility of child abuse, from the history, the parents emphasized that there was no history of trauma making the possibility of child abuse unlikely. On examination there was no evidence of violence and from the radiographs, all the fractures were at the same age, which again weakened this diagnosis. In addition, there was no history of a previous similar condition.

A pathologic fracture by definition is a fracture that occurs on a diseased bone without trauma or after minor trauma, and bone diseases such as osteomyelitis and vascular necrosis of the bone are well-known complications of sickle cell disease.

Management

The patient was admitted, transfused and received intravenous ciftrixone for 3 weeks, followed by oral augmentin for another 3 weeks. The fractures were managed conservatively, first by avoiding movement and relieving pain with paracetamol. After 3 weeks we encouraged the parents to mobilize him gradually; after 3 months he was able to walk alone without pain and his serial radiographs showed good bone healing (Fig. 3).

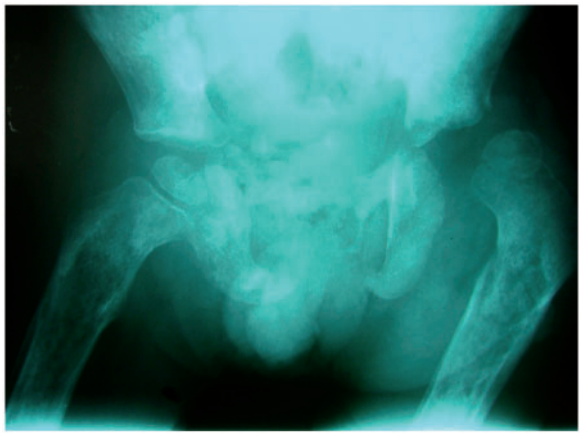

Fig. 1. Multiple bilateral fractures in the upper femurs with multiple osteolytic areas occupying the subtrochanteric region and the shafts.

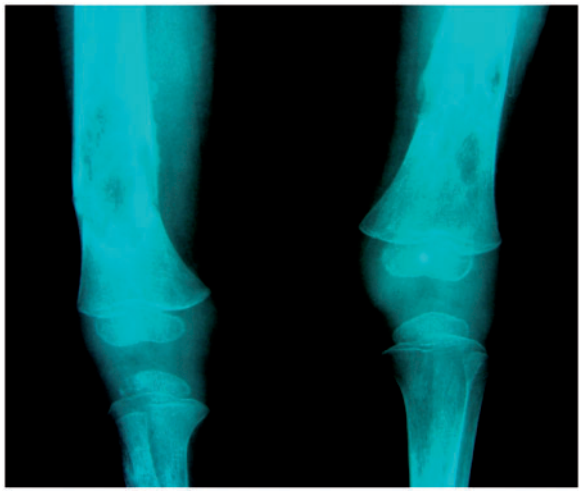

Fig. 2. Fracture in both lower femurs, right proximal tibia and fibula.

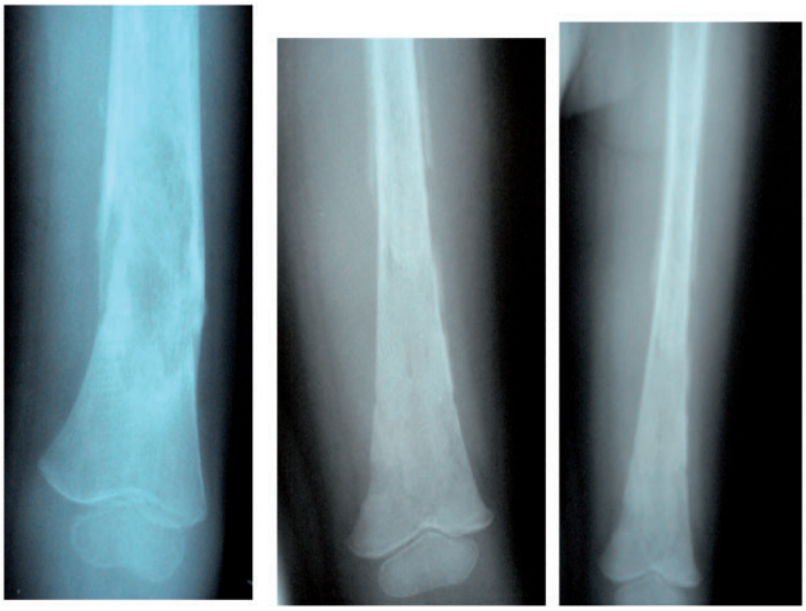

Fig. 3. Evidence of healing of fractures in both lower femurs 6 weeks later.

Discussion

Pathologic spontaneous fractures are fractures through weak or diseased bones of abnormal composition; most of the causes are benign[31]. The commonest cause of pathologic fracture in white children in most reports is simple or unicameral bone cysts[32]. In data from Africa, the most common cause is osteomyelitis[33], which is a well-known complication of sickle cell disease[2,3].

Pathologic fractures have been reported in patients with sickle cell disease who have osteomyelitis[34] or in occasional cases when osteomyelitis complicates pathologic fracture later after healing[35]. Pathologic fracture was reported as a comorbidity in sickle cell disease, where it accompanied splenic abscess[36].

Long bones are affected as well as the vertebral column[37,38]. This bone involvement usually indicates severe disease[39–41] and has been reported from different geographic locations[8,32,42,43], which reflects that it is universal in the natural history of the disease and hence represents a major chronic morbidity that may need invasive management, which itself may carry more risk to the patient[44].

In all the reports reviewed, the pathologic fracture involved one anatomic site with the exception of one report[34] that described bilateral fractures, but not in a patient with sickle cell disease. Here we report multiple pathologic fractures in a young patient with sickle cell disease. To our knowledge, multiple pathologic fractures in sickle cell disease have not been reported previously.

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