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University of Medical Science

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ABSTRACT

Bone infarction is a common presentation in sickle cell disease. Vertebral osteonecrosis is however not commonly reported, especially among children. We report a case of a 6 year old boy diagnosed with sickle cell anaemia, presenting with vertebral osteonecrosis involving multiple lumbar vertebrae. This report highlights vertebral osteonecrosis in childhood sickle cell anaemia which is an uncommon manifestation of sickle cell anaemia in the paediatric age group.

Author α ρ: Department of Paediatrics, University of Medical Science, Ondo, Nigeria.
Σ §: Department of Haematology and Blood transfusion, University of Medical Science, Ondo, Nigeria.
Ω χ: Department of Radiology, University of Medical Science, Ondo, Nigeria.
¥: Department of Microbial Pathology, University of Medical Science, Ondo, Nigeria.

I. INTRODUCTION

Sickle cell disease is a genetic disorder, and its mode of inheritance is autosomal recessive.[1] It is characterized by the inheritance of mutant haemoglobin genes from both parents. The disorder manifests with abnormally shaped red blood cells due to the distortion of the haemoglobin structure. The abnormally shaped red blood cells have reduced flexibility which results in occlusion within the microcirculation with resultant vaso-occlusion and bone ischaemia.

The bone is commonly involved in the manifestation of sickle cell disease.[2] Repetitive vessel occlusion involving most organ of the body including the skeleton is characteristic of the disease. Infarction, thrombosis, infection and marrow hyperplasia are some of the common bone manifestation of sickle cell disease.[3] There is usually an interplay between infarction resulting from repeated vaso-occlusion and increased susceptibility to infections, as the areas of necrosis serves as nidus for growth of organisms.[1] Avascular necrosis involving the femoral head is common in children and its prevalence increases with age.[4] Spinal necrosis is not common in children, especially in the younger age group. The clinical presentation of low back swelling and pain could connote tuberculosis infection in areas with high burden of the disease. We report this uncommon skeletal manifestation of sickle cell disease in a school age Nigerian boy.

II. CASE REPORT

A 6-year old boy with sickle cell anaemia presented at the pediatric haematology clinic of UNIMEDTH with two week history of pain and swelling over the lower back and pain involving both lower limbs. Swelling over the back had not significantly increased since it was noticed. Pain in the limbs was severe enough to cause impairment in walking and standing in an erect posture. There was no history of fever, cough, weight loss, night sweats or trauma to the lower back. He was diagnosed of sickle cell anaemia at four years and has had a fairly regular clinic attendance. He has had seven hospitalizations, mostly for vaso occlusion and haemolytic crisis. He has also had four previous blood transfusions. His last hospitalization was two months prior to the present complaint for which he was managed for vaso occlusion and haemolytic crisis with sepsis.

Physical examination revealed no fever (T-36.9◦C), there was obvious swelling with kyphosis over the lower back in the lumbar region (Fig 1) which was not tender, with no differential warmth. There was no tenderness along both lower limbs. There was normal muscle bulk.

Power and reflexes were normal in both lower limbs. He was unable to stand erect, had an anteflexion posture with an abnormal gait. He had hepatosplenomegaly. All other systemic examination findings were normal.

Blood investigation revealed a packed cell volume of 20%, raised total white blood count of 26.2 times 10^3mm and erythrocyte sedimentation rate (ESR) of 4mm/hr (1-10mm/hr). Mantoux test result was 1mm (0-4mm). A spine X-ray (Fig 2) revealed reversal of normal lumbar lordosis, osteolysis of L2-L4 with reduction in vertebral size. There was reduced intervertebral space with collapse at L2-L3. Magnetic resonance imaging and bone scans were not done in this patient, on account of financial constraints and limitation of facilities for bone scan. A diagnosis of Vertebral Osteonecrosis in childhood sickle cell anaemia was made based on clinical findings and the radiological investigations we were able to do in this patient

Conservative management included rest, analgesics, antibiotics and skeletal support with the use of lumbar braces. Patient subsequently got relieved of the pain in the lower limbs, with improved gait and was able to maintain an erect stance. Mild swelling still persists over the lower back, being site of angulation of the collapsed vertebrae. Patient is currently being followed up in the orthopaedic and haematology unit.

IV. DISCUSSION

Back pain is uncommon in the pediatric and adolescent age group unlike adults in whom it occurs commonly.[5] The presentation of low back pain however uncommon in children could suggest osteonecrosis in a child with sickle cell anaemia. The lumbar vertebrae have been reported as the commonest site of spine infarction among adults and children and this was demonstrated in this patient. Multiple spinal vertebrae involvement could occur in osteonecrosis. This patient had multiple spinal lumbar vertebrae involvement with associated vertebral collapse.

Bone infarction is a debilitating illness with significant impact in children with sickle cell anaemia. The repeated sickling of red blood cells results in irreversible and rigid red cells with resulting obstruction in microcirculation. This resultant effect of tissue ischaemia and hypoxia results in eventual cell death in the affected bone.
The presentation of bone infarction and infection could be difficult to distinguish apart from each other, due to similarity in clinical presentation. Spinal infections such as spondylitis and acute osteomyelitis occur commonly among people with sickle cell anaemia. Acute osteomyelitis is usually due to Staphylococcal aureus and Salmonella infection. In the case presented, fever was absent, and ESR was not elevated. However this patient had an elevated total white blood count. The patient had a mantoux test done to exclude tuberculosis, considering its high burden among the study population. There were no other features deferrable to tuberculosis in this child, and mantoux result was within normal limits, thus anti-tuberculous therapy was not considered in this patient.

Magnetic resonance imaging (MRI) is considered as the best imaging option when pathology involving the spine is being considered.[5] Marrow infiltration and H-shaped vertebrae as a result of central growth plate infarction has been reported as MRI findings in vertebral bone osteonecrosis.[6,7] Lumbar-sacral spine x-rays are useful in diagnosis where there are constraints to obtaining a MRI as it was in the reported case. The fish vertebrae sign and vanishing vertebrae have been demonstrated on X-ray.[8] The differences in the blood supply of the central part of the vertebrae and the peripheral parts are responsible for this findings. The longer arteries supplying the central vertebrae are more affected by infarction and the consequent bone destruction.[9] The findings of osteopenia, with reduced bone height, and vertebral collapse were highly suggestive in this case.

Vertebral osteonecrosis is amenable to conservative management including bed rest, analgesia and use of vertebral braces as it was in this case. A course of antibiotics was considered in this patient on account of elevated WBC. There is

a possible interplay of infection and infarction in sickle cell disease, also bone infarction and necrosis creates a nidus for bacterial growth and spread.[1]

Emodi anoye[10] had earlier reported two cases of nine year old Nigeria girls with sickle cell anaemia that had vertebral collapse, both were managed conservatively similar to this study. The patient in this study is however 6years and thus younger than the cases they reported. In a retrospective study of osteoarticular manifestation of sickle cell anaemia in Nigeria children, Chinawa et al reported vertebral collapse in one child in the 6-10years age group out of twenty –five children with musculoskeletal complications of sickle cell anaemia. However details of clinical presentation and management was not discussed in their study

V. CONCLUSION

Vertebral osteonecrosis is an uncommon presentation in children with vaso-occlusion. It should be considered in young children with sickle cell anemia, presenting with low back pain. Careful evaluation and exclusion of other common causes of low back pain should be done with imaging and other relevant studies. Early orthopaedic consultation and conservative management as seen in this case is associated with good patient outcome.

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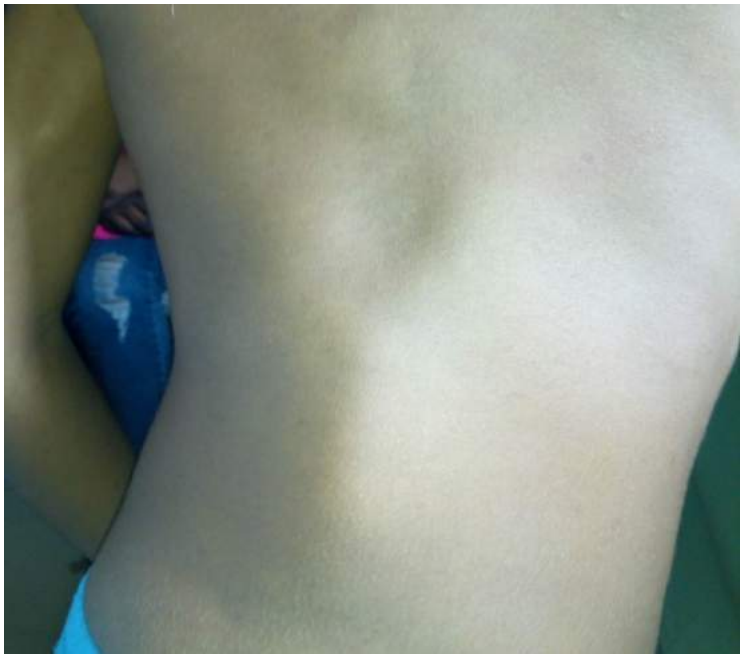

Figure 1: Swelling over the lower back and lateral curvature of the spine

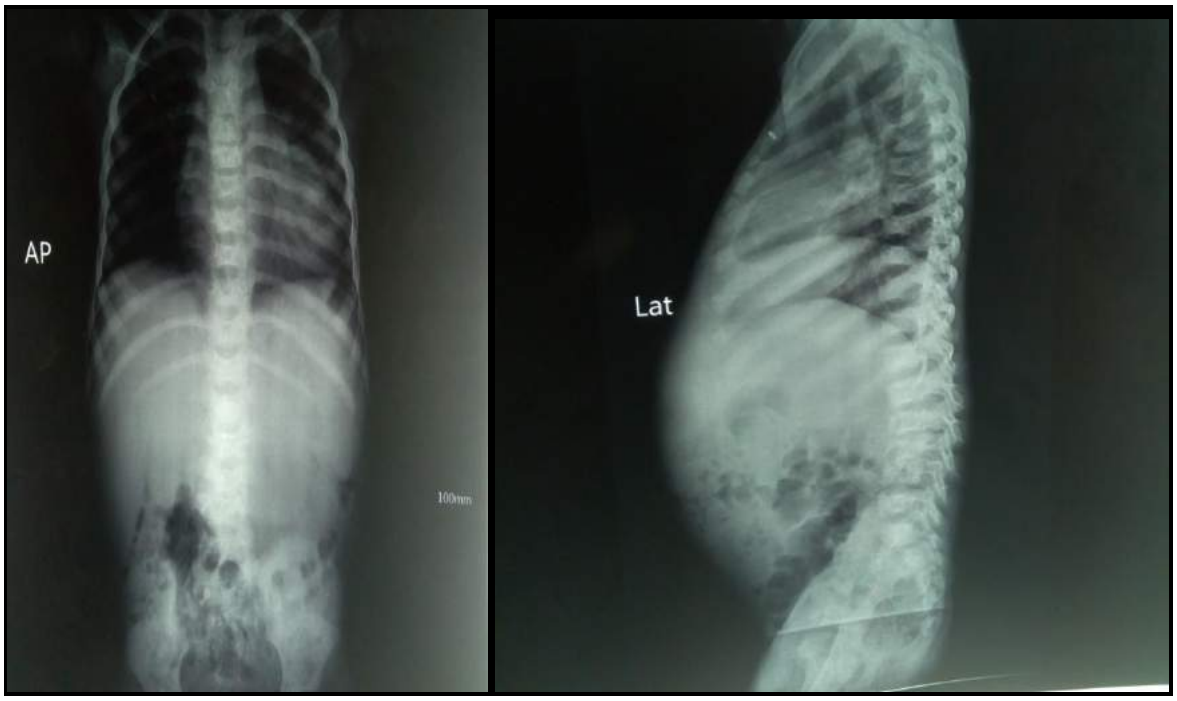

Figure 2: AP and lateral X-ray view of spine showing mild scoliosis, reduction in vertebral heights L2-L5 with irregular outlines and reduction in intervertebral disc space with collapse of L2-L3