Doi: 10.32677/IJCR.2017.v03.i03.014

Case Report

An unusual case of spinal manifestations of sickle cell disease

Chetana Ratnaparkhi, Sushupti Milind Chaukhande, Avinash Dhok, Devashis Barick

From Department of Radiology, NKP Salve Institute of Medical Sciences, Nagpur, Maharashtra, India

Correspondence to: Sushupti Milind Chaukhande, NKP Salve Institute of Medical Sciences, Nagpur, Maharashtra, India. E-mail: sushupti90@gmail.com

Received - 26 March 2017 Initial Review - 06 May 2017 Published Online - 22 July 2017

ABSTRACT

Sickle cell disease (SCD) results from the presence of abnormal beta globin chains within hemoglobin. Overtime, the disease produces various spinal abnormalities as a result of chronic anemia which include marrow hyperplasia, reversion of yellow marrow to red marrow, and occasionally extramedullary hematopoiesis. Vaso-occlusion may manifest as ischemia and infarction whereas superimposed infection may lead to osteomyelitis. There is paucity of literature illustrating the spinal manifestations of the SCD in a single case. Hence, in the present work, we report the case of a 30-year-old male with SCD who presented with almost all the manifestations of the disease in the spine.

Key words: *Bone infarction, Magnetic resonance imaging, Sickle cell disease, Spinal manifestations*

Sickle cell disease (SCD) is an autosomal recessive genetic condition in which a defective form of hemoglobin (Hb), (Hbs), results from a single amino acid substitution (Valine for glutamic acid at position 6) in the globin gene [1]. Bones are the second most affected organs by SCD, after the spleen [2]. However, bone involvement is the most common clinical manifestation of the SCD [3]. Spine represents the second most common area of bone involvement (26%) exceeded only by Knee (35%) [4]. The vertebra affected are lumbosacral in 66% of the cases, followed by thoracic in 22% and cervical in only 12% [4]. Minimal physical signs are noted, other than a local tenderness over the spinous process in 71%, and a decreased range of back motion in 17% cases [4].

The different spinal manifestations in SCD include diffuse marrow replacement from anemia, epidural abscess, osteomyelitis/discitis, facet osteomyelitis, bone infarct, H-shaped vertebra, fish-mouth vertebra, and tower vertebra. We also reviewed the literature which illustrated the spinal manifestations of the disease in different patients of SCD. However, there was scarcity of literature illustrating all the spinal manifestations of the SCD in a single case [5]. Here, we discuss the radiographic and magnetic resonance (MR) findings of spine in a case of sickle cell anemia. The unique nature of this case is that all the spinal manifestations of SCD were seen in a single patient.

CASE REPORT

The patient was a 30-years-old male, a known case of SCD (SS pattern) who came with the chief complaints of severe low back pain radiating to the right hip and swelling over it for 1 month. Pain and swelling had increased in the past 8 days and the pain was aggravated on sitting or standing. The patient gave history of 2 blood transfusions in the past. He was hospitalized and treated for subacute intestinal obstruction and pneumonitis 5 years back. He had undergone total left hip replacement for avascular necrosis of the femoral head 1 year back.

On local examination, diffuse swelling was present over back region. Straight leg raising was 90° on right side and 80° on left side. Power was 3/5 in right knee, ankle, and toe. Knee, ankle, and plantar reflexes were normal on both sides. On general examination, pallor and icteric tinge was noted. Vitals including pulse and blood pressure were within normal limits. On per abdominal examination, tenderness was present in the right hypochondrium. However, there was no organomegaly.

His blood investigations revealed reduced Hb (5.8 g/dl) levels and raised total leukocyte count (17500/cu mm) with neutrophil predominance. On X-ray imaging of the lumbar spine, osteopenia with end plate sclerosis of all the lumbar (L) vertebrae was noted (Fig. 1) along with the reduction in height of L4 vertebra. Mild scoliosis of the lumbar spine was also noted with convexity toward right side.

On MR imaging of the spine, old bone infarcts were noted in the lumbar and thoracic (T3, T6, T9, T10, T11, and T12) vertebral bodies and posterior elements of the cervical (C4), and T1, T5, T7, T9, L1, L2, and L4 vertebrae (Fig. 2). Marrow conversion was noted in the form of hypointensity on T2-weighted (T2WI) and short tau inversion recovery (STIR) and isointensity on T1WI in visualized spine including sacral vertebrae (Fig. 2). Paravertebral abscess formation was seen bilaterally along the L3, L3-4, and L4 vertebrae appearing hyperintense on STIR and T2WI and hypointense on T1WI showing peripheral enhancing rim with central non-enhancing area (Fig. 3). Cortical destruction was noted in the L4 vertebra on left side. Epidural enhancing soft tissue component was noted at the level of L3, L4, and L5

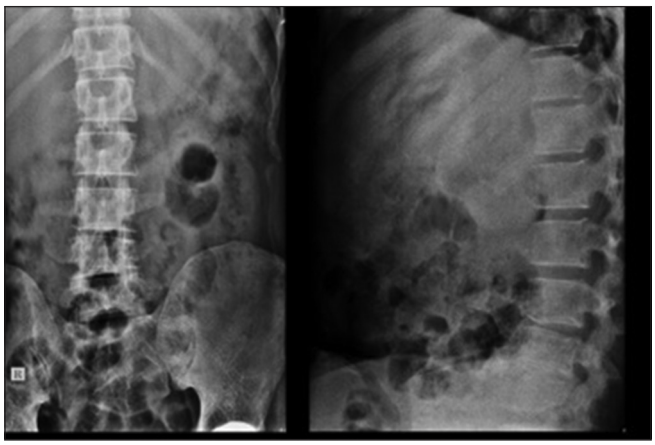

Figure 1: Anterior-posterior and lateral X-ray of the lumbar spine: Osteopenia with end-plate sclerosis of all the lumbar vertebrae, reduction in the height of L4 vertebra, and mild scoliosis

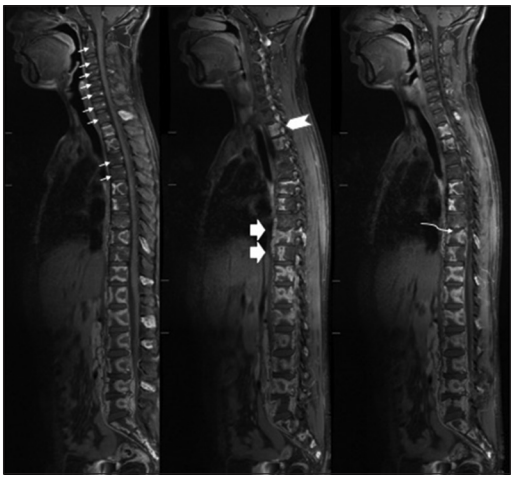

Figure 2: Sag T1C + magnetic resonance images showing bone infarcts (thin arrows), marrow conversion (arrow heads), changes of discitis at T8-9 level (curved arrow), and subligamentous-enhancing soft tissue component (thick arrows)

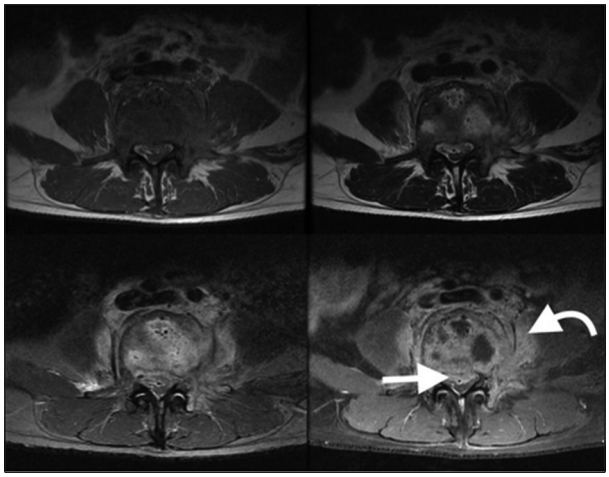

Figure 3: Ax T1-weighted (T1WI), T2WI, short tau inversion recovery and T1C + magnetic resonance images showing paravertebral abscess formation (curved arrow) and epidural enhancing soft tissue component (straight arrow)

vertebral bodies and intervening intervertebral levels appearing hyperintense on STIR and T2WI and hypointense on T1WI (Fig. 3). Reduction in the disc height of the T8-9 intervertebral level was noted with inferior end plate irregularities of T8 vertebrae with the adjacent part of the T8 vertebra showing mild enhancement post-contrast suggestive of discitis (Fig. 2). Subligamentous enhancing soft tissue component was noted, elevating the all extending from the level of mid part of T8 vertebra to superior one-third of T10 vertebra (Fig. 2). The collection was drained surgically and aggressive medical management of painful crisis was established.

DISCUSSION

According to a study by the American society of radiology, diagnoses in the five collected cases included diffuse marrow replacement from anemia (n = 5), epidural abscess (n = 2), osteomyelitis/discitis (n = 3), facet osteomyelitis (n = 1), and bone infarct (n = 3) [5]. Most of these imaging findings were found in a single patient in our study. There are multiple studies which mention all the different manifestations of the SCD in different patients. However, there is paucity of literature which describes all the spinal manifestations of the SCD in a single patient. The purpose of this case is to list almost all the spinal manifestations of SCD in a single patient which is not served by any of the above-mentioned studies.

The varied manifestations of SCD on spinal MR are as follows: Effects of intramedullary hyperplasia anemia results from the rapid removal of abnormal red blood cells by the reticuloendothelial system, which reduces the red cell life span to one-tenth its normal duration [1]. After birth, the red marrow undergoes a gradual conversion into yellow or fatty marrow. Increased red cell destruction and consequent anemia are the main stimuli for the reconversion of yellow to red marrow. The constant stimulation of red cell production leads to the widening of medullary spaces and thinning of cortical bone, which may result in pathologic fractures. Coarsening of the normal trabecular pattern is seen in both long and flat bones. Osteopenia also results from this process and may be visible on radiographs [1]. All these findings were very well visualized in our case.

In the spine, cortical thinning and softening of bone produce a smooth biconcave deformity of the vertebral bodies and adjacent intervertebral discs compress the endplates, giving the vertebrae the characteristic "fish-mouth" appearance [1]. Vertebral collapse with resultant kyphosis is also seen. Although it is more common in other hemolytic anemias and in sickle variants such as Hbs-thalassemia, extramedullary hematopoiesis occasionally is seen in sickle cell anemia [1]. The most common site is the liver, but the spleen also may be affected, and soft-tissue hematopoietic masses may develop in the thorax, adrenal glands, and skin.

Deoxygenation of Hbs-containing red blood cells results in the aggregation of abnormal Hb molecules into long chains [1]. This irreversible process distorts the red blood cell into a rigid sickle shape. The sickling of red blood cells in the bone marrow

causes stasis of blood and sequestration of cells. Ischemia and tissue hypoxia are the consequences and, in turn, worsen the sickling process. The eventual result is cell death leading to infarction. Infarction is a debilitating and significant complication of SCD [1]. Bone infarcts are seen in our case involving almost whole of the spine.

H-shaped vertebral deformity is a result of central growth plate infarction. It can be distinguished from marrow hyperplasia by the characteristic sharp step-like appearance of the vertebral endplate. In addition, compensatory lengthening of the vertebrae adjacent to the H-shaped vertebrae may occur which has been described as "tower" vertebrae [6]. Bone infections are a serious complication of SCD and important causes of hospitalization [1]. The high frequency of infection in patients with SCD is due hyposplenism, which is secondary to infarction in childhood and subsequent fibrosis, phagocytosis, and complement dysfunction [3]. Infarction and necrosis of medullary bone create a good culture medium for bacterial growth and spread. In addition, multiple hospital admissions may increase the patient's exposure to certain bacterial pathogens [7].

Imaging findings of discitis and osteomyelitis are seen in our case. MR findings of discitis/osteomyelitis include [5]: (1) Increased disc signal on STIR and T2WI sequences as well as enhancement on T1WI images following contrast administration, (2) erosion of the adjacent cortical end plate identified on STIR, T1WI and T2WI sequences, (3) low signal on T1WI images, high signal on STIR and T2WI sequences and contrast enhancement of the end plates and vertebral bodies adjacent to the infected disc, (4) adjacent para spinal soft tissue enlargement and enhancement, and (5) facetal arteriopathy.

CONCLUSION

A patient with SCD presenting with all the spinal manifestations of SCD on Spinal MR is rare. Spinal MR is critical for detection and differentiation of the various types of spinal pathologies seen in SCD patients. MR allows the clinician to make an appropriate decision regarding surgical or medical treatment given its ability to identify cord compression caused by epidural abscesses as well as to differentiate discitis/osteomyelitis from bone infarcts.

REFERENCES

1. Ejindu VC, Hine AL, Mashayekhi M, Shorvon PJ, Misra RR. Musculoskeletal manifestations of sickle cell disease. Radiographics. 2007;27(4):1005-21.
2. da Silva Junior GB, Daher Ede F, da Rocha FA. Osteoarticular involvement in sickle cell disease. Rev Bras Hematol Hemoter. 2012;34(2):156-64.
3. Almeida A, Roberts I. Bone involvement in sickle cell disease. Br J Haematol. 2005;129(4):482-90.
4. Roger E, Letts M. Sickle cell disease of the spine in children. Can J Surg. 1999;42(4):289-92.
5. Candocia FJ, Bowen BC, Post JD. Spinal MR in Sickle Cell Patients: Range of Pathology and Diagnostic MR Features, 2006. American Society of Spine Radiology. Available from: http://www.theassr.org/abstract/spinal-mr-in-sickle-cell-patients-range-of-pathology-and-diagnostic-mr-features. [Last accessed on 2017 Jun 14].
6. Marlow TJ, Brunson CY, Jackson S, Schabel SI. 'Tower vertebra': A new observation in sickle cell disease. Skeletal Radiol. 1998;27(4):195-8.
7. Resnick D, editor. Hemoglobinopathies and other anemias. In: Diagnosis of Bone and Joint Disorders. 4th ed. Philadelphia, PA: Saunders; 2002. p. 2146-87.

Funding: None; Conflict of Interest: None Stated.

How to cite this article: Ratnaparkhi C, Chaukhande SM, Dhok A, Barick D. An unusual case of spinal manifestations of sickle cell disease. Indian J Case Reports. 2017;3(3):153-155.