

## Imaging Bone Manifestations of Sickle Cell Disease: Case Report

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DOI: [10.36347/sjmcr.2021.v09i05.017](https://doi.org/10.36347/sjmcr.2021.v09i05.017)

| Received: 03.04.2021 | Accepted: 10.05.2021 | Published: 14.05.2021

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### Abstract

### Case Report

Sickle cell disease (SCD) is the most prevalent hemoglobinopathy. Medical imaging plays a very important role in the diagnosis of complications and sometimes to diagnose the initially misunderstood disease.

**Keywords:** Sickle cell disease, medical imaging, bone manifestations.

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## INTRODUCTION

Sickle cell disease (SCD) is an inherited abnormality of the  $\beta$ -globin chain resulting in a spectrum of haemolytic anaemias [1]. HB S is prevalent in Africa, the Middle East, Mediterranean countries, and India. Clinical manifestations are the results of the polymerization of 1-HB S. under hypoxic conditions the erythrocyte loses its flexibility and consequently the formation of sickle cell. The slightest deformability of sickle cells will cause microvascular occlusion and hemolytic anemia. Complications generally appear at the late stage, especially osteo-articular complications.

Imaging is essential in exploring the complications of sickle cell anemia which are numerous and of variable locations. MRI and CT offer a rich semiology to assess the lesions. The purpose of this case report is to show late-revealed sickle cell anemia in a 22-year-old man with no history of disease.

## CASE REPORT

Young man of 22 years, with no particular pathological history, called to the military service, referred to the emergency services for acute abdominal pain with vomiting. The biological assessment showed, Anemia; Increase in LDH; Increase in predo total bilirubine, unconjugated inesnce, a lipasemia at 1161 IU / L without lithiasis detectable on ultrasound. An abdominal CT scan was performed, which showed stage a pancreatitis with asplenia (because of repeated

infarctions, splenic sequestration). The evolution was marked by the appearance of a paraplégie, which required an emergency spinal MRI. Spinal MRI showed inversion of the spinal cord signal with individualization of multiple signal abnormalities of the vertebral bodies and psotterior arches in hyposignal T1-T2, limited by desquamation in T1 hyposignal, and a double border in T2 sequence. On the bone window, the main anomaly observed is the sign of the H-shaped vertebra or the “Tower” vertebra (Figure 1).

The spontaneous evolution was marked a few days later by a spontaneous regression and resumption of walking with lameness. An MRI of the pelvis is requested.

MRI of the pelvis showed a loss of sphericity of the femoral head with signal abnormality: heterogeneous hypersignal in T1-T2 limited by a line of scaling. Hip joint effusion has also been observed (Figure 2).

Subsequently, an assessment of protein hemolysis and electrophoresis was requested, which showed anemia at 8 and HBS at 46%. The diagnosis of sickle cell disease is retained.

## DISCUSSION

Sickle cell disease (SCD) is the most prevalent hemoglobinopathy. Normal hemoglobin has two alpha and two Beta globin chains. If both the beta globin chains are defective, the disease is called homozygous hemoglobin sickle cell anemia (SCA) (HbSS), these sickled and rigid cells lack the flexibility required to

<sup>1</sup> G. Madani et al., « The Radiological Manifestations of Sickle Cell Disease », *Clinical Radiology* 62, n° 6 (juin 2007): 528- 38, <https://doi.org/10.1016/j.crad.2007.01.006>.

flow in the circulation, leading to vasoocclusion, ischemia, and infarcts.

SCD can affect many organs in the body, but brain injury is one of the most devastating and feared complications leading to mortality and morbidity. The cerebrovascular complications of SCD include stroke, transient ischemic attack, silent cerebral ischemia (SCI), and brain atrophy. Abdominal pain is a common component of sickle cell painful crises and may result from micro-occlusive infarcts of the mesentery and abdominal viscera. Acute pancreatitis has been described as a rare manifestation of vaso-occlusive painful crisis. Its diagnosis is based on the presence of abdominal pain and biochemical evidence of pancreatic injury specifically Serum lipase who has a higher sensitivity and specificity. CT can illustrate changes of acute pancreatitis and enable assessment for complications. In uncomplicated cases, the pancreas may be mildly enlarged, but enhances normally with peripancreatic inflammatory changes and fluid. Complications include necrotizing pancreatitis, in which the pancreatic parenchyma does not enhance peripancreatic fluid collections, pseudocyst formation, aneurysms, and venous thrombosis. Most patients with SCD respond to conservative management, though complications of pancreatitis such as pseudocyst formation have been reported. In this case report we describe patient who has acute pancreatitis without lithiasis in the gallbladder, for that we suggest that pancreatitis was secondary to sickle injury ischemia. Shahid Ahmed and his colleagues make the same observation in four patients with pancreatitis without lithiasis [2]. Pancreatitis should always be considered among the differential diagnoses in patients with sickle cell anemia who have acute abdominal pain.

Osteonecrosis or infarction is one of the consequences of sickle cell anemia in the bone marrow which can cause blood stasis. Blood vessels can become blocked, with development of linear osteosclerotic strands parallel to the cortical border. These osteosclerotic strands are visible on imaging and lead to a "bone in bone" appearance. Initial radiographs are usually normal with an acute infarction [3].

Due to edematous changes in the bone marrow, MRI may show areas of increased signal. Over time, these chronic infarct regions become hypointense on T1 and T2 imaging due to sclerosis and the fibrotic changes accompanying this process. In the T2

sequence, the infarct zone appears in the form of a hypersignal zone as in the case of osteomyelitis, which sometimes makes it difficult to differentiate between them [4].

Long bones are the most common sites of bone infarction. The heads of the humerus and femur are the sites most frequently affected by this avascular necrosis, as they report Jean-Baptiste and De Ceulaer in their case series [5]. Our case presented the same signs in MRI which made it possible to diagnose his unknown pathology at the beginning.

Finally the standard radio may be normal at the start and may be inconclusive. Untreated, 87% of affected femoral heads will collapse within 5 years of diagnosis [6]. MRI remains the gold standard for early diagnosis and prevention of complications of avascular infarction.

Another important manifestation of bone infarction in sickle cell anemia is the H-shaped vertebrae in the spine. The vertebral bodies exhibit depression of the central terminal plate due to chronic infarction.

This deformation can be differentiated from spinal cord hyperplasia by a specific step-like appearance of the end plates of the vertebrae. As a result of this depression of the end plate, the vertebrae adjacent to the H-shaped vertebrae may have an elongation to compensate for the change in shape and to support the spine. This deformation was called "tower" vertebrae [7]. The same radiological signs were observed in our case in the spinal MRI performed at the start of the symptomatology.

<sup>4</sup> Vijaya Kosaraju et al., « Imaging of Musculoskeletal Manifestations in Sickle Cell Disease Patients », *The British Journal of Radiology* 90, n° 1073 (mai 2017): 20160130, <https://doi.org/10.1259/bjr.20160130>.

<sup>5</sup> Georges Jean-Baptiste et Karel De Ceulaer, « Osteoarticular Disorders of Haematological Origin », *Best Practice & Research Clinical Rheumatology* 14, n° 2 (juin 2000): 307- 23, <https://doi.org/10.1053/berh.2000.0067>.

<sup>6</sup> Hernigou, P., Bachir, D. & Galacteros, « The Natural History of Symptomatic Osteonecrosis in Adults with Sickle-Cell Disease », 2003.

<sup>7</sup> Malizos KN, Sifakakos MS, Fotiadis DI, Karachalios TS, Soucacos PN, « An MRI-based semiautomated volumetric quantification of hip osteonecrosis. », *Skeletal Radiol*, 2001.

<sup>2</sup> Shahid Ahmed et al., « Acute Pancreatitis during Sickle Cell Vaso-Occlusive Painful Crisis », *American Journal of Hematology* 73, n° 3 (juillet 2003): 190- 93, <https://doi.org/10.1002/ajh.10344>.

<sup>3</sup> A. Ganguly, W. Boswell, et H. Aniq, « Musculoskeletal Manifestations of Sickle Cell Anaemia: A Pictorial Review », *Anemia* 2011 (2011): 1- 9, <https://doi.org/10.1155/2011/794283>.

In our case, medical imaging made it possible to orient the diagnosis of sickle cell anemia thanks to the different radiological signs observed on spinal and pelvic MRI. The blood protein electrophoresis confirmed the diagnosis later.

## CONCLUSION

Sickle cell anemia is a common hereditary disease in our region; its diagnosis is sometimes late. Medical imaging plays a very important role in the diagnosis of complications and sometimes to diagnose the initially misunderstood disease which will be confirmed by protein electrophoresis.

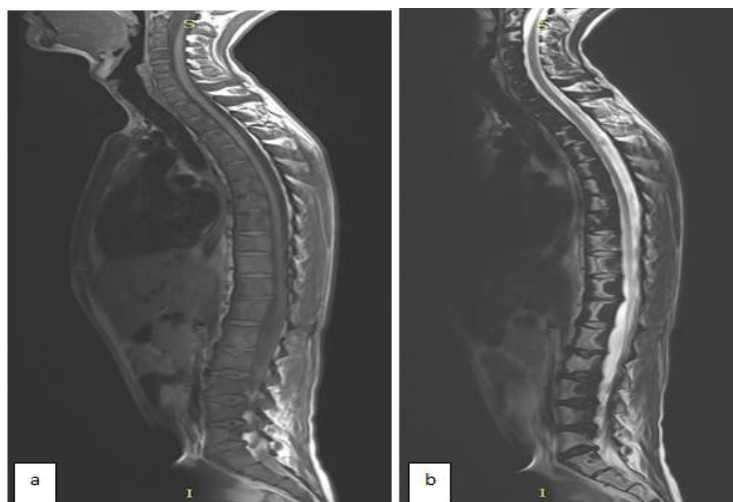


Fig-1: MRI of the entire bone spine in sagittal section in T1 (a) and T2 (b) showing H-shaped vertebrae or "tower" vertebrae

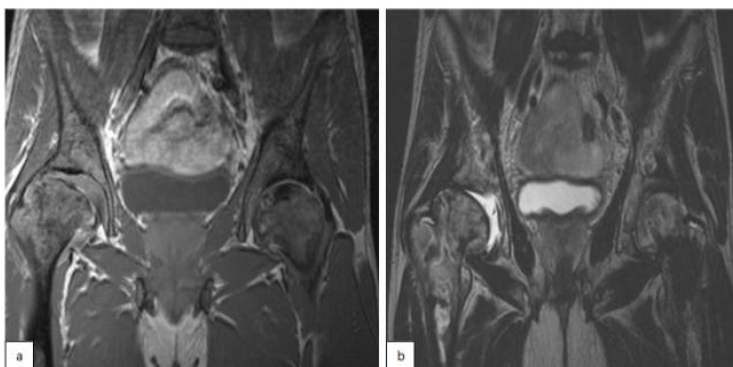


Fig-2: MRI of the pelvis in coronal section showed a loss of sphericity of the femoral head with signal abnormality: heterogeneous hypersignal in T1 (a)-T2(b) limited by a line of sclerotic in favor of osteonecrosis. Hip joint effusion has also been observed

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