Osteo-articular manifestations in 97 children with sickle cell disease - imaging value

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Authors: P. L. Pegado¹, S. Serpa², R. Perry¹, C. A. S. Ruano¹, A. Nunes¹, A. P. Petinga¹, P. Alves¹, E. Soares¹; ¹Lisbon/PT, ²São Miguel/PT
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Learning objectives

To illustrate the imaging of musculoskeletal manifestations of sickle cell disease (SCD) in 97 children followed in our center.

To demonstrate usefulness of imaging in the diagnosis of the various musculoskeletal manifestations in SCD.

Background

Sickle cell disease disease is one of a group of genetic disorders known as hemoglobinopathies, and is characterized by the inheritance of sickle haemoglobin (HbS). A mutation in the beta-globin gene on 11th chromosome, resulting in replacement of glutamic acid in position 6 of the beta-globin chain by valine, resulting in an abnormal haemoglobin HbS molecule. The term sickle cell disease applies to those patients who have at least one abnormal HbS chain and another abnormal beta chain. If the second abnormal beta chain is also an HbS chain then the patient is considered to be homozygous HbSS. Alternatively, other abnormal haemoglobin chains like HbC or thalassemia result in HbSC and HbS-thal, respectively.

Sickle cell disease is the most common blood disease, with worldwide distribution (1).

Deoxygenation of Hb S results in the aggregation of abnormal haemoglobin molecules into long chains. In the beginning this is a reversible process that turns into an irreversible process that tends to distort the red blood cell membrane into a rigid sickle shape (2) (Fig. 1 on page 4.).
Fig. 1: SCD physiopathology - The abnormal HbS protein chain polymerizes reversibly in deoxygenated environment into a gelatinous network of fibrous polymers that stiffen the RBC (red blood cell) membrane, increases the viscosity, and causes dehydration resulting in a sickle shape. These abnormal cells are abnormally sticky provoking microvascular occlusions and premature haemolysis.

References: Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT

The consequences are obstruction of the microcirculation, ischemia, and infarction. 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. Thus, disease manifestations can be roughly attributed to two phenomena: haemolysis and vaso-occlusion. Haemolytic anaemia occurs in all forms of SCD and results in destruction of the sickled cells, from cell dehydration and direct membrane damage by rigid haemoglobin polymers (2). Vaso-occlusion is due to entrapment of sickled cells in the microcirculation and leads to tissue ischaemia and damage in almost all organs (2). Several organs can be affected with alterations in the central nervous system, bone and joints, cardiovascular system, respiratory system, gastrointestinal tract, and kidneys which increases morbidity and mortality in this group of patients (1).
Bones are the second most affected organs by SCD, after the spleen (3). Despite the importance of osteoarticular involvement in SCD, these complications are still little studied and the physiopathology of alterations is not fully understood.

Nevertheless, the most frequent complications that require hospitalization in SCD patients are painful vaso-occlusive crises and osteomyelitis (4).

This poster discusses the manifestations related to musculoskeletal involvement in SCD, not only in the acute setting, but also the long term complications, with particular reference to differentiating infection from infarction, where imaging is playing a crescent important role on acute bone pain algorithm management.

Images for this section:

**Fig. 1:** SCD physiopathology - The abnormal HbS protein chain polymerizes reversibly in deoxygenated environment into a gelatinous network of fibrous polymers that stiffen the RBC (red blood cell) membrane, increases the viscosity, and causes dehydration resulting in a sickle shape. These abnormal cells are abnormally sticky provoking microvascular occlusions and premature haemolysis.
This retrospective study reports the osteo-articular manifestations associated with sickle-cell disease encountered in our institution, in children with at least one hematologic appointment since 2013 till the end of 2014. Osteo-articular manifestations with imaging findings were reported in 28 patients out of 97. There were 14 females and 14 males. There were 23 homozygous (SS) patients and 5 composite heterozygous (SC, Sb) out of 28 patients with osteo-articular manifestations. These cases were retrospectively studied and we depicted 11 major manifestations like infections and avascular necrosis. The remaining 17 cases were reported as minor manifestations such as intramedullary marrow replacement and hyperplasia and also growth effects.

Marrow replacement and hyperplasia

Increased destruction of red blood cells and consequent anemia causes persistence and expansion of the red (haematopoietic) marrow. Marrow hyperplasia results in widening of the medulla and subsequent cortical thickening, resulting in coarsening of the normal trabecular pattern with loss of corticomedullary differentiation (Fig. 2 on page 24).
Fig. 2: Effects of medullary hyperplasia. Posteroanterior chest radiograph, of a 14 year old girl, homozygous HbSS, showing coarsening of trabecular pattern with loss of corticoedullary differentiation particularly on the ribs.

References: Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT

This process may cause the bone to appear osteopaenic and make the bone prone to pathologic fractures (1).

Cranial diploe proeminence is observed in the skull (Fig. 3 on page 25), as well as flattening of the external and internal laminae and increased thickness of the frontal and parietal bones. The best example of bone softening is seen in the vertebral bodies, where
the end plates assume a smooth concavity described as fish mouth vertebra (5) (Fig. 4 on page 25). On T1 weighted image, normal fatty marrow shows high signal intensity, while haematopoietic red marrow is low in signal (Fig. 5 on page 26).

Fig. 3: Bone marrow hyperplasia within the skull vault. A-Posteroanterior radiograph from a 10 year old girl, homozygous sickle cell disease (Hb SS), there is widening of the diploic space (arrows). B- T1W Sagittal MR image of the same patient, the medullary cavity is widened with low signal intensity.

References: Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT
**Fig. 4:** Thoracic Computed Tomography (CT), reformatted on a sagittal plane in bone window of a 18 year old girl HbSS. Smooth concavity of the vertebral endplates at multiple vertebral body levels resulted from bone softening (fish mouth vertebra). Note patchy sclerosis of the vertebrae and sternum, caused by medullary infarction.

**References:** Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT
**Fig. 5:** T1W MRI sagittal view of the spine in a 12 year old boy HbSS, shows low signal intensity indicative of cellular (red) marrow hyperplasia.

**References:** Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT
Extramedullary hematopoiesis is also described in SCD, however we didn´t find it in our series. 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 in the skin.

**Bone Infarcts**

Abnormal RBC shape causes stasis of blood and sequestration of cells in bone capillaries resulting in ischemia and tissue hypoxia which worsens the sickling process. Bone infarction can take place in both the diaphysis, causing medullary infarcts and in the epiphyses, causing avascular necrosis. This can present as the classical painful bone crisis, although they also may be clinically silent and discovered incidentally at radiography (6).

Initial radiographs are usually normal with an acute infarction (7). Chronic infarcts demonstrate intramedullary lucency and patchy sclerosis with or without periosteal reaction, depending if cortical bone is also affected. MRI is more sensitive than CT and plain radiographs in the detection of bone infarcts (7). On T2 weighted MRI, infarction is seen as an area of high signal intensity. Infarcts may also show peripheral enhancement post gadolinium and soft tissue involvement, complicating the differential diagnosis with osteomyelitis in the acute setting (8) (Fig. 6 on page 27 Fig. 7 on page 28 Fig. 8 on page 30).

**Fig. 6**: A- T1W FS MRI, sagittal view of a 12 year old boy heterozygous HbS# with vertebral infarction at different levels, on L3 L4 and L5, showing abnormal heterogeneous signal intensity, with collapse of the vertebral bodies. B- CT Axial view
on bone window of the same patient showing patchy sclerosis of the vertebral body consistent with chronic infarction

References: Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT
**Fig. 7:** T2W FS MRI, lumbar sagittal view of a 10 year old boy homozygous HbSS, showing high signal intensity at L2, L3 and L4 vertebral bodies consistent with osseous edema in the setting of acute bone pain consistent with acute infarction.  
**References:** Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT

**Fig. 8:** A- DP FS MRI of the right knee on sagittal view of a 13 year old boy, homozygous HbSS, with acute right knee pain, where we can see a serpiginous double line at the distal femoral diaphysis and proximal tibia diaphysis, not affecting the articular surface, consistent with medullary infarcts. B - Knee plain radiograph on lateral projection of the same patient, done at the same time of the MRI, no radiographic alterations were depicted.  
**References:** Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT

In young children, infarction occurs within the small bones of the hand and feet resulting in painful dactylitis termed "hand-foot" syndrome (7). Sickle cell dactylitis is common between the ages of 6 months and 2 years but is rare after the age of 6 years because of the regression of red marrow in these areas with increasing ages (9) (Fig. 9 on page 30). In our series we only depicted 2 cases of dactylitis, all of them corresponding to a homozygous HbSS children.
Fig. 9: A - Left hand plain radiograph on AP projection of a 5 month boy homozygous HbSS, with periosteal new bone formation along the diaphysis of the first and third metacarpal (arrow) and also lucencies (circles) in keeping with early destructive lesions consistent with dactylitis. B- T1 FS MRI, where we can see hyperintensity at the base of the third metacarpal (circle) with soft tissue involvement consistent with sequestered blood in the marrow space in the subacute phase of an infarction.

References: Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT

Another pattern of bone infarction, as it was referred before, is the epiphysis involvement. This process is more usually known as avascular necrosis (10). Again, initial radiographs are usually normal. As osteonecrosis progresses, changes become evident at radiography. Early radiographic signs include lucency and sclerosis within the epiphysis; subsequently, crescent-shaped subchondral lucencies develop, and later a depression of the articular surface with collapse and eventually fragmentation may occur (Fig. 10 on page 31 Fig. 11 on page 32). The earliest signs of avascular necrosis are seen on MRI. Particularly T2 weighted images with fat supressed sequences showing
regions of high signal intensity indicative of bone marrow edema. A serpiginous line is classically seen, that consists of a hyperintense inner border and hypointense periphery. This double line results from the high signal intensity inflammatory response of bone with granulation tissue, inside the low signal intensity reactive bone interface (11, 12) (Fig. 12 on page 32).

**Fig. 10**: Plain radiographs on antero-posterior (AP) projection of a nine year old male HbS homozygous HbSS with alpha thalassemia too, showing left humeral head AVN progress within 4 months. In A there are signs of epiphysis lucencies. In B there are subcondral lucencies with depression of articular surface. In C there is collapse of the articular surface with flattening of the femoral head.

**References**: Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT
Fig. 11: Plain radiographs on AP projection of a 10 year old boy homozygous HbSS with alfa thalassemia, presented with left femoral head osteonecrosis with advanced degenerative changes due to the collapse of the femoral head. B- A Valgisation osteotomy was performed.

References: Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT
Fig. 12: Twelve year old boy homozygous HbSS, presented with acute pain on the left hip. A- T2W FS MRI, coronal view, high signal intensity was depicted on the left femoral head consistent with osseus edema. B- T1 W FS post gadolinium, coronal view, no enhancement on post contrast was depicted on the left femoral head in keeping with infarction. C- Plain radiograph on AP projection, several months later where we can find flattening of the femoral head. D- Total hip arthroplasty was performed

References: Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT

Seven cases of avascular necrosis were encountered in our series. The sex ratio was 5 female : 2 males. Six cases were on the femoral head and one case at the humeral head. Four of the ones with femoral head necrosis needed surgery.

Osteomyelitis
Osteomyelitis is a serious complication of sickle cell disease and usually occurs in long bones e.g. femur, tibia, and humerus. The high frequency of infection in patients with SCD is due to a number of factors. Hyposplenism due to autosplenectomy results in a degree of immunocompromise (5). Infarcted or necrotic bone is a good environment for bacterial growth (3). In addition, multiple hospital admissions may increase the patient's exposure to certain bacterial pathogens (5) (Fig. 13 on page 33). The most common infectious agents of osteomyelitis in patients with SCD are *Salmonella* organisms or *Staphylococcus aureus* (1).

![Fig. 13: Factors that may contribute to high frequency of infection in SCD](image)

**References:** Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT

Osteomyelitis occurs in 18% while septic arthritis occurs in 7% of patients with SCD according to a study (13).

Six cases of infection were observed in our series, five cases of osteomyelitis and 1 cases with arthritis. Only in one case a microorganism was cultured and it was a *Staphylococcus sp*. No bacteria were cultured in the other 5 cases. One case was reported as a multifocal osteomyelitis.

The classical clinical findings of pain, fever, and raised inflammatory markers can also be seen in infarction, which can cause diagnostic difficulty in the acute setting (14).
Plain film findings of osteomyelitis include osteopaenia, periosteal reaction with or without associated cortical destruction, sinus tract formation, and soft tissue extension. There are features such as osteopaenia and periosteal reaction that are not specific of osteomyelitis and can also be seen in acute bone infarction. But the main question with plain films is that the earliest changes may not be evident for 8-10 days from the beginning of the clinical complains (15).

Sonography has been reported to have high sensitivity in diagnosing osteomyelitis in children with SCD. In a recent study, it was reported that sub-periosteal fluid collection with diameter superior of 4 mm is a strong indicator of osteomyelitis (16).

MR imaging is an increasingly useful tool for the diagnosis of osteomyelitis in SCD. On T1 weighted sequences osteomyelitis is of low signal intensity, care must be taken because areas of red marrow will also be of low signal intensity. On T2 weighted fat saturated sequences may show fluid collections as an area of high signal intensity within the bone marrow. These sequences also are useful for demonstrating the communication of soft-tissue fluid collections with medullary fluid collections through cortical defects. Osteomyelitis will also show areas of enhancement postgadolinium. This will tend to be more diffuse than in infarction. It has been proposed that the degree of the thickness of rim enhancement differs between osteomyelitis and acute infarction (8). Thus in osteomyelitis, there is a thick irregular rind of enhancement whereas acute infarction results in thin linear rim enhancement (8) (Fig. 14 on page 34 Fig. 15 on page 36 Fig. 16 on page 36 Fig. 17 on page 37).
Fig. 14: Osteomyelitis of the left femur in an 11 year old female, homozygous HbSS, no agent was identified. A - Sonography at the left tight, there is a subperiosteal liquid collection of the femur with a depth superior than 4 mm. There is also some irregularity of the cortical, and a thickening of subcutaneous tissue. B- Plain radiograph on AP projection showing subperiosteal reaction. C- CT - Sagital reconstruction on bone window where we can depict the subperosteal collection and the thickening of the cortical bone. D- Coronal T2W FS MRI, obtained at a follow-up examination shows
medullary high signal intensity as well as a periosteal collection that were later found to be an abscess.

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Fig. 15: Osteomyelitis of the left humerus in a 1 year old boy heterozygous HbSC. No agent was isolated. A- Plain radiograph of the left arm, no pathological features were depicted. B-Sonography on a tranversal view, where we can see a liquid periosteal collection on the left humerus with 7 mm diameter deph. C- T2-weighted fat supressed MR image, shows high signal intensity on the humerus diaphysis in keeping with osseous edema, note also the high signal intensity of the soft tissues around. D- T1 weighted fat supressed MR image post gadolinium showing geographic enhancement at the proximal aspect of humerus diaphysis and also in the distal portion involving the distal epiphysis, consistent with an osteomyelitis enhancement pattern.

References: Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT
Fig. 16: Osteomyelitis of the left radius in a two year old girl, homozygous HbSS with alfa Thalassemia, no agent was isolated. A- Sonography, there is a liquid subperisosteal collection, with cortical irregularity. B- Plain radiography on AP projection, showing multifocal bone destruction in the radio characterized by medullary hyperlucencies with cortical irregularities. C- T2 weighted fat supressed MR image, shows high signal intensity within the radio medullary cavity and a liquid colections of the soft tissues around. D- T1 weighted fat supressed MR image post gadolinium shows a thick and irregular enhancement on the medullary cavity.

References: Radiologia, Hospital de São José, Centro Hospitalar Lisboa Central - Lisbon/PT

Although there is no reference standard for diagnosing sickle cell osteomyelitis, and even the culture of biopsy is not completely reliable (15). The lack of a reference standard makes a comparison of imaging modalities difficult. In this way, the interpretation of radiologic findings must be part of a careful ongoing multidisciplinary assessment. However it must be referred that acute bone infarction is about 50 times more frequent that acute osteomyelitis (4).

Recently it was proposed that the acute bone infarcts in sickle cell disease are caused by sequestration of red blood cells in the bone marrow (17). Furthermore it was also proposed that T1 weighted fat saturated sequence alone is diagnostic for acute bone infarcts, when a high signal intensity is depicted and in the acute bone pain setting. The authors also propose that contrast enhancement sequences should be added if no high signal intensity is depicted on T1 weighted fat saturated in order to diagnose or exclude osteomyelitis (Fig. 18 on page 38).
Fig. 18: Management algorithm of acute bone pain in SCD (8,17) - In strong equivocal cases of acute bone pain a MRI should be performed. High signal intensity on T2 and T1 FS sequences, acute bone infarction should be diagnose. In the setting of high signal intensity on T2 and a low or isointense signal on T1 FS, contrast enhancement should be performed, to confirm or exclude the diagnosis of osteomyelitis.

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Septic arthritis is less common than osteomyelitis. It often arises in conjunction with vaso-occlusion and bone infarction. Both US and MRI are useful and may include features like articular effusion and peri-synovial edema.

Soft tissue abnormalities

Occlusion of vessels leads to inflammation, edema, and myonecrosis. Fluid collections, hematomas, and fat necrosis may occur in soft tissue. No cases of soft tissue abnormalities alone were encountered in our series.

US and MRI have good sensitivity for identifying soft-tissue changes. On MRI, high signal intensity within muscle on T2 weighted sequences is the main feature of soft tissue involvement.
Growth effects

Patients with SCD have reduced height. This is believed to be due to bone marrow hyperplasia. Bones are generally shorter due to epiphyseal shortening subject to ischemia /infarction and vascular compromise to the growth late. Premature closure of growth plates also occurs.

Images for this section:

Fig. 2: Effects of medullary hyperplasia. Posteroanterior chest radiograph, of a 14 year old girl, homozygous HbSS, showing coarsening of trabecular pattern with loss of corticoedullary differentiation particularly on the ribs.
Fig. 3: Bone marrow hyperplasia within the skull vault. A-Posteroanterior radiograph from a 10 year old girl, homozygous sickle cell disease (Hb SS), there is widening of the diploic space (arrows). B- T1W Sagittal MR image of the same patient, the medullary cavity is widened with low signal intensity.
Fig. 4: Thoracic Computed Tomography (CT), reformated on a sagittal plane in bone window of a 18 year old girl HbSS. Smooth concavity of the vertebral endplates at multiple vertebral body levels resulted from bone softening (fish mouth vertebra). Note patchy sclerosis of the vertebrae and sternum, caused by medullary infarction.
Fig. 5: T1W MRI sagittal view of the spine in a 12 year old boy HbSS, shows low signal intensity indicative of cellular (red) marrow hyperplasia.
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**Fig. 17:** Osteomyelitis of the posterior L3 arch in a five year old boy, homozygous HbSS. T2 weighted MR image sagittal View (A) and axial view (B), showing high signal intensity on the posterior L3 arch (circle), there is also an epidural collection (arrow).
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Conclusion

Familiarity with the imaging features of SCD is important for the diagnosis and management of musculoskeletal complications.

Acute osteomyelitis may be particularly difficult to distinguish from bone infarction, and MRI could be an important tool in those cases.

References


Personal Information

Pedro Luís Figueiredo Pegado
MD, Radiology Lecturer at New University of Lisbon, Faculty of Medicine.
Department of Radiology, Centro Hospitalar Lisboa Central - Hospital de São José, Rua Antonio Serrano 1000-199, Lisboa, Portugal
pedropegado@hotmail.com

Sara Serpa
MD
Department of Radiology, Hospital do Divino Espírito Santo - Av. D. Manuel I, 9500-370 Ponta Delgada, Portugal

Rosário Perry
MD
Department of Pediatrics, Centro Hospitalar Lisboa Central - Hospital Dona Estefânia, R. Jacinta Marto, 1169-045 Lisboa

Carina Ruano
MD, Radiology Lecturer at New University of Lisbon, Faculty of Medicine.
Department of Radiology, Centro Hospitalar Lisboa Central - Hospital Stº António Capuchos, Alameda de Santo António dos Capuchos, 1169-050 Lisboa

Ana Nunes
MD
Department of Radiology, Centro Hospitalar Lisboa Central - Hospital Dona Estefânia, R. Jacinta Marto, 1169-045 Lisboa

Ana Paula Petinga
Pedro Alves

MD, Radiology Lecturer at New University of Lisbon, Faculty of Medicine.

Department of Radiology, Centro Hospitalar Lisboa Central - Hospital de São José, Rua Antonio Serrano 1000-199, Lisboa, Portugal

Eugénia Soares

MD

Department of Radiology, Centro Hospitalar Lisboa Central - Hospital Dona Estefânia, R. Jacinta Marto, 1169-045 Lisboa