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Imaging of Bone Involvement of Sickle Cell Disease in Children: About a Case and Literature Review

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Abstract

Sickle cell disease is a genetic disease transmitted in an autosomal recessive way. Acute bone pain crises frequently occur in children with sickle cell disease and may be related to bone infarction or osteomyelitis.

Acute bone infarction can simulate osteomyelitis clinically and biologically as well as in imaging. However, the distinction is essential because the diagnosis of osteomyelitis requires long-term emergency antibiotic therapy, while bone infarction justifies treatment of few days. MRI with Gadolinium injection remains the key examination for making the diagnosis.

Through this work, we report the case of a 12-year-old child with sickle cell disease who presents for bone pain not relieved by paracetamol palliative II by Identifying the semiological elements MRI allowing to differentiate bone infarction of osteomyelitis, faced with acute febrile bone pain in a child with sickle cell disease.

Keywords: Sickle cell disease; MRI; Osteomyelitis; Vaso occlusive crisis

Introduction

Sickle cell disease (SCD) is an autosomal recessive hereditary disease linked to the production of Hemoglobin S (HbS) by mutation of the 6th amino acid of the β chain of globin and changes in the physicochemical properties of globin.

It is the most common genetic disease in the world with 275,000 newborns born every year (1). SCD affects 1 in every 2000 births in England and it is estimated that there are about 12–15,000 SCD patients in the UK [1,2]. However this pales in significance when compared to Sub-Saharan Africa where over 75% of the world's patients live.

3 main categories of clinical manifestations can coexist: chronic hemolytic anemia with acute episodes, vaso-occlusive phenomena and susceptibility to infections giving rise to the three clinico-radiological manifestations of damage to the skeletal system: medullary hyperplasia, vaso-occlusive crises (bone infarction and aseptic osteonecrosis of the femoral head) as well as osteomyelitis.

MRI with injection is the gold standard for diagnosis with a sensitivity and specificity varying respectively from 88% to 100% for osteomyelitis [2], although some studies have shown that MRI cannot differentiate acute osteomyelitis (AOM) from Vaso-occlusive crisis (VOC). On the other hand, for others, this differentiation may be possible [3]. Both conditions are associated with a rise in inflammation such as C-reactive protein (CRP) and white cell count (WCC). Although VOC bone infarction is reported to be up to 50 times more common than osteomyelitis in patients with SCD [4].

In this article, we review the epidemiological, clinical, and pathophysiological features of bone involvement in sickle cell disease and chronic bacterial osteomyelitis. We also describe the pearls and pitfalls of imaging and discuss differential diagnoses.

Observation

We report the case of a 12 year old child, 2nd of a sibling of 3, from a first degree consanguineous marriage, with no significant pathological history, followed 4 years ago for chronic anemia under martial treatment then the diagnosis of sickle cell disease was retained by hemoglobin electrophoresis showing an Hb F:15%, Hb S:82% and Hb A2:3%. Hospitalized in pediatrics for the management of bone pain initially in the spine then in the pelvis not relieved by palliative paracetamol II taken for two weeks. The symptomatology was aggravated by the appearance of a 38.5° low grade fever resolved after administration antibiotic therapy made of injectable flucloxacillin 150mg/kg/day for 4 days then relay orally, patient had benefited from a blood culture which came back negative and in front of the suspicion of osteomyelitis (OM), patient underwent an MRI of the pelvis looking for bone involvement of infacrtus or OM, showing multiple bilateral femoral epiphyseal metaphyseal areas, iliac wings and sacrum with infiltration of the iliac psoas muscles and the left gluteus minimus without clearly visible collection related to bone infarctions.

Discussion

Sickle Cell Disease (SCD) is a clinically significant haemoglobinopathy with increasing incidence in developed countries. SCD affects 1 in every 2000 births in England and it is estimated that there are about 12–15,000 SCD patients in the UK [1]. However this pales in significance when compared to Sub-Saharan Africa where over 75% of the world's patients live. is the most common hereditary hemoglobinopathy, which results in abnormally shaped and rigid red blood cells. These sickle-shaped red blood cells cause vaso-occlusion and ischemic phenomena that can affect any organ in the body.

The involvement of the skeletal system in children with sickle cell disease can be summarized in three main areas: spinal cord hyperplasia, vaso-occlusive crises and osteomyelitis. Vaso-occlusive events are the

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most common acute clinical presentation of SCD in children [5]. A combination of tissue infarction, immunodeficiency due to splenic dysfunction and excess iron leads to increase risk of osteomyelitis in SCD.

Imaging and mainly MRI with gadolinium injection allows a diagnostic approach showing In children without sickle cell disease [6] : At birth; a red marrow throughout the skeleton, which converts to yellow marrow in the peripheral skeleton, completed around 5 years in the diaphyses and epiphyses of the long bones with a complete conversion around 20-25 years. The red marrow is hematopoietic, active-in hyposignal T1, intermediate T2 signal and the yellow marrow is adipose, not active, in Hypersignal T1 [Figure 1].

In children with sickle cell disease, the persistence of red marrow due to a lack of fatty conversion due to chronic anemia and constant stimulation of the red marrow is responsible for marrow hyperplasia manifested by an enlargement of the marrow cavity, a bone thinning, cortical thinning and gross thickening of the residual trabeculation.

Vaso-occlusive crises are secondary to tissue ischemia by slowing of the flow and microvascular occlusion leading to a foot-hand syndrome from 6 months to 2 years by pain and inflammatory swelling of the metacarpals and Metatarsals, to a metaphyseal-diaphyseal infarction from 3 to 10 years, and to an epiphyseal infarction beyond 10 years. Imaging in the initial phase are not indicated, 1-2 weeks later, a periosteal reaction, cortical thinning and speckled osteolysis of the metacarpals, metatarsals and phalanges appear then the evolution is done towards the bilaterality of the lesions with disappearances of the osseous contours, then with a massive infarction with extensive necrosis [7].

The bone infarction reaches the spongy bone of the long bones, in a bilateral and sometimes symmetrical way, the standard radiography shows delayed lesions compared to the clinic with speckled osteolysis, osteocondensation or Mixed, a dense border or radio- transparent on the long bones and signs of cortical infarction, in particular anomaly of transparency and periosteal reaction [8].

MRI remains the most sensitive examination for the positive diagnosis showing signs of infarction in Hyposignal T1, hypersignal T2 initially with a hypo-intense peripheral border of secondary appearance enhanced in the periphery in a heterogeneous way with possible abnormalities of periosteal signal and soft parts, a secondary fibrosis appears in hypointense on all the sequences [9] result compatible with our case [Table 1].

Osteomyelitis is a contamination mainly by hematogenous route in the event of digestive ischemia most frequently by BGN (gram-negative bacillus), salmonella, , as shown by a recent study by Carol M.Cao [10] on a sample of 3553 cases, 19 of which were retained on microbiological evidence. Staphylococcus Aureus is the most common organism to cause osteomyelitis irrespective of a diagnosis of sickle cell disease with preferential diaphyseal involvement of the long bones : Femur, tibia, humerus and involvement tubular bones of the extremities specific to children most often under 5 years of age with a higher incidence in boys than in girls, which could be linked to greater exposure to trauma [11-12]. Hematogenous or non-hematogenous osteomyelitis can be classified as acute or chronic. Hematogenous spread is the most common form of osteomyelitis in children with an estimated incidence of 1 case per 5,000 children per year in the United States [11].

In clinical practice, chronic osteomyelitis is defined by signs and

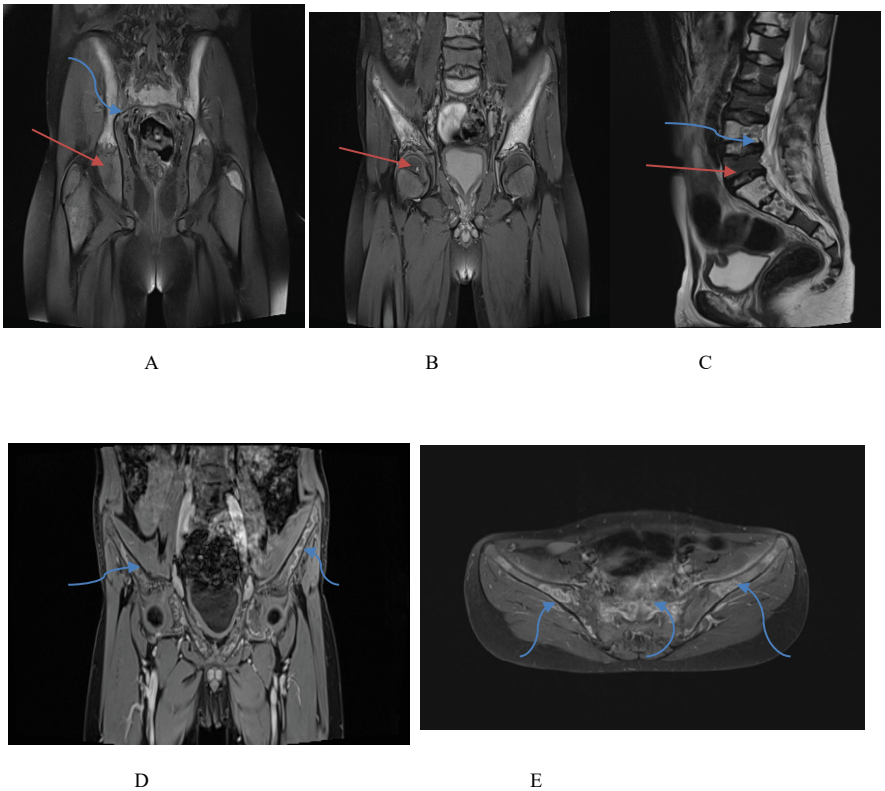

Figure 1: MRI in T1 coronal (A), STIR (B), T2 sagittal (C) and T1 gado (D,E) sequences showing metaphyseal bone signal abnormalities in T1 hyposignal related to medullary hyperplasia with multiple infarct areas bone involving the iliac wings, the sacrum, the apophyseal nuclei and the lumbosacral spine in T1 iso-signal, STIR hypersignal with peripheral enhancement.

Table 1: Bone infarction Vs osteomyelitis on MRI. Cotten, Musculoskeletal imaging, General pathologies 2013.

	Bone infacrtion	osteomyelitis
multifocal and synchronous bone involvement	++	rare
Contrast enhacement	Peripheral enhancement	Extended enhacement
Spontaneous T1 hyperintense	+++	No
Intraosseous collection	NO	+
Periosteal abnormalities	Stratified detachement possible without collection	Subperiosteal abscess
Soft tissue damage	+	++ (collections)

symptoms of bone inflammation for at least two weeks with radiological evidence of devitalized bone [12]. On imaging, features of chronic osteomyelitis are osteosclerosis and sequestrum (dead bone) with inflammatory damage to the soft tissues and adjacent sinus tract [13]. osteomyelitis, to document the extent of the disease and to guide any intervention such as an image-guided biopsy or surgical debridement [14].

The MRI reveals one or more intramedullary Collections in Hyposignal T1, hypersignal T2, peripheral contrast enhancement realizing the target aspect with a cortical defect communicating with a collection of soft tissues [15]. However, in imaging the semiology is also close to the early phase: poorly limited osteolysis, periosteal reaction .

The diagnosis of osteomyelitis vs infarction remains a clinical and imaging challenge ranging from diagnosis to long-term management. Currently, the interest of T1 FS IV- sequences makes it possible to decide the diagnosis by showing a hypersignal of compacted red blood cells compared to the marrow with visualization of the signal of red blood cells sequestered during an infarction [16].

Thus, the discovery of an intra-osseous micro-abscess, the abscess of the soft parts and the periostitis circumferential are in favor of osteomyelitis [17].

Synchronous involvement of several bones, heterogeneous contrast uptake and discreet involvement of the soft tissues of the inflammatory type would be MRI signs that would rather orient towards the diagnosis of bone infarction [16].

Blood cultures, radiographic imaging, surgical incision, and drainage of areas of suspected OM are all important diagnostic considerations in distinguishing osteomyelitis from the more common diagnosis of Vaso-Occlusive Crisis in children with sickle cell disease. Obtaining blood cultures before antibiotic therapy and operative cultures when possible are potential steps to increase the likelihood of identifying a definitive diagnosis of osteomyelitis and thus to guide treatment strategy [18,19].

Conclusion

Bone vaso-occlusive crises and osteomyelitis, sources of intense pain, are the signature of sickle cell disease, on the basis of our case, the clinical signs, the negative culture and the absence of bone abscesses or soft tissues allow us to exclude the diagnosis of osteomyelitis. Knowledge of imaging methods, in particular MRI, is essential to detect the occurrence of potentially severe complications and to guide management.

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