Tardive hip disease diagnosis in a young adult with sickle cell disease

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Abstract

Background: Hip disease is a complication of Sickle Cell Disease most commonly occurs during adolescence and early adult life that constitutes from 3.3% to 26.7% % of cases in SCD patients. Damage to mature epiphysis may cause persistent symptoms requiring surgery which may need revision if it becomes loose with wear.

Case presentation: We report a 27-year-old male who was diagnosed with hip disease following investigations for persistent pain in right hip associated with limp and painful limitation of movement. Avascular necrosis of the right femoral head was identified on a hip X-ray.

Conclusion: Further investigation will be necessary to identify risk factors in SCD Congolese patient and each SCD patient must be screened for predisposing factors.

Keywords: Hip disease- diagnosis-sickle cell anemia- young adult-Kinshasa

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Introduction

Necrosis of the femoral head represents a special form of aseptic bone necrosis because it develops in the high bearing region of the hip joint. This complication is a common cause of morbidity in sickle cell disease and increases with age. The incidence and prevalence is unknown and epidemiological data are rare in Africa[1]. Despite the high prevalence of sickle cell disease in Democratic Republic of Congo[2], no comprehensive care management for bone and joint complication exists to screen sickle cell patients for early detection and monitor this complication. In this context diagnosis was generally delayed and management in limited resource-settings was accordingly difficult[3]. We report a case of hip disease in a young Congolese adult who presented late with advanced disease.

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Case presentation

A 27-year –old black boy with sickle cell disease was referred to Military Hospital of Kinshasa, in Kinshasa, Democratic Republic of Congo. He has had multiple hospitalizations for sickle cell crises. The patient admitted to feeling increasingly persistent pain in that right hip radiating to the right knee for over 6 months associated with limp, worse on walking, limitation of movement and relieved by lying down. On examination in Military Hospital of Kinshasa the patient was a fully developed Congolese male, 1.78 m in height and 55 kg for weight. The right was shorter than the left with a difference of 50 mm. X-ray revealed (figure 1) changes to the articular surface of the right femoral head. The left femoral head were normal. A complete blood count showed Hemoglobin 7.9 g/dl, leukocytes 9.10^9/ L, platelets 280.10^9/L, creatinine 80 µmol/L, urea 4.8 mmol/ L, uric acid 2.65µmol/L, cholesterol 4.1 mmol/L, and HDL 1.2 mmol/L. A diagnosis of hip disease was established. The persistent symptoms due to delayed diagnosis limit quality of life, perform total hip replacement was performed.

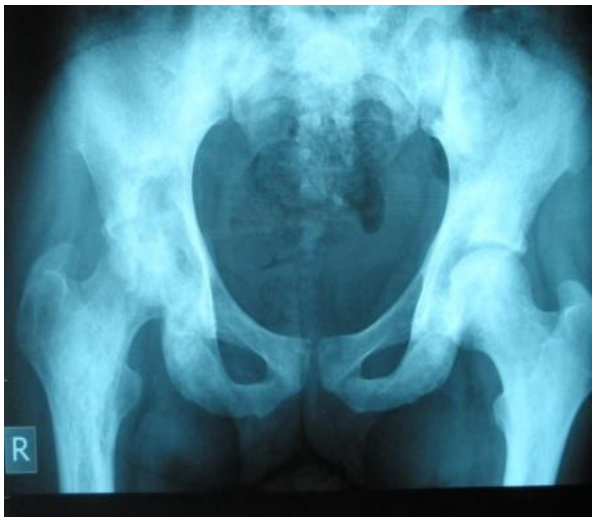

Figure 1: Diffuse necrosis and destruction of the right femoral head

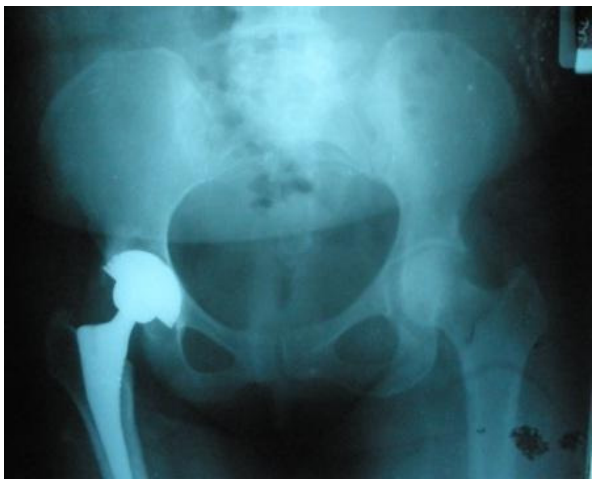

Figure 2: 2 weeks after a cement right total hip arthropalsty

Discussion

Osteonecrosis of the femoral head is an important complication of sickle cell disease with prevalence from 2.9% to 41 % [1,3] and with an Incidence ranging from about 2 to 4.5 cases per 100 patient-years [4]. We have reviewed the literature on osteonecrosis and sickle cell disease. Some aetiopathogenetic mechanisms have been suggested for osteonecrosis in sickle cell disease. Some studies have implicated increased intraosseous pressure due a disorder in circulation supplying the proximal femur with occlusion in to femoral head sinusoid by sickle red cells [5].

This phenomenon has been implicated in the cause of pain and evolution of osteonecrosis. In our case, patient reported pain with progressive limited range of movement at time of diagnosis. Others studies have implicated thrombophilia and decreased fibrinolysis as a result of decreased levels of natural coagulation inhibitors [6]. Some studies showed a relation between higher number of hospitalizations and osteonecrosis [7]. This case revealed a history of multiple hospitalizations due to severe sickle cell crisis.

Our patient presented late. This case revealed the problematic of early diagnosis, regular follow-up and early detection of complications in African SCD patients' especially asymptomatic osteonecrosis of the femoral head [8,9]. At start, this disease was frequently asymptomatic and it is reported found that 60 % of these patients with this complication were asymptomatic and was significantly associated with a history of leg ulcer[10]. It is known that untreated asymptomatic osteonecrosis of the femoral head in patient with sickle cell disease has high likehood of progression to pain and collapse. We are not sure of the result of arthroplasty in our cause because of high prevalence of complications after total hip (figure 2). Arthroplasty in SCD patients should be given to early surgical intervention in attempt to retard progression of this complication.

Conclusion

This case report pointing out the problem encountered in the specific diagnosis of hip disease in limited resource-settings as in Democratic Republic of Congo. There is need for early institution of preventive and therapeutic protocol for global management of sickle cell disease. Considering the high prevalence of sickle cell disease in Democratic Republic of Congo, the paucity of facilities available for total hip replacement we recommend community educational program for early detection of this complication and regular screening of patients with SCD particularly during childhood.

References

1. Ndugwa CM. Aseptic necrosis of the head of the femur among sickle cell anemia patients in Uganda. *East African Medical Journal*. 1992; 69(10): 572-6.
2. Tshilolo L, Aissi LM, Lukusa D, Kinsiama C, Wembonyama S, Gulbis B, Vertongen F. Neonatal screening for sickle cell anaemia in the Democratic Republic of the Congo: experience from a pioneer project on 31 204 newborns. *Journal of Clinical Pathology*. 2009; 62(1): 35-8.

3. Milner PF, Kraus AP, Sebes JI, Sleeper LA, Dukes KA, Embury SH, et al. Sickle cell disease as a cause of osteonecrosis of the femoral head. *New England Journal of Medicine* 1991; 325(21): 1476-8.
4. Mukisi MM, Bashoun K, Burny F. Sickle-cell necrosis and intraosseous pressure. *Orthopaedics & traumatology, surgery & research*. 2009; 95 (2): 134-8.
5. Cenni E, Fotia C, Rustemi E, Yuasa K, Caltavuturo G, Guinti A, Baldini N. Idiopathic and secondary osteonecrosis of the femoral head show different thrombophilic changes and normal or higher levels of platelets growth factors. *Acta orthopaedica* Acta Orthop. 2011; 82(1): 42-9.
6. Akinyoola AL, Adediran IA, Asaleye CM, Bolarinwa AR. Risk factors for osteonecrosis of the femoral head in patients with sickle cell disease. *International orthopaedics* 2009 Aug; 33(4): 923-6.
7. Akakpo-Numado GK, Gnassingbe K, Sakiye KA, Boume MA, Amadou A,Tekou H. Aseptic osteonecrosis of the femoral head in children with sickle- cell disease. *Sante* 2008; 18(4): 231-3.
8. Mont MA, Zywiel MG, Marker DR, McGrath MS, Delanois RE. The natural history of untreated asymptomatic osteonecrosis of the femoral head: a systematic literature review. *J Bone Joint Surg Am.* 2010; 92(12): 2165-70.
9. Hernigou P, Habibi A, Bachir D, Galacteros F .The natural history of asymptomatic osteonecrosis of the femoral head in adults with sickle cell disease. *Journal of bone and joint surgery. American volume.* 2006; 88(12): 2565-72.
10. Mukisi-Mukaza M, Saint Martin C, Etienne-Julan M, Donkerwolcke M, Burny ME, Burny F. Risk factors and impact of orthopaedic monitoring on the outcome of avascular necrosis of the femoral head in adults with sickle cell disease: 215 patients case study with control group. *Orthop Traumatol Surg Res.* 2011; 97(8): 14-20.