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Research Article

Sickle Cell Osteonecrosis, Report of 31 Cases in Congo Brazzaville

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Abstract

Goal: Improve the management of osteonecrosis in sickle cell patients.

Materials and Method: Retrospective study, conducted from January 1, 2001 to June 30, 2010, i.e. 9 years and 6 months in the departments of Rheumatology, Clinical Hematology, Orthopedic Traumatology, Childhood Surgery and Pediatrics Grand Children of the University Hospital of Brazzaville-Congo. The diagnosis of sickle cell osteonecrosis was made based on anamnestic, clinical and paraclinical arguments (radiological classification by Arlet and Ficat).

Results: We included 31 cases of sickle cell osteonecrosis (0.6%), including 18 female (58.1%) and 13 male (41.9%), with a sex ratio of 0.72 (M/F). The mean age was 25.6 years (range 10 and 44). The hip was the seat of the most common osteonecrosis: left unilateral in 15 cases (48.4%), right unilateral in 12 cases (38.7%) and bilateral in 4 cases (12.9%), follow-up of the shoulder, the location of which was bilateral in 2 cases (6.4%). According to the Arlet and Ficat classification, osteonecrosis was stage III in 15 patients (47.4%), stage II in 14 patients (45.1%) and stage IV in 2 cases (6.4%). The treatment combined medical treatment in all patients and surgical treatment in 6 patients (19.3%).

Conclusion: This work underlines that osteonecrosis is a serious and disabling complication of sickle cell anemia. Adequate management involves analgesics and NSAIDs in our context.

Keywords: Osteonecrosis; Sickle Cell Disease; Hemoglobinopathy S

Introduction

Sickle cell disease was discovered about 100 years ago. It remains a public health problem around the world [1]. Its prevalence is estimated at 7% of the world's population. Each year, 40,000 children are born with sickle cell trait worldwide, including 30,000 in Africa [2,3]. In its homozygous form, mortality is significant before the age of 5 years. Congo Brazzaville is one of the most affected African countries, around 20 to 30% of the population carries S and 1 to 2% on average is homozygous [1,4]. Among the most frequent and serious chronic complications of sickle cell anemia, osteonecrosis ranks first. Sickle cell osteonecrosis affects younger subjects from the age of 10 and is bilateral in half of cases [2,5]. The difficulty lies in the fact that its predictive factors are not known and that its management is uncertain in underdeveloped areas and osteonecrosis lacks standardized preventive treatment. It has no effective treatment in the early stages before the onset of structural changes. Landfill in the initial phase would require complete and prolonged bending, illusory in young active patients. That is to say

the interest of current techniques such as drilling associated with a cellular contribution by hematopoietic bone marrow or cement. Sickle cell osteonecrosis has been the subject of several studies in other countries. There is a classification linking the radiological stage of osteonecrosis of the femoral head and the treatment proposed in sickle cell patients. In Congo Brazzaville, there are no studies on sickle cell osteonecrosis. The aim of our study was to improve the management of osteonecrosis in sickle cell patients at the Brazzaville-Congo CHU.

Patients and Methods

This is a descriptive retrospective study, conducted from January 1, 2001 to June 30, 2010, i.e. 9 years and 6 months, in the departments of Rheumatology, Clinical Hematology, Orthopedic Traumatology, Infant Surgery and Pediatrics Large Children of the University Hospital of Brazzaville-Congo. From the data collected in the medical files, the patients were contacted individually by telephone or reached at home. After informed consent, the patients were seen in consultation in the rheumatology department of the CHU, in order to collect the socio-demographic characteristics (age, sex, profession, origin), the site of the osteonecrosis, to assess the clinical evolution, in particular the impact functional according to Lequesne's algofunctional score:

- Anamnestic plan, antecedents of foot-May syndrome, vaso-occlusive crises, blood transfusion and homozygous or heterozygous parents.
- On the clinical level: vaso-occlusive and deglobulization crises in the critical period, a sickle cell facies, jaundice, splenomegaly and failure to thrive in the inter-critical period.
- Biologically, by electrophoresis of hemoglobin with an alkaline pH of 8.5 + -0.1 on cellulose acetate.

And that of osteonecrosis in front of:

- Clinically, on the presence of localized mechanical pain or inflammatory pseudos without general signs, with or without limitation of joint amplitudes.
- In terms of imaging:
- Standard x-ray, a lesion showing signs of osteonecrosis according to the ARLET and FICAT classification.
- Magnetic resonance imaging, by highlighting a peri necrotic border appearing as a thin band in hypointense in T1 in serpentine going from one cortical to another or a double border of low segment double of a hypointense on T2-weighted or stir sequences which circumscribe the lesion.

A necrotic zone limited by the hypointense border, a medullary edema in T1 hypointense, T2 hypersignal enhanced after injection of gadolinium and a hypointense T1 and T2 hypersignal joint effusion without enhancement after injection of the contrast product.

- Scan by the presence of a heterogeneous zone with juxtaposition of hypo and hyperdense zone.

31 documented cases are the subject of this study. Were excluded from our study, patients with osteonecrosis of other etiologies, sickle cell patients without osteonecrosis, chronic inflammatory, degenerative, microcrystalline or metabolic rheumatic pathologies, infectious and unusable medical records. The SPSS 22 software enabled the analysis of clinical data.

Results

There were 31 cases, with a hospital prevalence of 0.6%, of which 18 were female (58.1%) and 13 were male (41.9%). The average age was 25.6 years +/- 10.1 with extremes of 10 and 40 years. The sex ratio (M/F) was 0.72. 22 (70.9%) of our patients were pupils and students, traders in 5 cases (16.1%), teachers, the unemployed, hairdressers and accountants respectively in 1 case (3.2%). The clinical manifestations were dominated by pain (100%), followed by lameness in 27 cases (87.1%). The pain was inflammatory in 18 cases (58.1%) and mechanical in 13 cases (41.9%) (Table 1). The hip was the seat of the most common osteonecrosis: left unilateral in 15 cases (48.4%), right unilateral in 12 cases (38.7%) and bilateral in 4 cases (12.9%), follow-up of the shoulder, the location of which was bilateral in 2 cases (6.4%). The radiological evolutionary stages of osteonecrosis at the time of diagnosis according to the Arlet and Ficat classification for the hip were the most found: stage 3 (Figure 1) in 47.4%, stage 2 in 45.1% and stage 4 in 6.4% of cases and for the shoulder were stages 2 and

Symptoms	NOT	%
Pain	31	100
Limitation of amplitudes		
Extension	7	22.5
Abduction	4	12.9
Adduction	4	12.9
Internal rotation	4	12.9
External rotation	4	12.9
Lameness	27	87.1
Quadricipital atrophy	3	9
Shortening of the lower limb	2	6.45

Table 1: Clinical manifestations.

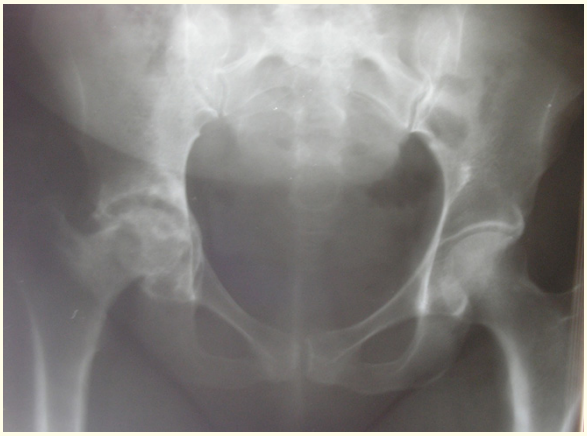

Figure 1: X-ray of the pelvis showing osteonecrosis of the right femoral head stage III (subchondral fracture in the form of linear clarity in eggshell, Loss of sphericity of the femoral head when stepping and a pseudo widening of the coxofemoral space).

3 respectively in 3.2% of cases. Biologically, the mean hemoglobin level at the time of diagnosis was 8.4 g/l + -2.4 with extremes of 4.3 and 12.4 g/l. The mean hospital stay was 17.8 days + -9.5 with extremes of 6 and 42 days. On the therapeutic level, medical treatment was instituted in all patients combining 1st, 2nd or 3rd level analgesics of the WHO, non-steroidal anti-inflammatory drugs, medical rest and a prescription for an English cane in one case. Surgical treatment in 19.3% of cases combining traction at stages II and III respectively in 2 cases (33.3%) and a Moore prosthesis at stage IV of Arlet and Ficat in 2 cases (33.3%). The outcome after surgical treatment was favorable in 4 cases (66.8%) and unfavorable in 2 cases (33.2%).

Discussion

We found 31 cases of sickle cell osteonecrosis in 4995 patients hospitalized at the CHU/B, i.e. a hospital prevalence of 0.6%. This prevalence is close to that of Coulibaly, which found 0.73% in Mali [6]. The difference is probably explained through recruitment.

In our study, the average age was 25.6 years + -10.1, lower than that of Coulibaly, 31 years old [6] and slightly higher than that of K. N'Dri in Côte d'Ivoire [7] who finds an average age of 22.7 years. This could be explained could be explained by the fact that K N'Dri included single adults with only one site of osteonecrosis and the patients were explored by the scanner. Sickle cell osteonecrosis occurs above all in young subjects, the age group most affected in our study is between 26 and 35 years old, just like Homawoo in Togo [8]. For Carayon and Hernigou in France [5,9], osteonecrosis appears between 7 and 15 years old. This is due to the fact that Carayon's patients are accessible to CT and magnetic resonance imaging. In Coulibaly's study [6] the youngest patient was 11 years old and in our series 10 years old. The fundamental reason for the attack of the young person suffering from sickle cell anemia is the fragility of the bone due to the multiple ischemia. Carayon [5] and Hernigou [9] found subjects aged on average 7 years. The MRI performed for the diagnosis could have explained this difference.

The sex ratio was 1.4 in favor of women, identical to that of Coulibaly in Mali [6] on 32 patients. Our two target populations are identical, perhaps the real reason lies in the structure of our societies whose population is predominantly female. On the other hand, Elira in Congo [10], out of 12 patients found a predominance of men, this can be explained by the fact that we included younger subjects from the age of 5 years, whereas in her study only subjects over age 30 were included.

Students and pupils were the most represented at 70.9%. This corroborates with the results of Coulibaly in Mali [6] who finds a student predominance in 40.6% of cases.

Pain was the main symptom, it was present in all cases of sickle cell osteonecrosis, followed by lameness in 87.1%. The shortening

of the lower limb was objectified in 6.45%. The pain was mechanical in 41.9% and pseudo-inflammatory in 58.1%. In the study by Coulibaly [6] both types of pain are observed with predominance on mechanical rhythm. Any persistent pain and lameness in a sickle cell patient of at least 10 years should suggest osteonecrosis.

Osteonecrosis preferentially affects the hip, 31 patients in our study, this result is comparable to that of Goudote in Benin on 22 patients [11], Coulibaly in Mali [6] on 32 patients and K'Dri in the Ivory Coast [7] in 38 patients. Their studies focused exclusively on osteonecrosis of the femoral head.

15 patients or 48.4% have unilateral left hip involvement and 12 patients or 38.7% have right involvement. Bilateral hip involvement was found in 12.9% (4 patients) in our series. Coulibaly [6] found the right side predominant in 12 patients, i.e. 37.5%, and the left side in 9 patients, i.e. 28.1%. The involvement was bilateral in 11 patients, i.e. 34.4% of cases.

The shoulder involvement is bilateral and associated with hip involvement in 6.4% of cases in our series.

Osteonecrosis was diagnosed mainly at stage III of Arlet and Ficat, in 47.4% of cases in our series. This result is identical to that of Coulibaly in Mali [6] who found 59.4% of patients consulted in stage III. This can be explained by a delay in consultation, the financial insecurity of our patients and ignorance.

The hemoglobin level was 8.4 g/dl + - 2.4, as reported by Elira who found a hemoglobin level between 7 and 8.5 g/dl in these sickle cell patients in these patients [10].

The results of hemoglobin electrophoresis at acidic pH could have enlightened us whether osteonecrosis is more frequent in the SC form or in the SS form. Carayon in France [5] and Goudote in Benin [11] find that the SS form is predominant. For Homawoo in Togo [8] SC hemoglobinosis is more frequent in osteonecrosis of the femoral head. This can be explained by the predominance of the SC form in West Africa. It is considered to be a homozygous form.

The medical treatment in our series combined analgesics according to the WHO levels, non-steroidal anti-inflammatory drugs and discharge by carrying a cane. Lafforgue [12] advises joint economy: rest wearing elastic heels, physiotherapy, carrying a cane, weight reduction and prohibition of sport except swimming and cycling.

In our study, no surgical treatment by osteotomy, drilling, drilling associated with bone grafting or cementoplasty was performed. However, 4 pull-ups were done followed by a plaster splint placed for 3 months in older children, 2 of whom were in stage III of Arlet and Ficat. One patient in Alert and Ficat stage III had an unfavorable outcome. GK Akakpo-Numado., *et al*. [13], found 6 out of 8 patients in stage III with a favorable outcome after traction followed by a thomas splint for an average of 14 months, 2 patients in stage III of Arlet and Ficat had an unfavorable outcome. Traction followed by a splint in older children gives immediate favorable clinical results. 2 cases of Moore prostheses were noted in our series. Lafforgue [12] notes that osteotomies are complex and abandoned, drilling of the femoral head and neck by the Trans trochanteric approach has finally been found ineffective in randomized trials even at the prefractural stage. Adding a graft doesn't make it any better. On the other hand, in a series of Hernigou [14], injection by drilling of an autologous stem cell transplant gave favorable results. The technique is not available in Congo.

Conclusion

Osteonecrosis is a serious and disabling complication of sickle cell anemia. Our study made it possible to determine the hospital prevalence which is 0.6% with an average age of 25.6% years. Adequate management involves analgesics, NSAIDs and joint economy. The appropriate treatment is impossible in our context, due to the fact of an under-equipped technical platform. Traction followed by immobilization by a splint has its place in the therapeutic arsenal of sickle cell osteonecrosis in children to avoid surgery. The evolution is towards osteoarthritis. The hip prosthesis remains the treatment of choice in IV.

Conflicts of Interest

The authors declare no conflict of interest.

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