

Sociodemographic and Functional Evaluation of Patients with Osteonecrosis of the Femoral Head Secondary to Sickle Cell Disease

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Abstract

Objective: To determine the sociodemographic and functional profile of patients with hip osteonecrosis secondary to sickle cell anemia.

Materials & Methods: A cross-sectional, observational, qualitative, and quantitative study performed in individuals with sickle cell disease affected with osteonecrosis of the femoral head from April to December 2016. A sociodemographic structured questionnaire and the Lequesne Algofunctional Questionnaire were used.

Results: A total of 101 hips were evaluated in 61 patients, with a mean age of 31.8 years, predominantly women and black, with low literacy, presenting with hemoglobin electrophoresis with HbSC expression and bilateral hip involvement, and receiving some benefit modality. Using the Lequesne questionnaire, patients were classified as having extremely serious impairment.

Conclusion: Osteonecrosis of the femoral head secondary to sickle cell disease affects young patients, preferably female, and has a high risk of bilaterality. Functional impairment is severe in most patients and associated with the inability to perform job activities.

Keywords: Sickle cell anemia; Osteonecrosis; Hip injuries

Abbreviations: SCD: Sickle cell disease; USA: United States of America; MRI: Magnetic resonance imaging; HbSC: Hemoglobin SC; HbSS: Hemoglobin SS

Introduction

Sickle cell disease (SCD) is the most commonly inherited hematological disease in the world [1]. It encompasses a group of inherited hemoglobinopathies of great clinical and epidemiological importance [2], and sickle cell anemia, among the several forms of SCD, is most commonly found in Brazil, occurring predominantly among Afrodescendants [3]. SCD emerged some 50 to 100 thousand years ago, between the Paleolithic and Mesolithic periods, in the countries of Central West Africa, India, and East Asia [4].

SCD is the most frequent hemoglobinopathy in our country and constitutes a public health problem, due to the expressive number of patients with this disease. In Brazil, especially in Bahia, its real incidence is still the object of studies. However, considering the racial characteristics of the Brazilian population, it is probable that the high prevalence of this disease is correlated with the high percentage of racial miscegenation [5].

In Brazil, there is an annual birth of about 700 to 1,000 people with SCD. About 0.1–0.3% of the Brazilian black population is affected by the disease in the homozygous form, with a tendency to increase, due to the process of miscegenation [6]. The heterozygous genotype affects Afrodescendants with a percentage of 10% in the USA, 6% in the South and Southeast of Brazil, and approximately 15.7% in the state of Bahia. In Bahia, one out of every 650 children born has SCD, and one in 17 has a sickle cell trait [7].

Aseptic necrosis of the femoral head, also recognized in the literature as avascular necrosis or osteonecrosis, affects mainly young adults in the 30- to 50-year age group with a prevalence among men [8]. The exact prevalence of femoral head osteonecrosis is unknown. In the United States of America (USA), it is estimated that there are between 10 and 20 thousand patients diagnosed per year and represents approximately 10% of the arthroplasties performed. When it's associated with corticosteroid therapy, usually affects young adults, with a mean age of around 40 years [9]. The incidence of osteonecrosis is 10% to 30% of the population with SCD, and often there is bilateral impairment [5].

Osteonecrosis primarily affects the hip more frequently but can also affect other sites such as the knee, shoulder, ankle, spine, and other joints. The greater the extent of the disease in several joints, the greater the limiting and painful effect for the patient. SCD is directly related to multifocal osteonecrosis [10].

Ficat and Arlet were the first authors to classify osteonecrosis initially in four stages, in which the pre-radiographic phase (currently diagnosed by magnetic resonance imaging -MRI) is recognized as stage I; the stage of repair or pre-collapse as stage II; the phase of collapse as stage III; and the sequela phase or arthrosis as stage IV [11]. Ficat, in 1985, added stage 0, in which the hip contralateral to the one already diagnosed with osteonecrosis is considered suspect, due to the high frequency of bilaterality, but it is truly silent. This author also recognized a phase, called the transition phase, which corresponds to the subchondral collapse characterized by the sign of the crescent. Two years later, Warner classified the stage of subchondral (transitional phase) collapse as IIB and segmental collapse as III [12].

The Lequesne questionnaire was developed in France in the 1970s and published for the first time in the 1980s. This questionnaire aims to assess hip functionality and was updated in 1997 and revised again in 2003. This index is composed of 11 questions on pain, discomfort, and function, having six questions on pain and discomfort, one over distance when walking, and four on activities of daily living. Scores range from 0 (no severe impairment) to 22 (extremely severe impairment). The instrument assesses symptoms and physical disability and was translated and validated into Portuguese in the year 2006 [13].

Objective

The aim of the study was to determine the sociodemographic and functional profile of patients with hip osteonecrosis secondary to sickle cell anemia. We used the Lequesne Somewhat Functional Questionnaire for the functional evaluation.

Methodology

This is a cross-sectional, observational quantitative study of 61 patients of both sexes and all ages, with SCD affected by osteonecrosis of the femoral head attended at the Orthopedics and Traumatology Department in the period of 01/02/2017 to 01/02/2018.

A research instrument containing questions on sociodemographic

aspects and the Lequesne Somewhat Functional Questionnaire were used.

The work was approved by the Research Ethics Committee under registration number 06/2005 and by National Commission for Research Ethics according to registration number 1457/2005. The patients participating in the study provided written informed consent.

Results

We interviewed 61 patients with SCD with clinical and radiographic diagnoses of femoral head osteonecrosis. The maximum age of the patients was 59 years, and the minimum was 12 years, with a mean of 31.8 years. There were 24 men (39.3%) and 37 women (60.7%). Of the patients, 55 came from the state of Bahia, 4 from the state of Pará, and 2 from Rio de Janeiro. As for ethnic groups, 36 patients identified themselves as black and 25 as brown. Regarding education, 20 patients did not complete primary level and 7 completed primary level, 11 did not complete secondary level and 13 completed middle level, 7 did not complete tertiary level and 3 completed tertiary level (Table 1).

Hemoglobin expression was verified through previous examinations of the patients, where it was observed that 59.3% of the patients had HbSC and 40.7% HbSS. Regarding the affected side, we found bilateral involvement in 38 patients (62.3%). Of the patients evaluated, 41 (67.2%) had no other joint affected by osteonecrosis, and 20 (32.8) had multifocal osteonecrosis.

Regarding issues related to work, occupation, and employment, we found that 32 (52.4%) patients received social security benefits. 24

patients received sickness benefits, and 8 received disability retirement benefits. Among these patients, the mean age was 37.4 years. Five (8.2%) patients did not work because they were minors, and 11 (18%) were unemployed. Of all patients, 13 (21.3%) were employed.

The evaluation of family income, the sum of the income of each household member, showed that 12 families had income up to minimum wage, 35 families had earnings between 1 and 3 minimum wages, 12 families earned between 3 and 4 minimum wages, and only 2 reported that their family income exceeded 5 minimum wages.

Of all the evaluated patients, 40 had bilateral osteonecrosis (65.57%), while 21 had unilateral, with a total of 101 hips evaluated. Table 2 shows the distribution of the hips according to the classification of Ficat and Arlet.

The evaluation of patients with the Lequesne questionnaire (Table 3) showed that 32 (52.45%) of the 61 patients interviewed in the study were classified as having extremely severe impairment, 4 (6.55%) very severe, 5 (8.19%) severe, 9 (14.75%) moderate, and 11 (18.03%) little functional impairment.

Discussion

Sex seems to influence femoral head osteonecrosis severity, having a higher frequency in women than men, and there is an increased risk of the osteonecrotic process in women as evidenced by this sample⁵. The mean age of the participants was 31.8 years, which indicates an early onset of the disease in this population.

With respect to education and income, we observed a low level of socioeconomic status and formal education, with the majority of patients not having completed high school. More than half of the patients received social security benefits (52.4%) related to the disease, and among these patients, the average age was only 37.4 years. This aspect shows the serious consequence of the disease for not only patients and health systems but also welfare since young patients of productive age become incapable of exercising paid activity that guarantees them subsistence.

The literature shows that corticosteroid-induced femoral head osteonecrosis presents about 55% bilaterality [14]. In the studied sample, the bilaterality was 65.57%, significantly higher than that found in the literature for osteonecrosis due to other causes.

The evaluation of radiological severity of the disease was initially evaluated in the study by means of lesion classification using the Ficat and Arlet classification. Considering the disease at

Table 1: Sociodemographic aspects.

Age	
Maximum	59
Minimum	12
Mean	31.8
Sex	
Male	24
Female	37
Naturalness	
Bahia	55
Pará	4
Rio de Janeiro	2
Black	36
Brown	25
Education	
Incomplete primary level	20
Complete primary level	7
Incomplete secondary level	11
Complete secondary level	13
Incomplete tertiary level	7
Complete tertiary level	3
Marital Status	
Married	22
Single	39

Table 2: Ficat and Arlet Classification.

Ficat and Arlet Classification	Hips
I	9 (8.91%)
II A	32 (31.68%)
II B	10 (9.9%)
III	23 (22.77%)
IV	27 (26.73%)

Table 3: Index of severity for osteoarthritis of the hip by Lequesne.

Severity	Points	Number of patients
Extremely severe	≥ 14	32 (52.45%)
Very severe	11–13	4 (6.55%)
Severe	8–10	5 (8.19%)
Moderate	5–7	9 (14.75%)
Mild	1–4	11 (18.03%)

early stages I, IIa and IIb, we observed that only 51 (50.4%) had early-stage disease. Twenty-three (22.77%) patients in stage III had an increased risk of secondary coxofemoral osteoarthritis, while 27 (26.73%) had coxarthrosis.

The functional evaluation of disease severity was assessed using the Lequesne questionnaire. Forty-one patients (67.21%) were classified as having severe functional impairment (severe, very severe, and extremely severe). Only 11 (18.03%) patients reported little functional impairment.

In a study by Asnani et al. [15], the mean life expectancy of patients with SCD was 53 years for men and 58.5 years for women. In another study, it was observed that about 11% of patients with SCD do not survive until adulthood [16]. Thus, SCD is responsible for the high risk of mortality in its carriers. The results show that, in addition to this previously known risk, femoral head osteonecrosis increases morbidity with consequent functional limitation and work restriction.

Conclusion

Osteonecrosis of the femoral head secondary to SCD affects young patients, preferably female, and has a high risk of bilaterality. Functional impairment is severe in most patients and related to a great inability to perform work.

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Author's contributions

Study design: DAVR, TBF, MRGP. Data collection: MRGP, TBF, LFSL. Data analysis: TBF, LFSL, BAFMF. Study supervision: VAF, GD. Manuscript writing: TBF, DAVR. Critical revisions for important intellectual content: GD.

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