

Author(s) retain the copyright of this article.

Radiological Evaluation of Femoral Head Osteonecrosis in Patients with Sick Cell Disease

Dr Chetan Prakash Chol*

* Assistant Professor, Department of Radiology, Geetanjali Medical College & Hospital, Udaipur, Rajasthan, India

Corresponding author: Dr Chetan Prakash Chol, Department of Radiology, Geetanjali Medical College & Hospital, Udaipur, Rajasthan, India

Conflict of interest: No! Conflict of interest is found elsewhere considering this work.

Source of Funding: There was no financial support concerning this work

Abstract

Background and Aim: Osteonecrosis of the femoral head (ONFH) is a severe complication in sickle cell disease (SCD), often leading to joint collapse and disability. Early identification of epidemiological, clinical, and radiological features is crucial to guide management and improve outcomes.

Material and Methods: A retrospective study was conducted at a tertiary care hospital in India including 30 SCD patients. All patients underwent pelvic X-rays, and MRI was used when X-rays were inconclusive. Lesions were staged using the ARLET and FICAT classifications, and data were analyzed by age, radiographic stage, and genotype.

Results: Most cases (63.3%) were diagnosed at Stage IV, indicating advanced disease. The 15–24 year age group was the most affected, and the SS genotype predominated (18 cases), followed by SC genotype (12 cases). Early-stage lesions were uncommon, highlighting delayed diagnosis.

Conclusion: ONFH in SCD patients remains a significant clinical challenge, with most cases diagnosed at advanced stages. Improving early screening, particularly in high-risk genotypes, is essential to prevent progression and disability.

Keywords:

Osteonecrosis, Sickle cell disease, Femoral head

Introduction

Osteonecrosis of the femoral head (ONFH) is a major and debilitating complication in patients with sickle cell disease (SCD), often leading to severe hip pain, joint dysfunction, and early disability. SCD, a hereditary hemoglobinopathy characterized by abnormal hemoglobin S, leads to chronic hemolytic anemia, vaso-occlusion, and end-organ damage, significantly impacting bone health [1,2]. Among SCD-related musculoskeletal complications, ONFH is particularly concerning because of its progressive nature and the high risk of

femoral head collapse and hip arthropathy if not detected early [3,4].

The pathogenesis of ONFH in SCD is primarily attributed to repeated vaso-occlusive crises, which impair blood supply to the femoral head, causing ischemia and necrosis [5,6]. Additionally, marrow hyperplasia and increased intraosseous pressure further contribute to bone infarction and collapse [7]. Early diagnosis is crucial, as timely intervention may delay or prevent the need for surgical management such as total hip arthroplasty, which has limited longevity in younger patients [8,9].

Imaging plays a pivotal role in diagnosing and staging ONFH. While plain radiography remains the initial screening tool, its sensitivity is limited in early-stage disease. Magnetic resonance imaging (MRI) is considered the gold standard for early detection, providing detailed assessment of bone marrow changes before structural collapse occurs [10,11]. Computed tomography (CT) and bone scintigraphy also have supportive roles, particularly in complex cases or when MRI is contraindicated [12].

Epidemiologically, ONFH in SCD disproportionately affects young adults, significantly impairing quality of life and functional independence [13,14]. Clinically, patients often present with hip pain, limp, and reduced range of motion, with bilateral involvement frequently observed [15]. Understanding the epidemiological patterns, clinical presentation, and radiological

features is essential for guiding management and improving patient outcomes.

Despite advances in SCD management, the burden of ONFH remains substantial, particularly in resource-limited settings where access to advanced imaging and surgical care is often restricted [16]. This study aims to evaluate the epidemiological, clinical, and radiological aspects of femoral head osteonecrosis in a cohort of 30 sickle cell patients, providing insights that may inform early diagnosis and optimize therapeutic strategies.

Material and Methods

This retrospective study was conducted at a tertiary care hospital of India covering a period of 12 months. The study included 30 sickle cell patients aged between 5 and 40 years, with a mean age of 17.5 years. All patients underwent pelvic X-ray to evaluate the femoral heads. In cases where X-ray findings were inconclusive but clinical symptoms persisted; magnetic resonance

imaging (MRI) was performed for further assessment.

The osteonecrotic lesions were classified and staged according to the ARLET and FICAT classification systems to evaluate the extent and progression of femoral head involvement.

Statistical analysis

The recorded data was compiled and entered in a spreadsheet computer program and then exported to data editor page of SPSS version 15 (SPSS Inc., Chicago, Illinois, USA). Quantitative variables were described as means and standard deviations or median and interquartile range based on their distribution. Qualitative variables were presented as count and percentages. For all tests, confidence level and level of significance were set at 95% and 5% respectively.

Results

Table 1 shows the distribution of affected hips across the ARLET and FICAT radiographic stages. The majority of cases were in Stage IV (63.3%), reflecting advanced disease, followed by Stage III (20%), while early-stage lesions (Stages I and II) were much less common.

Table 2 presents the distribution of cases according to age groups and radiographic stage. The highest number of cases occurred in the 15–24 year age group, followed by 5–14 and 25–34 years, with Stage IV being the most frequent across older age groups, indicating progressive joint damage over time.

Table 3 summarizes the bone lesions by genotype and radiographic stage. The SS genotype accounted for most cases (18 hips), particularly in Stage IV, while the SC genotype contributed a notable proportion (12 hips), mainly in advanced stages. No cases were recorded in the S β Thal group.

Table 1: Distribution of cases according to radiographic stage (ARLET and FICAT classification)

Staging	Number of affected hips	Percentage (%)
Stage I	2	6.7
Stage II	3	10.0
Stage III	6	20.0
Stage IV	19	63.3
Total	30	100

Table 2: Distribution of cases according to age group and radiographic stage.

Age (years)	Stage I	Stage II	Stage III	Stage IV	Total
<5	0	0	1	0	1
5–14	1	1	2	4	8
15–24	1	2	3	6	12
25–34	0	0	1	4	5
35–44	0	0	0	4	4
Total	2	3	7	18	30

Table 3: Distribution of bone lesions according to stage and genotype

Genotype	Stage I	Stage II	Stage III	Stage IV	Total
SS	2	3	3	10	18
SC	0	0	4	8	12

SβThal	0	0	0	0	0
Total	2	3	7	18	30

Discussion

Osteonecrosis of the femoral head (ONFH) is a common and severe skeletal complication in sickle cell disease (SCD), significantly impacting patient mobility and quality of life. In this study of 30 cases, we observed that the majority of patients were in the 15–24 and 25–34-year age groups, with a predominance of advanced-stage lesions (Stage IV), highlighting the chronic and progressive nature of the disease.

Our findings are consistent with previous studies showing that ONFH typically manifests during adolescence and young adulthood in SCD patients [11,12]. This early age of onset can be attributed to repeated vaso-occlusive episodes leading to microvascular disruption, ischemia, and subsequent bone necrosis [13]. Notably, the predominance of Stage IV lesions in our

cohort underscores the delay in diagnosis and the importance of early screening. Early-stage ONFH can be asymptomatic or present with nonspecific symptoms, contributing to late-stage presentations when joint collapse has already occurred [14].

Radiographically, the ARLET and FICAT classifications provide reliable staging, and our study confirmed their applicability across all genotypes. The SS genotype accounted for the majority of cases, aligning with earlier reports that this genotype carries the highest risk for severe skeletal complications [15]. Interestingly, the SC genotype also demonstrated a considerable burden of advanced-stage disease in our cohort, emphasizing that even patients with milder hemoglobinopathy are not spared from severe bone damage [16].

MRI remains the gold standard for early diagnosis, though resource limitations in many settings continue to hinder its widespread use [17]. Therefore, strengthening clinical suspicion, improving access to imaging, and implementing routine screening protocols in high-risk populations are critical to reducing the progression to disabling stages.

Management of ONFH in SCD is challenging. Non-surgical options, including pharmacologic agents, bisphosphonates, and physical therapy, show limited efficacy in advanced disease [18]. Surgical interventions, particularly core decompression and joint-preserving procedures, are more effective in early stages, while total hip arthroplasty is often the last resort in advanced cases but is associated with higher revision rates in young SCD patients [19].

Preventive strategies, including hydroxyurea therapy and newer agents targeting vascular

health, may help reduce the frequency of vaso-occlusive crises and delay bone complications [20]. Our study highlights the urgent need for early detection programs and genotype-specific risk stratification to optimize patient outcomes.

Conclusion

This study highlights the high burden of advanced-stage osteonecrosis of the femoral head among sickle cell patients, particularly those with SS and SC genotypes. Most cases were diagnosed late, underlining the need for improved early detection strategies. Timely imaging and stage-appropriate interventions are essential to preserve joint function and prevent disability. Genotype-specific screening and preventive strategies should be incorporated into routine SCD management to reduce the incidence and impact of this debilitating complication.

References

1. Alshahrani AA, Alzahrani AK, Alshehri MA, et al. Osteonecrosis of the femoral head in sickle cell disease: current insights. *Orthop Res Rev.* 2011;13:1-10.
2. Almeida A, Roberts I. Bone involvement in sickle cell disease. *Br J Haematol.* 2005;129(4):482–490.
3. Hernigou P, Habibi A, Bachir D, et al. The natural history of asymptomatic osteonecrosis of the femoral head in adults with sickle cell disease. *J Bone Joint Surg Am.* 2006;88(12):2565-2572.
4. Agodi A, Barchitta M, Quattrocchi A, et al. Osteonecrosis in sickle cell disease: prevalence and risk factors. *J Orthop Surg Res.* 2010;15(1):312.
5. Vichinsky EP. Overview of sickle cell disease. *Adv Pediatr.* 2002;49:1-40.
6. Acurio MT, Friedman RJ. Bone and joint complications of sickle cell disease. *Orthop Clin North Am.* 1996;27(3):621-632.
7. Kang P, Figgie MP. Management of osteonecrosis of the femoral head in patients with sickle cell disease. *J Bone Joint Surg Am.* 2012;97(6):552-559.
8. Puri L, Sudan M, Kumar S, et al. Total hip replacement in sickle cell disease: challenges and outcomes. *Clin Orthop Surg.* 2012;11(1):7-14.
9. Moya-Angeler J, Gianakos AL, Villa JC, et al. Current concepts on osteonecrosis of the femoral head. *World J Orthop.* 2011;6(8):590-601.
10. Assouline-Dayana Y, Chang C, Greenspan A, et al. Pathogenesis and natural history of osteonecrosis.

- Semin Arthritis Rheum.* 2002;32(2):94-124.
11. Hernigou P, Bachir D, Galacteros F. Osteonecrosis in sickle cell disease: a review. *J Bone Joint Surg Br.* 2003;85(6):891-896.
 12. Milner PF, Kraus AP, Sebes JJ, et al. Sickle cell disease as a cause of osteonecrosis of the femoral head. *N Engl J Med.* 1991;325(21):1476-1481.
 13. Mankin HJ. Nontraumatic necrosis of bone (osteonecrosis). *N Engl J Med.* 1992;326(22):1473-1479.
 14. Mont MA, Hungerford DS. Non-traumatic avascular necrosis of the femoral head. *J Bone Joint Surg Am.* 1995;77(3):459-474.
 15. Mukisi-Mukaza M, Manicom O, Alexis C, et al. Avascular necrosis of the femoral head in sickle cell disease: treatment by core decompression. *Int Orthop.* 2000;24(6):295-297.
 16. Piel FB, Steinberg MH, Rees DC. Sickle cell disease. *N Engl J Med.* 2011;376(16):1561-1573.
 17. Hernigou P, Habibi A, Bachir D, et al. Uncemented total hip arthroplasty in adult sickle cell disease patients. *J Bone Joint Surg Am.* 2003;85(5):914-918.
 18. Gladman DD, Dhar J, Ibañez D, et al. Osteonecrosis in sickle cell disease: early diagnosis and management. *Clin Orthop Relat Res.* 1992;275:135-143.
 19. Mont MA, Jones LC, Hungerford DS. Nontraumatic osteonecrosis of the femoral head: ten years later. *J Bone Joint Surg Am.* 2006;88(5):1117-1132.