Total hip arthroplasty in sickle cell disease
(case report)

Abstract
Sickle cell disease (SCD), often leads to skeletal complications such as avascular necrosis (AVN) of the femoral head, osteoporosis, fractures, and infections. Despite available joint-preserving surgeries, many SCD patients require total hip arthroplasty (THA). However, THA carries significant risks for these patients, including hypoxia and vascular. This case report describes a 29-year-old female with SCD who presented with recurrent hip pain after a previous surgery was canceled due to a sickle cell crisis. Physical examination revealed restricted hip motion, and imaging confirmed AVN of the right femoral head. After a thorough hematology consultation, uncemented THA was performed. The surgery was uneventful, with minimal blood loss and no need for transfusion. The patient recovered well and was stable at her first follow-up. THA is common in SCD patients due to the high incidence of femoral head osteonecrosis, but they face higher risks of postoperative complications. Therefore, careful preoperative planning and coordination among surgical, anesthetic, and hematology teams are crucial for optimizing the patient for THA.
Keywords: Sickle Cell Anemia, Total Hip Arthroplasty, Osteonecrosis.

Accepted: 50 days before printing

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Introduction

Sickle cell disease (SCD) is a common autosomal recessive disorder caused by a point mutation in the β-globin chain of hemoglobin[1,2]. his genetic alteration induces an abnormal configuration of red blood cells, which tend to deform into a sickle shape under conditions of reduced oxygen levels. This impedes the blood supply to tissues and leads to vaso-occlusion, which is central to the pathophysiology of SCD. SCD patients exhibit a wide range of clinical manifestations, with key features including anemia, vaso-occlusive crises, and chronic organ failure[1].

Skeletal manifestations are common in sickle cell disease (SCD) and encompass conditions such as avascular necrosis (AVN) of the femoral head, osteoporosis, pathological fractures, and infections, including septic arthritis and osteomyelitis[2,3]. AVN of the femoral head poses a significant and debilitating challenge, affecting 20-50% of SCD patients, predominantly in their second or third decade of life[4]. This condition has a high probability of progressing to hip osteoarthritis (OA), usually in the third or fourth decade of life[2].

Despite the implementation of joint-preserving surgical strategies like core decompression, osteotomy, and bone grafting, many patients with SCD experience advanced osteonecrosis, necessitating total hip arthroplasty (THA)[5,6]. Recent advancements in surgical techniques, hardware, drug therapysuch as hydroxyurea, and stem cell transplantation have greatly enhanced the life expectancy and quality of life for SCD patients, with many living comfortably into their seventh decade[6,7].

Nevertheless, these patients continue to be high-risk candidates for surgery. The physiological stress associated with surgical procedures can trigger cytokine release, leading to hypoxia, hypoperfusion, and acidosis, conditions that facilitate red blood cell sickling and the subsequent occlusion of the microcirculation[8,9].

Given the unique challenges faced by SCD patients undergoing THA, we will present a detailed case study of a patient with SCD who was a candidate for this procedure. Furthermore, we will review current guidelines for the management of SCD patients in the context of THA.

Case Description

A 29-year-old female was referred to our orthopedic center at Imam Khomeini Hospital Complex, Tehran University of Medical Sciences, presenting with a chief complaint of right hip pain and limping for the past year. The patient is a known case of SCD. She had previously presented with the same complaint two months earlier, but her operation was canceled due to a sickle cell crisis, which included a decrease in hemoglobin from 10.7 to 8.9 g/dL and severe bone pain. She had a history of blood exchange and platelet injection two years prior, but no history of surgery, particularly orthopedic surgery. The patient had been taking tramadol and hydroxyurea irregularly. Her body mass index (BMI) was 21, and her Charlson Comorbidity Index (CCI) was zero (Figure 1).

On physical examination, Stinchfield's test was positive, indicating pain in the anterior groin. The range of motion (ROM) in various movements had decreased, with the greatest decrease observed in internal rotation (20 degrees below the lower limit of the normal value). A flexion contracture was noted at 10 degrees. The neurovascular examination was intact, and no skin scars were observed. Examinations of other systems were unremarkable. Laboratory investigations revealed hemoglobin at 10.6 g/dL, white blood cell count at 4,100/µL, platelets at 151,000/µL, erythrocyte sedimentation rate at 53 mm/h, C-reactive protein level at 39 mg/L, and an international normalized ratio (INR) of 1.07. Imaging studies showed avascular necrosis (AVN) of the right femoral head with a diameter of 38 mm (more than 50%), internal mild edema, and focal sclerosis on magnetic resonance imaging (MRI). A hip X-ray revealed a 1 mm leg length discrepancy and was classified as Dorr type B (Figure 2).

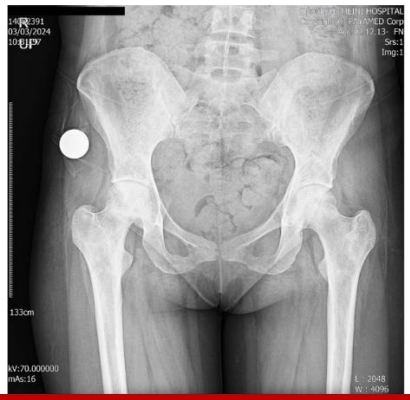

Figure 1: Preoperative pelvis anteroposterior (AP) radiograph

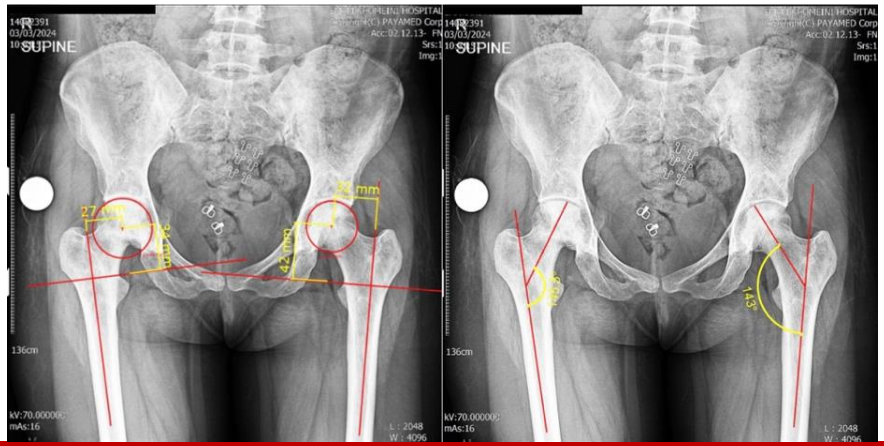

Figure 2: Preoperative planning. Right: medial offset = 27 mm, vertical offset = 34 mm, caput-collum-diaphyseal (CCD) angle = 145.5°; Left: medial offset = 32 mm, vertical offset = 42 mm, CCD angle = 143.0°.

With the diagnosis of AVN, THA was planned for the patient. A consultation with a hematologist was conducted to ensure the operation's safety, and after reviewing the laboratory tests, the hematologist approved the surgery. Four units of packed red blood cells and Intensive care unit (ICU) bed were reserved for the patient.

Preoperative vital signs were a blood pressure of 111/80 mmHg, heart rate of 95 bpm, and oxygen saturation of 95% on room air. Two grams of cefazolin were administered to the patient as prophylaxis one hour prior to the procedure. The uncemented THA was performed by the senior author (SMJ.M, a fellowship-trained knee surgeon). After the administration of anesthesia, 1.5 grams of tranexamic acid is given before the preparation and draping process begins. A thermal cautery is used to make the incision through the subcutaneous tissue and fat, followed by superficial hemostasis. The ascending and lateral circumflex artery branches, located between the Tensor Fasciae Latae and Sartorius muscles, are identified. These vessels are clamped using a Babcock clamp and cauterized for ligation. Once the neck is cut, the osteotomy site is temporarily sealed with bone wax, which remains in place until broaching of the femoral canal. At that point, the wax is removed with a curette. The implants used included a Zimmer Trilogy 52 mm cup, fixed with two screws; an M/L Tapered size 5 stem; and a Neutral (0 mm) VerSys 36 mm head. C-arm navigation was not utilized. The operation lasted 1.5 hours. During surgery, the patient's blood pressure, heart rate, and oxygen saturation remained stable, with 350 cc of blood loss and no transfusion required. Postoperatively, the patient was admitted to the ward and experienced an uneventful hospitalization and the leg length discrepancy in the patient was three millimeters. She was stable during her first follow-up visit one week after discharge (Figures 3 & 4).

Figure 3: Collapsed necrotic femoral head

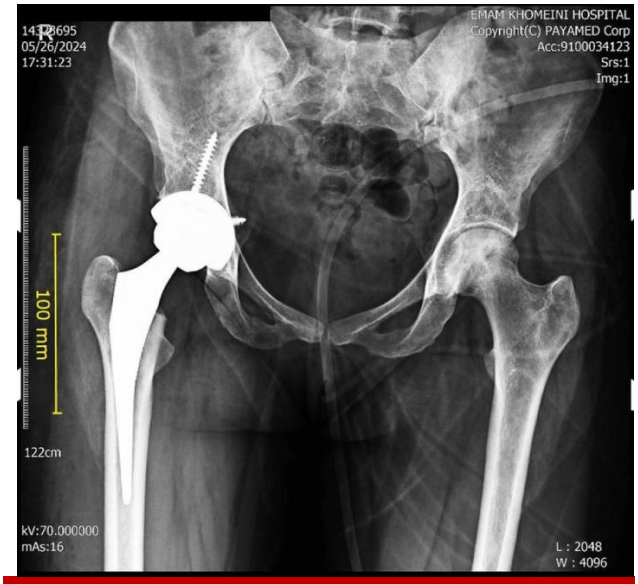

Figure 4: Postoperative pelvic anteroposterior (AP) radiograph.

At the final follow-up, conducted four months postoperatively, a radiograph was obtained, which revealed no complications. The patient also reported no dissatisfaction with the outcome.

Discussion

THA is frequently performed in patients with sickle cell disease (SCD) due to the high incidence of femoral head osteonecrosis, with some studies reporting rates as high as 50%[10]. Systematic reviews have shown that THA is an effective intervention for alleviating pain and enhancing mobility in patients with SCD. However, these patients face a greater risk of postoperative complications, both medical and surgical, compared to those without SCD[1,11]. Although various treatment options exist for femoral head osteonecrosis, most SCD patients inevitably progress to advanced osteoarthritis of the hip, rendering conservative management largely ineffective[12]. Due to the pathophysiology associated with SCD, alternative surgical treatments, such as core decompression, have shown limited success, with failure rates exceeding 40%[13]. Despite these challenges, studies have consistently demonstrated significant improvements in functional outcomes, as measured by the Harris Hip Score and Merle d'Aubigne score, following THA in SCD patients[11].

Preoperative assessment and effective communication among the surgical, anesthetic, hematologic, hospitalist, infectious disease, and cardiology teams are essential in managing these

patients. A thorough preoperative evaluation should routinely include laboratory tests to assess serial hemoglobin S percentage (Hb S%), renal and liver function, and oxygen saturation levels. These results guide the decision regarding the need for preoperative blood transfusion. Red blood cell exchange, aimed at reducing Hb S levels to below 30%, is reserved for patients with a history of severe acute chest syndrome, prior cerebrovascular events, or severe anemia with hemoglobin levels under 5 g/dL[14,15]. Adequate hydration is also vital to reduce blood viscosity and mitigate the risk of vaso-occlusive episodes[11]. Special attention is required for SCD patients on hydroxyurea therapy. Given the potential for myelosuppression, a complete blood count should be obtained preoperatively, and if any signs of toxicity are present, hydroxyurea should be temporarily discontinued for approximately one week to allow for recovery of blood counts[14].

The distinct pathophysiology of SCD presents several intraoperative challenges during THA. Extensive infiltration of fatty bone marrow into the femur often results in sclerosis and narrowing of the femoral canal, which increases bone fragility and the risk of intraoperative fractures[11]. In extreme cases of femoral canal narrowing, a dysplasia-type, narrow, straight cylindrical stem may be useful[12]. On the acetabular side, difficulties in properly seating the acetabular component are common, often accompanied by Paprosky type I acetabular defects. These factors can make multi-hole cups with screw fixation especially useful[16]. Concerning the use of cement, a systematic review found that patients with SCD undergoing cemented THA faced a higher risk of requiring revision surgery. Additionally, cemented THA was associated with an elevated rate of aseptic loosening[11]. Therefore, press-fit fixation for both femoral and acetabular components may be a more suitable approach for SCD patients. For those who undergo press-fit THA, surgeons may recommend a postoperative period of reduced activity to promote bony ingrowth of the implants, potentially reducing the likelihood of early aseptic loosening[17,18].

Post-operatively, SCD patients should be placed in a closely monitored unit. Optimal care is provided in a tertiary care center where the anesthesia team is experienced in managing and preventing vaso-occlusive crises. Nonsteroidal anti-inflammatory drugs (NSAIDs) offer both analgesic and antiplatelet effects and can enhance the pain-relieving properties of opioids, thereby minimizing narcotic use and lowering the risk of inducing vaso-occlusive episodes[19]. Due to the potential need for blood transfusions or exchange transfusions, significant blood bank resources may be required to maintain safe hemoglobin levels while preventing excessive blood viscosity[20]. Additionally, functional asplenia in SCD patients increases their susceptibility to infections, including wound infections and sepsis. It is crucial to involve the infectious disease team promptly if an infection is suspected to prevent these life-threatening complications. Clinicians must remain vigilant for signs of acute chest syndrome, such as cough, chest pain, fever, and hypoxia, as this condition is the leading cause of mortality among SCD patients[21].

SCD patients undergoing THA tend to have a longer LOS and higher rates of readmission compared to those undergoing THA for primary osteoarthritis [11]. This difference is largely attributed to the higher frequency of medical complications in SCD patients, such as pain crises, sepsis, acute chest syndrome, and other respiratory issues[22,23]. The recurrent polymerization and depolymerization of hemoglobin in SCD patients lead to increased red blood cell rigidity and stasis, which can obstruct blood flow and trigger venous thromboembolism (VTE) events[24]. Therefore, clinicians may opt for more aggressive deep vein thrombosis (DVT) prophylaxis and maintain a lower threshold for ordering duplex ultrasound evaluations. Regarding the management of pain crises and acute chest syndrome, careful monitoring of intraoperative blood loss and perioperative fluid balance can reduce these risks. This approach aligns with the recommendations of Vinchinsky et al., who emphasized the importance of optimizing oxygenation, pain control, and hydration in THA patients with SCD to minimize the occurrence of vaso-occlusive crises and acute coronary syndrome[25].

Patients with SCD face a heightened risk of surgical complications compared to those without SCD. Notably, the rates of wound complications, infections, and aseptic loosening are significantly elevated in this population. To mitigate infection risks, antibiotics, such as first- and second-generation cephalosporins at a dosage of 2.5 grams per day, should be administered intraoperatively and for three days postoperatively. The increased incidence of wound complications and infections may be linked to the impaired microvasculature and weakened immune response in SCD patients, which can result in

delayed wound healing and a reduced ability to prevent early wound contamination from progressing to periprosthetic joint infections. The higher rate of aseptic loosening in SCD patients is likely due to their younger age at the time of THA compared to patients undergoing THA for primary osteoarthritis[11,26,27]. Younger patients tend to have higher activity levels and greater functional demands, which place additional stress on the implant-bone interface, increasing the likelihood of aseptic loosening and implant failure. Consequently, surgeons might consider using press-fit fixation to encourage durable biological ingrowth, and patients should be informed of their potentially increased risk for future revision surgery due to aseptic loosening.

Conclusion

In conclusion, THA is frequently performed in SCD patients due to the high prevalence of femoral head osteonecrosis. Research has shown that individuals with SCD face an elevated risk of postoperative medical and surgical complications compared to non-SCD patients. As a result, comprehensive care is essential, necessitating thorough preoperative evaluation and coordinated communication among the surgical, anesthetic, hematologic, hospitalist, infectious disease, and cardiology teams to optimize outcomes for this high-risk patient population.

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