Case Report

A rare case of osteonecrosis of hip in sickle cell disease

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Abstract

Avascular necrosis of bone is a severe complication of sickle cell disease (SCD) and Management of these problems is often difficult because of the diagnostic imprecision of most laboratory and imaging investigations and because of the lack of evidence for most surgical procedures in sickle cell disease. Its treatment is not standardized. The objective in this case is to determine the impact of core decompression and PRP infusion in the management of Avascular necrosis of hip. In this case, a young Indian male with a known history of sickle cell disease presented to the clinic with severe bilateral hip pain. The pain had lasted for several months and had not improved with anti-inflammatory medication and starting on alandronate. There was severe pain with internal and external rotation of the hip. MRI of the femur showed stage 2 or 3 avascular necrosis of the femoral head, while X-rays of the femur were unremarkable. Patient managed conservatively by Non weight-bearing for several weeks and oral medication shortly thereafter, the patient underwent core decompression of the bilateral femoral head as well as continuing on Alendronate, a bisphosphonate. The patient improved temporarily but regressed shortly thereafter. His avascular necrosis worsened radiographically over the next several months. At this point, the only other option would be to do a total hip arthroplasty, but the patient may need several more throughout his lifetime due to the lifespan of the artificial replacement. There have only been scarce reports of avascular necrosis in patients with sickle cell trait. This manuscript presents such a case and includes the trials and tribulations associated with its management.

Key words

Sickle cell trait, Avascular necrosis, Sickle cell disease, Total hip arthroplasty, Hip, Core decompression.

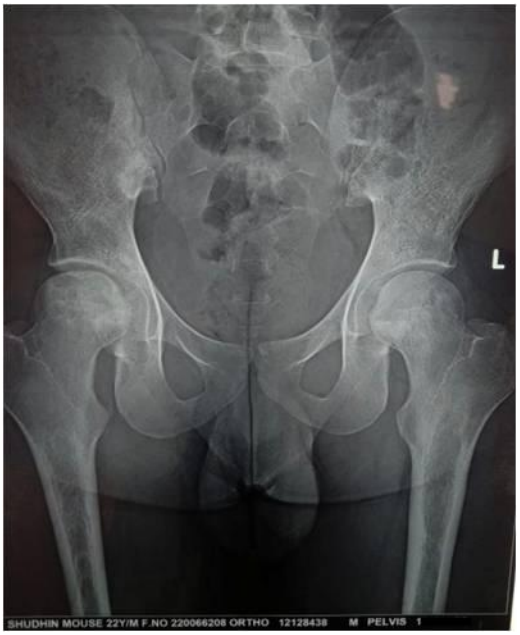
Introduction

Osteonecrosis, also known as avascular necrosis (AVN), is a disorder in which there has been an infarction of bone tissue due to ischemia. This can happen as a result of trauma, such as a fractured femoral neck, when the watershed areas that supply the femoral head are insufficiently large enough to prevent reduced blood flow from happening without ischemic repercussions. Once an infarction occurs, it just takes a short while for oxidative phosphorylation to become inefficient and necrosis to set in. In order for metabolic processes to function properly, the majority of biological tissues require oxygen. About 20,000 to 30.000 new diagnosis of AVN occur each year. Systemic lupus erythematosus (SLE), radiation therapy, coagulopathy caused by the factor V leiden mutation, excessive alcohol and glucocorticoid usage, and sickle cell anemia are just a few of the atraumatic related etiologies [1-4]. AVN is a multifactorial condition that can start with a disruption in the flow of blood and oxygen to the blood vessels in and around the bone before progressing to trabecular thinning (also seen in cases of osteoporosis), and ultimately, bone collapse. In the case of sickle cell disease, an infarction is brought on by red blood cells (RBCs) occluding the vasculature because they have altered their shape to flow less easily in the blood arteries. The RBCs are not as spherical and easily passable as typical RBCs; instead, they have a form resembling a crescent. Because of their form, they can stick to endothelium walls and other RBCs, causing vaso-occlusion. This may result in ischemia, bone marrow blockage, and ultimately progression to AVN. In sickle cell disease, it occurs fairly frequently. By the time they turn 35, up to 50% of sickle cell individuals may develop AVN. In contrast, sickle cell trait (SCT), a considerably more benign form of sickle cell disease in which patients are frequently asymptomatic, it is extremely uncommon. This case report intends to shed light on a rarely reported instance of AVN in sickle cell trait and the dilemma encountered during its treatment in a young Indian male.

Case report

A 22-year-old Indian man with a history of sickle cell trait arrives at the orthopaedic clinic complaining of throbbing bilateral hip pain. His father is a sickle cell trait carrier. He had never experienced an acute episode of sickle cell symptoms like severe, abrupt chest discomfort prior to two years ago. He did, however, have a history of chest trouble that was treated with medication and a hospital visit two years earlier. He claimed that he had the same hip pain about a year earlier. He saw his primary care physician, who prescribed him an over-the-counter pain reliever. These were beneficial for a while. He then had a history of slip and fall injuries that aggravated his pre-existing pain, prompting him to return to his primary care physician. He was referred to the orthopaedic clinic at this time. He had severe groin pain as well as internal and external hip rotation. X-rays of his hip (**Figure – 1**) revealed a necrotic area suggestive of stage 1 or 2 AVN in the head of the bilateral femur. **Figure - 2** shows an MRI that revealed stage 2 AVN on the left and stage 3 AVN on the right.

Figure - 1: Pre-operative X-ray.



He was then given bisphosphonates, instructed to walk with no weight bearing and support, and scheduled for a decompression procedure with PRP infusion several weeks later.

Figure - 2: Pre-operative MRI.

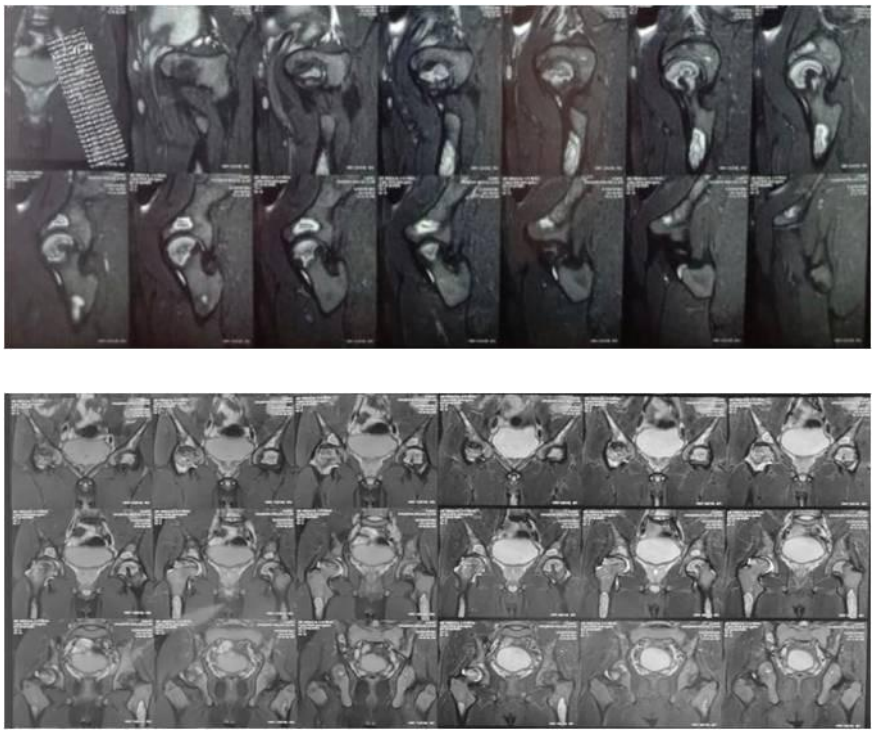
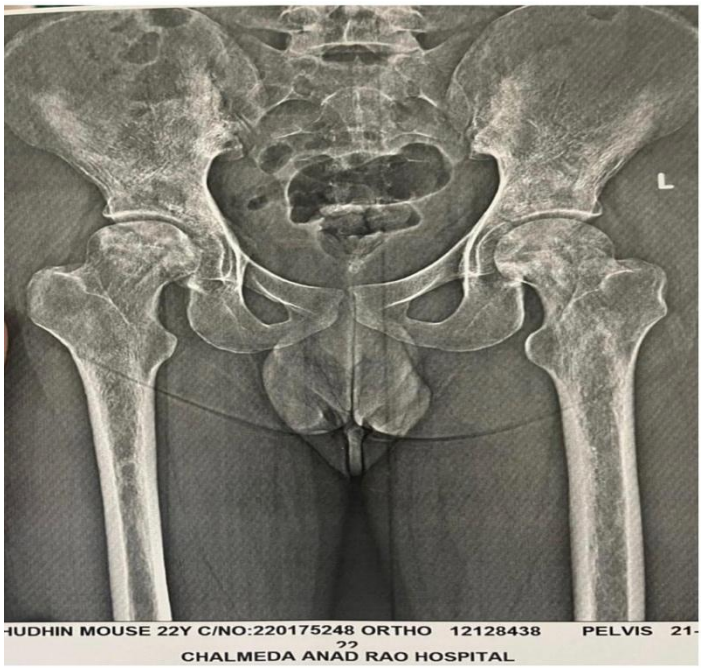


Figure – 3: Post-operative X-ray.



Following surgery, the plan was to limit weight bearing for 6 weeks while also taking Alendronate, a bisphosphonate. The patient's pain subsided after decompression. The patient returned for a check-up and suture removal two weeks after surgery. At the time, his hip x-rays were normal. **Figure - 3** depicts X-rays taken two weeks after surgery. He was able to bear weight on the hip six weeks after surgery. There was no pain during range of motion or internal and external rotation testing. He was then scheduled to begin physical therapy soon thereafter.

However, eight weeks after surgery, he returned to the clinic for a check-up, claiming fresh onset pain. He was able to bear weight, but the discomfort in his hip and groin had returned. There were also fresh X-ray results (AP Pelvis and Frog Hip views) that revealed a serpiginous line consistent with AVN development with no femoral head collapse. Even after bisphosphonates and decompression, the patient's AVN had spread to encompass the majority of the femoral head. At 3 months post-surgery, an MRI revealed that the femoral head was flattening, as well as volume loss and bone marrow edema.

There were no other conservative options for this patient at this stage. The issue is his age. Osteotomy is sometimes used to treat AVN, however given the necrosis of this patient's femoral head, this procedure is not recommended. This patient's treatment will remain non-weight bearing. Total hip arthroplasty is the only definite treatment for this patient (THA). Because this patient is so young, he was referred to a hip joint revision specialist, with a follow-up appointment set for 6 weeks later. At his age, a THA would be challenging because he would most certainly have to undergo the surgery numerous times throughout his life. Hip replacements have an average lifespan of 15-20 years, thus preserving as much of the joint as possible and avoiding THA at such a young age is critical. It would be catastrophic for the patient to require 4-6 hip replacements throughout the course of his life. Time will tell whether conservative therapy, such as reduced weight-bearing, bisphosphonates, and vitamin D, will have any discernible impact on this patient's hip. Two months passed between a clinically and radiographically effective surgery and the onset of pain.

Discussion and Conclusion

Two alpha chains and two beta chains make up the typical adult hemoglobin. Sickle hemoglobin is the name given to hemoglobin that can sickle (HbS). A single point mutation on the beta globin chain, from Glutamine to Valine, causes sickle hemoglobin. A person will develop sickle cell disease if they are homozygous, which means they inherited two beta globin chains that are mutated [5]. RBCs become weak and change shape as a result of the hypoxia, acidity, and dehydration caused by these aberrant beta chains. Sickled RBCs repeatedly occluding blood vessels will eventually result in ischemia, infarction, edema, and end-organ damage in any system of the bone [6, 7]. An individual with the sickle cell trait who is heterozygous will only acquire one mutant beta globin chain. Cells typically need 50% of HbS in order to be able to sickle. Individuals with the sickle cell trait typically exhibit no symptoms and have a prevalence of 40% [8]. However, there have been cases of foetal death, venous thromboembolism (VTE), renal papillary necrosis, and exercise-induced rhabdomyolysis recorded.

It is an extremely uncommon event, with only seven to eight cases of AVN in sickle cell trait having been documented in the literature [9]. Any individuals who complain of hip discomfort must be carefully monitored by doctors. There is always a chance that the patient has AVN as a result of sickle cell illness. Even if the patient simply carries the trait, there is still a theoretical possibility that they will develop AVN. To preserve what we can of the patient's anatomical and structural integrity of the bone, we must be prepared to have a lower threshold for thinking about receiving conservative treatments as soon as possible.

To determine why sickle cell trait occasionally causes severe exacerbations, more in-depth research must be done. Although the RBCs do not appear to have HbS or HbSC on electrophoresis, there may be an underlying genetic or epigenetic component that allows them to undergo shape modification. Future assessments of younger patients who show similarly to the one described above will be difficult because of the need to take extra precautions to prevent them from needing multiple hip replacements. Given the importance

of the case's economic and quality of life implications, prompt treatment is required.

Abbreviations

AVN: Avascular necrosis; SCT: Sickle cell trait; SLE: Systemic lupus erythematosus

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