Bilateral Avascular Necrosis of Femoral Head in Sickle Cell Trait: A Case Report

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**Abstract:**
Background: Sickle cell trait is usually an asymptomatic presentation of a patient with a slightly different hemoglobin molecule structure other than normal. It is similar to sickle cell disease, in which a person’s hemoglobin is mutated which causes their red blood cells to change their normal shape to a shape of the sickle in certain environmental and internal states. This causes red blood cells to adhere to the walls and occlude the lumen of the arteries in which they travel. It is leading to downstream effects secondary to ischemia. Sickle cell trait does not have these ischemic effects, usually.

Case presentation: In this case, a 42 years male patient presents to the orthopedic OPD with severe right hip pain and difficulty in walking. The patient is a known case of Sickle cell trait. His past history includes THR done on LT Hip joint 3 years back. The pain has lasted for several months and has not improved with NSAID. There is severe pain with internal and external rotation of the LT hip. The neurovascular of the lower extremities are intact bilaterally. MRI of the Right Hip shows stage IV avascular necrosis of the femoral head, while X-rays of the HIP are suggestive of Arthritis of Right hip with periarticular osteoporosis. The patient underwent THR of the Left Hip as well as starting bisphosphonates. The patient improved temporarily but regressed shortly thereafter. His avascular necrosis worsened radiographically over the next several months. At this point, the only option would be to do a total hip arthroplasty on the Right side, but the patient may need several more throughout his life as far as the age of the patient and the lifespan of the artificial replacement is concerned.

Conclusion: 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.
INTRODUCTION:
Avascular necrosis (AVN), is also known as osteonecrosis. Avascular necrosis is a disease that results from the temporary or permanent loss of blood supply to the bone. When blood supply is cut off, and ischemia of bone tissue has taken place, leading to infarction, ultimately the bone tissue dies and the bone collapses. If avascular necrosis happens near a joint, the joint surfaces may get involved. This condition may happen in any bone. This can result from trauma such as femoral neck fracture, where there occur ischemic consequences because of the compromised blood flow to the femoral head. Most living tissues need oxygen, without which there is inefficient metabolic functioning. Once infarction takes place, oxidative phosphorylation cannot take place and necrosis ensues.

More than 20,000 people each year enter hospitals for treatment of osteonecrosis of the hip. In many cases, both hips are affected by the disease. The prevalence of AVN is around 20,000–30,000 new diagnoses per year. There are a number of atraumatic associated etiologies, such as excessive alcohol, glucocorticoid use, Sickle Cell anemia, systemic lupus erythematosus (SLE), radiation therapy, and coagulopathy such as factor V Leiden mutation. [1–3]. AVN is multifactorial but can begin with interruption of blood and oxygen supply to vasculature in and around the bone. It progresses to trabecular thinning (seen in osteoporosis also) and it results in the collapse of the bone. In the case of sickle cell disease, this infarction results from occlusion of the vasculature by red blood cells (RBCs) which have changed their form from biconcave or round to crescent or sickle shape and flow less smoothly in the blood vessels. Their shape allows them to adhere to other RBCs as well as the endothelial walls, worsening vaso-occlusion. This leads to occlusion of bone marrow, ischemia, and progression to AVN. It is common in sickle cell disease. As much as 50% of sickle cell patients can develop AVN by the time they reach the age of 35. However, it is very rare in sickle cell trait (SCT), a much milder form of sickle cell disease in which patients are usually asymptomatic. This case report intends to focus on a rarely reported instance of AVN of Bilateral Femoral Head with Arthritis of the both hip joints in case of sickle cell trait. The dilemma is encountered during their treatment because of the age of the patient.

Case presentation
The patient is a 42-year-old Indian male, known case of sickle cell trait, with a past history of THR done 3 years back on the Left Hip joint. The patient presents with complaints of Right hip pain and difficulty in walking in the orthopedic OPD. He has sickle cell trait-like his father. Throughout his so, he has not had any acute episode of sickle cell-related symptoms like severe, acute chest pain, abdominal pain, and joint pain other than the hip joint. However, he states about 2 years prior, he had left hip pain. He went to his primary care provider, who gave him NSAID. These helped for a few weeks. His feet became swollen during this time, prompting him to visit MGIMS Sewagram(Wardha, Maharashtra, India), where he was operated for THR 3 years back on the Left side. He had severe groin pain with internal and external rotation of the hip joint. The range of movement at the Right Hip joint is as Flexion 10-45, Abduction 10-20, Extention, Internal rotation, External rotation, and adduction not possible. He had a 1cm true shortening on the RT side. X-rays were done of his Pelvis with both Hips AP and Lateral view. It showed an area of necrosis suggestive of stage 4 AVN in the head of the right femur. An MRI was not done as the diagnosis was confirmed on X-ray.
Anteroposterior radiographyc view of the pelvis shows flattening of the outer portion of the right femoral head from avascular necrosis, with adjacent joint space narrowing, juxta-articular sclerosis, and osteophytes representing degenerative Arthritis (stage IV). He was prescribed bisphosphonates in view of prevention of AVN on another side when he came 2 years back for follow-up of his THR performed 3 years back on the left side. He was told to walk with a non weight bearing on the right leg. He had not felt pain on the Right side at that time. range of movements was pain-free and within normal limits. His hip x rays were normal at that time. He was then advised to begin physical therapy soon thereafter. However, after eight months of this visit, he presented to Ortho OPD with stating new-onset pain on the Right Hip. He had weight-bearing but the pain had started again in the right hip and groin. There were also new x-ray findings (AP Pelvis and Frog Hip views), which showed a serpiginous line that was consistent with the progression of his AVN with no collapse of the femoral head. Even after bisphosphonates, the patient's AVN had now grown to involve the majority of the head of the femur. MRI showed that the head of the femur was beginning to flatten as well as loss of volume and bone marrow edema, which is seen in Fig. 2. Further progression of her AVN can be seen with follow up x-rays at a later appointment of 6 months.
This coronal T1-weighted image shows the progression of AVN with necrosis, flattening of the right femoral head, and post decompression evidence in the femoral neck. There is also edema, which corresponds to the increasing pain the patient was experiencing at this time. There are obvious differences between the THR done and right femoral heads. AP view of the right femoral head shows areas of hyperlucency and surrounding sclerosis, as well subtle changes in the shape of the articular surface. The necrosis also spreads into the acetabulum. This x-ray was done just before the patient was referred for THR. At this point, there were no other conservative options available for this patient. Patient’s age was the only dilemma. Osteotomy is sometimes a treatment for AVN, but given the area of necrosis of this patient’s femoral head, this procedure is not indicated. The plan for this patient is THR as it would be the only definitive treatment for this patient. Because this patient is was comparatively young. At his age, it would be challenging to do a THR, given that he would most likely have to undergo the same procedure several more times throughout his life. Hip replacements have an average life of 15–20 years, so the prospect of conserving as much of the joint as possible and not undergoing THA at such a young age is paramount. It would be devastating for the patient to have 4–6 hip replacements in his life as he had bilateral THR. As there was no alternative patient was posted for THR. UAAP with posterior Moore's approach, an incision was taken, Femoral head excised, Acetabulum and femoral canal prepared, THR done using acetabular shell - 50 mm, modular head component - 32 by 6 mm, femoral stem - 1.4 by 140 mm,2 dome screw - 6.5 by 20 mm and 6.5 by 30 mm, wound closed in layers under sterile dressing done. The procedure was uneventful intraoperative and postoperative. Pathological investigations were done preoperatively just for confirmation of the diagnosis of SST.His Sickling Test was Positive.HB Electrophoresis was done and AS pattern was found which was indicating a Sickle Cell Trait.

DISCUSSION AND CONCLUSIONS
The normal adult hemoglobin has two alpha and two beta chains. Hemoglobin that has the ability to sickle is referred to as sickle hemoglobin (HbS). Sickle hemoglobin is the result of a single-point mutation from Glutamine to Valine on the beta-globin chain. If an individual is homozygous, it means they inherit two mutated beta-globin chains and they will have sickle cell disease [4]. These abnormal beta chains cause RBCs to become fragile and change shape. It results in conditions of hypoxia, acidity, and dehydration at the tissue level. Repeated vaso-occlusion by sickled RBCs will eventually cause ischemia, infarction, edema, and end-organ damage of bone [5, 6] which ultimately leads to AVN. If an individual is heterozygous, they only inherit one mutated beta-globin chain and will have a sickle cell trait. It usually takes 50% of HbS for cells to have the ability to sickle. Sickle cell trait individuals have around 40% HbS and therefore are usually asymptomatic [7]. However there are certain instances of exertional rhabdomyolysis, exercise-induced sudden death, renal papillary necrosis, venous thromboembolism (VTE), and fetal demise are reported. There have only very few cases of AVN in sickle cell trait reported in the literature. Hence it is an extremely rare occurrence of bilateral AVN of the Hip joint. [8] There are always the high chances of the patient having AVN secondary to sickle cell disease. However, there is still theoretically a chance of the patient having AVN even if they only have the sickle cell trait. We must be ready to have a lower threshold to think about getting conservative treatments. So as soon as possible, to salvage what we can have the patient's anatomical, structural integrity of the bone. More detailed studies need to be undergone to assess the reason why sickle cell trait sometimes has severe exacerbations. There may be an underlying genetic or epigenetic component that allows the RBCs to undergo a change of shape even though on electrophoresis they do not appear to have HbS or HbSC. It will be challenging down the road to assess younger patients who present like the aforementioned one. It is because their situation
exacts special care so that they may not have to undergo multiple hip replacements in the future. The quality of life and economic aspects of this case is very important, so care must be given to treat quickly. A major issue in managing sickle cell disease and sickle cell trait patients is their prevalence is more in tribal and hilly populations. Poverty, illiteracy, and difficulty in approach are the major hurdles in early diagnosis and timely treatment. Hopefully with newer technology and pharmacologic therapies, along with conservative approaches like non-weight-bearing osteophos, patients who develop AVN will have a plan in place to salvage bone and quality of life.

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Author’s contributions
Dr. Prasad Bhandari worked on this case report and did all of the research groundwork as well as typing and other statistical work.

Abbreviations
AVN- Avascular necrosis,
SCT- Sickle cell trait
SLE- Systemic lupus erythematosus,
THR- Total Hip Replacement.

Notes
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