Core Decompression Surgery for Avascular Necrosis Can Delay Femoral Head Collapse in Patients with Sickle Cell Disease: A Case Report

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Introduction

Avascular necrosis (AVN) of the femoral head is a common orthopaedic complication in patients with sickle cell disease (SCD). By age 35, approximately half of patients homozygous for the sickle cell gene mutation have AVN, with involvement of both hips in 40-91% of patients.[1] Without early intervention, the rate of femoral head collapse is high and often necessitates total hip arthroplasty (THA).[1]

Various treatment options exist for AVN, including physical therapy, pharmacotherapies, core decompression, and arthroplasty.[2]the possibility has been raised that bone marrow containing osteogenic precursors implanted into a necrotic lesion of the femoral head may be of benefit in the treatment of this condition. For this reason, we studied the implantation of autologous bone-marrow mononuclear cells in a necrotic lesion of the femoral head to determine the effect on the clinical symptoms and the stage and volume of osteonecrosis.METHODS:We studied thirteen patients (eighteen hips However, outcomes of most treatments have been varied,[3]and core decompression is a relatively safe and possibly effective option. To optimize core decompression further, we have tested a new technique involving thorough decompression of the osteonecrotic zone under endoscopic visualization (TDEV and as a result, there is no consensus on the best method to effectively prevent collapse and postpone the need for THA. Despite the lack of consensus, surgical core decompression has shown potential in the management of these cases. Core decompression with thorough debridement under endoscopic visualization followed by bone grafting and fixation with a nail/plate device produced encouraging results in a cohort of pediatric patients after a mean follow-up of 28 months, particularly for the treatment of lesions graded lower than Steinberg Stage IIIB.[3]and core decompression is a relatively safe and possibly effective option. To optimize core decompression further, we have tested a new technique involving thorough decompression of the osteonecrotic zone under endoscopic visualization (TDEV Meanwhile, a recent modification of core decompression involving autologous bone marrow mesenchymal stem transfer and multiple smaller drill holes resulted in statistically significant improvement in pain and hip survival compared to standard decompression in patients with Association Research Circulation Osseous (ARCO) Stage I/II hips in a recent randomized control trial.[4]

In this case report, we present two patients with SCD whose disease course was complicated by AVN for which they subsequently underwent decompression procedures. One patient underwent multiple core drilling decompression with iliac crest bone marrow grafting, and the other underwent the standard core single bore drilling and iliac crest bone grafting with pin stabilization. Both patients had significant improvement in symptoms and radiographic evidence of at least arrest of femoral head collapse at three years of follow-up.

Case Report

Case 1

A 12-year-old African-American male with hemoglobin SC variant SCD presented to our institution with a one-month history of progressive left-sided hip pain without any history of trauma or steroid use. On physical exam, he had normal gait but exhibited a mildly stiff left hip. Left hip range of motion was particularly notable for limited internal rotation in flexion at 20° and painful abduction to 35°. This was in contrast to a painless right hip with range of motion within normal limits.

Antero-posterior (AP) and frog lateral images of the left hip revealed femoral head flattening, confirmed by subsequent magnetic resonance imaging (MRI) (Figure 1) as osteonecrosis of the left hip with evidence of subchondral fracture, graded as Steinberg IIIb. His right hip was normal. Given advanced osteonecrosis on imaging and severity of his hip pain stiffness, the decision was made for a multiple drill hole core decompression and grafting with iliac crest bone marrow mesenchymal stem cells. The patient underwent the procedure as previously described by Gangji *et al*[2] to try to stimulate growth and healing of his osteonecrotic lesion.

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Figure 1. Preoperative MRI of left hip showing avascular necrosis in a 12-year-old male.

His immediate post-operative course was unremarkable. The patient began physical therapy at six weeks and continued to have improved range of motion. At twelve weeks, he had improved abduction to 45°. Index radiographs at eight and twelve-week intervals continued to show stable osteonecrosis. Follow-up Xrays (Figure 2) have shown a halt in progression of femoral head irregularities and improvement of the femoral head contour.

Case 2

A 9-year-old African-American female with history of SCD and asthma presented to our institution with left hip pain. At initial presentation, she had a three-month history of intermittent left hip pain and a progressive limp on the left side. There was no inciting trauma or injury. She had a history of sickle cell crises requiring inpatient admissions despite a medication regimen that included hydroxyurea and prednisone. Her initial exam was significant for a severely stiff left hip with flexion limited to 70°, abduction to 25°, external rotation of 5°, and internal rotation of 5°. This was in contrast to an asymptomatic right hip.

AP and frog lateral images revealed advanced osteonecrosis of the left hip (Figure 3a) confirmed by MRI, which further showed evidence of subchondral fracture. The left hip was graded as Steinberg IIIC. Early signs of osteonecrosis with bone marrow edema were evident in her right hip as well, although it was clinically stable.

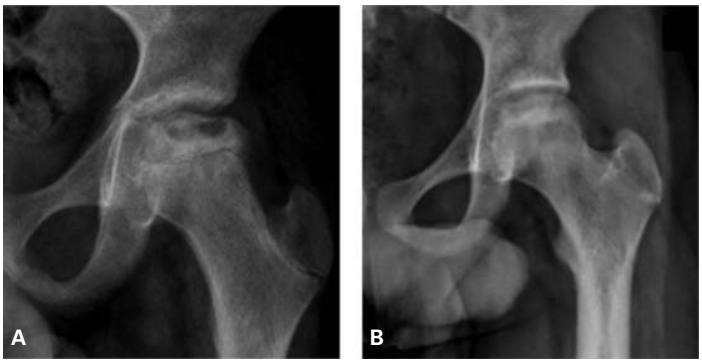

Figure 2. (A) 3 weeks postoperative AP image after decompression of left hip, and (B) follow-up AP of left hip 3 years after surgery showing minimal changes in femoral head contour.

Given the patient's age, degree of femoral head collapse and decreasing hip mobility, she was treated with core decompression, iliac crest bone grafting, and stabilization with a compression hip screw as previously described by Wells *et al.*[3] She did very well in the immediate postoperative period and had significant improvements in hip stiffness. A 0.5cm leg length discrepancy was noted at her 1.5-year follow-up visit, which was addressed with a shoe lift. At her most recent 5-year post-operative visit, a slight improvement in femoral head contour was noted (Figure 3b). Her physical exam at this visit demonstrated increased hip motion with improvement in flexion to 95°, abduction to 50°, adduction to 30°, external rotation to 40°, and internal rotation to 30°. There was no pain with range of motion.

Discussion

Treatment of advanced AVN with subchondral collapse is very challenging with various treatment options showing mixed results. We present two cases of adolescent African-American patients with advanced AVN, both of the left femoral head, who underwent successful decompression procedures: one with multiple drill hole decompression and iliac bone marrow grafting, and the other with a single drill core decompression, bone grafting, and stabilization with hip screw. The first patient was doing very well at his most recent 3-year follow up, and the latter was followed for five years with excellent improvement. Both have full, painless range of motion of both hips, and their radiographs show stable disease with improvement of femoral head contour.

Core decompression has become the most common way of preventing or prolonging the time to femoral head collapse in AVN, yet very few rigorous studies have validated long-term outcomes.[5] More specifically, for SCD patients, the studies that do exist demonstrate very conflicting results. Neumayr *et al*, in a prospective study, reported equal efficacy of physical therapy alone compared to decompression surgery, while Mukisi-Mukaza *et al*, in a more recent prospective study, demonstrated delayed need for arthroplasty with early decompression.[1,6] Both studies had a three-year follow-

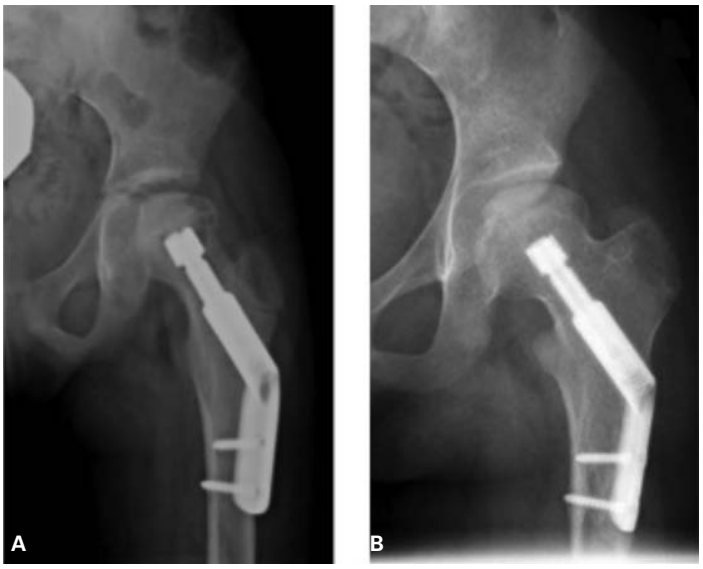

Figure 3. (A) Postoperative AP image of left hip in a 9 year-old female after core decompression, iliac crest bone grafting, and stabilization with a compression hip screw for AVN, and (B) AP Xray at 5 year follow-up visit showing no progression of AVN.

up period. In our own experience of treating AVN in SCD patients, limited weight bearing, physical therapy, and other non-operative treatments do not compare favorably to surgery.

Complicating the treatment picture are the numerous available core decompression procedure modifications, which include standard core decompression, multiple smaller drillings, and grafting with either autologous bone or mesenchymal bone marrow stem cells. While various studies of these procedure modifications have shown promising results in patients with AVN, no specific reports have addressed therapeutic benefits specifically in patients with SCD. Although this case report is only of two patients, the results are very encouraging. Both procedures were successful in preventing catastrophic collapse of the femoral head, improving gait, restoring motion, and alleviating pain in both patients. Both patients have radiographic evidence showing halting of disease progression for at least three years with no further femoral head collapse. Both standard core decompression with iliac crest bone grafting and pin stabilization, and multiple core decompression with iliac crest bone marrow grafting appear to be effective in prolonging femoral head collapse in SCD patients.

The few studies that compared multiple drill hole core decompression to conventional methods all claim improved outcomes, decreased fracture rate, lower rates of collapse, and longer time before collapse with the multiple drill hole method,[4,7,8] with Kim and associates demonstrating that the multiple drill hole technique for SCD had a lower rate of collapse (14.3%) than the conventional method (45%).[8] Bone marrow instillation in several small studies appears to be superior when added to core decompression.[2,4] To the best of our knowledge, no studies have compared standard bone grafting to marrow grafting. Thus it appears that, individually, core decompression with multiple drill holes and bone marrow mesenchymal stem cell graft confers a hip survival advantage to affected patients. Perhaps when both techniques are used in concert, greater benefits may be expected. Further studies are needed to compare methods of treating osteonecrosis in the sickle cell population, especially regarding bone marrow mesenchymal stem cell versus whole bone graft, as well as outcomes of standard decompression compared to multiple drill hole decompression.

Conclusion

Patients treated with either multiple drill hole decompression and iliac bone marrow grafting or single drill core decompression, bone grafting, and stabilization with a hip screw fare better than patients treated non-operatively. We feel that earlier disease detection will result in improved outcomes and longer preservation of the diseased femoral head. We recommend and encourage routine radiographic screening in patients with sickle cell anemia who are at high risk for osteonecrosis.

References

1. Neumayr LD, Aguilar C, Earles, AN, et al. Physical therapy alone compared with core decompression and physical therapy for femoral head osteonecrosis in sickle cell disease. Results of a multicenter study at a mean of three years after treatment. *J Bone Joint Surg Am* 88, 2573–2582 (2006).

2. Gangji V, Hauzeur, JP. Treatment of osteonecrosis of the femoral head with implantation of autologous bone-marrow cells. *J Bone Joint Surg* 87, 106–112 (2005).

3. Wells L, Hosalkar HS, Crawford, EA, et al. Thorough debridement under endoscopic visualization with bone grafting and stabilization for femoral head osteonecrosis in children. *J Pediatr Orthop* 29, 319–326 (2009).

4. Sen RK, Tripathy SK, Aggarwal S, et al. Early results of core decompression and autologous bone marrow mononuclear cells instillation in femoral head osteonecrosis. *J Arthroplasty* 27, 679–686 (2012).

5. Rajagopal M, Balch Samora J, Ellis TJ. Efficacy of core decompression as treatment for osteonecrosis of the hip: a systematic review. *Hip Int* 22, 489–493 (2012).

6. Mukisi-Mukaza M, Manicom O, Alexis C, et al. Treatment of sickle cell disease's hip necrosis by core decompression: A prospective case-control study. *Orthop Traumatol Surg Res* 95, 498–504 (2009).

7. Mont, MA, Zywiel MG, Marker DR, et al. The natural history of untreated asymptomatic osteonecrosis of the femoral head A Systematic literature review. *J Bone Joint Surg* 92, 2165–2170 (2010).

8. Kim, SY, Kim DH, Park IH, et al. Multiple drilling compared with standard core decompression for the treatment of osteonecrosis of the femoral head. *J Bone Joint Surg Br* 86-B, 149–149 (2004).