Scope assisted ankle arthrodesis in a young male with hemophilic arthritis: A case study

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ABSTRACT

Hemophilia is a congenital bleeding disorder caused by the absence or decrease of clotting factor VIII (Hemophilia A) or XI (Hemophilia B). When severe, this X-linked disease results in intra-articular bleeding which can progress to joint destruction. This is known to be the most common complication of severe uncontrolled hemophilia. We present a case of arthroscopic assisted ankle arthrodesis in a young male presenting with advanced ankle hemophilic arthritis. Our case report presents a unique pathology and the successful 1-year post-operative outcome that can be accomplished through surgical intervention and a multidisciplinary treatment approach.

Introduction

Arthritis is a debilitating condition that negatively affects an individual’s ability to carry out their activities of daily living. In the foot and ankle, there are numerous joints that are susceptible to disease. Osteoarthritis is a term many have become familiar with, as it is the most common form of arthritis encountered. Hemophilic arthritis on the other hand is a condition that is not as frequently seen or discussed. Hemophilia is a congenital bleeding disorder caused by the absence or decrease of clotting factor VIII (Hemophilia A) or XI (Hemophilia B). When severe, this X-linked disease commonly results in intra-articular bleeding which can progress to joint destruction.1-4 This is widely known to be the most common complication of severe uncontrolled hemophilia.

The presence of intra-articular blood has been shown to directly cause cartilage degeneration and loss. In the case of hemophilia, recurrent bleeding into the joint may occur leading to synovial tissue proliferation and neoangiogenesis. This in turn increases the joint’s susceptibility to bleeding and further perpetuates the process.4-5 With hemarthrosis, iron is released into the synovium through the breakdown of hemoglobin and promotes an inflammatory response. This subsequently leads to the progressive destruction of articular cartilage and bone.6-8 Ultimately, the changes seen in hemophilic arthropathy are manifested with symptomatic pain, stiffness, and articular deformity.

There are various treatments that have been described for patients suffering from hemophilic arthritis. While the use of clotting factor concentrations (CFCs) may reduce the risk of hemorrhaxis, the risk of bleeding cannot be completely avoided.9 Arthroscopic or open synoviectomies have been described to remove the pathogenic synovium. Chemical or radiosotopic synoviorthesis is an option for less severe cases but the treatment of choice remains to be joint fusion or arthrodesis for advanced stages of arthropathy.10 In the present study, we present a case of arthroscopic assisted ankle arthrodesis in a young male presenting with advanced ankle hemophilic arthritis.

Case report

A 25-year-old male presented to clinic with an 8-year history of worsening left ankle arthritis and pain. The patient was seen in a private practice orthopedic center from December 2019 until January 2021. He had a known history of Hemophilia type A with intra-articular involvement of the left ankle joint which subsequently led to traumatic arthritis secondary to this condition. At the time of presentation, the patient admitted that he had been medically managed for his condition which had slowed the progression of his musculoskeletal pathology. Unfortunately, due to the prolonged nature of his condition, the patient still reported debilitating ankle pain. The patient stated the symptoms had progressed to the point where he was no longer able to participate in any form of exercise and that standing at work or on uneven surfaces have begun to cause intolerable discomfort daily.

Conservative treatments for the patient were explored and ultimately exhausted. This treatment regimen included non-weightbearing...
immobilization, physical therapy, and AFO ankle bracing. Continued follow-up without significant improvement of the patient’s condition warranted advanced imaging via MRI. A discussion of surgical intervention to alleviate the symptomatic ankle was also completed following failed conservative measures and further understanding of the end stage ankle arthritis with the acquisition of advanced imaging.

**Physical exam**

Musculoskeletal exam revealed gross swelling noted to the left ankle region. Point tenderness was elicited throughout the joint line. There is limited range of motion available through the tibiotalar joint with accompanying crepitus. Subtalar joint evaluation also revealed stiffness without pain. The intrinsic and extrinsic extensors and flexors of the left lower extremity are within normal limits. The transverse tarsal joints demonstrate full motion without crepitus or pain. No significant cavus or planus deformity was noted.

Neurovascular status was intact with palpable dorsalis pedis and posterior tibial arterial pulses. Capillary fill time to the distal forefoot and digits were demonstrated. There were no signs of deep vein thrombosis, indurated vasculature, or exertional compartment syndrome. Protective sensations to the lower extremity were present and normal tendon reflexes were also exhibited.

**Imaging**

**Surgical intervention**

Arthroscopic assisted ankle arthrodesis with calcaneal autograft was performed by the senior author. Calcaneal autograft was acquired through a small linear incision along the lateral wall of the calcaneus, a power graft harvester was introduced into the medullary bone to procure the graft. The left lower extremity was then placed into a non-invasive Gould Ankle distraction system and the standard anteromedial and anterolateral ankle scope portals were established. A scope and shaver were utilized to extensively debride all visualized hemorrhagic synovium. The shaver was then exchanged for a burr following debridement to denude the articular cartilage from both the distal tibia and talar dome. The cartilage removal was supplemented utilizing curettage and subchondral preparation. Following adequate preparation of the joint margins for fusion, the ankle joint was filled with the calcaneal autograft and rhPDGF-BB. A 3-screw construct of two 6.5 mm partially threaded cannulated screws and a 6.5 mm fully thread screw was utilized. Figs. 1–3.

**Discussion**

The concern with intra-articular bleeding or hemarthrosis are the long-term effects and sequela that it has on the affected joint. Recent literature has shown that hemophilic arthropathy results in intra-articular inflammation and angiogenesis, resulting in articular cartilage degeneration. When blood is introduced into a synovial joint, hemoglobin breaks down and releases iron – which induces a chronic inflammatory process. This chronic inflammatory process thus leads to a progressive synovial pannus growth and subsequent articular cartilage damage.

The pathophysiology of hemophilic arthropathy is similar to rheumatoid arthritis (RA) and osteoarthritis (OA). There are both inflammatory and degenerative mechanisms that contributes to the disease process. Some authors believe that the initial pathogenic process involves articular cartilage damage due to the iron-catalyzed formation of destructive oxygen metabolites. These toxic metabolites then target chondrocytes and results in apoptosis, leading to synovial inflammatory changes as a secondary process.

The exact mechanism by which bleeding leads to subchondral bone destruction is not completely understood. Though, it is known that a critical regulator of bone biology is the osteoprotegerin (OPG), receptor activator of nuclear factor kB (RANK), and RANK ligand (RANKL). RANKL is a transmembrane ligand found on osteoblasts. It is synthesized by lymphocytes and synovial cells and is thought to induce osteoclastogenesis. RANK is expressed on the cell surfaces of osteoclast precursors and when bound to RANKL, induces osteoclast differentiation – thus RANKL induces bone resorption. OPG in this microenvironment can be thought of as a decoy as it competes for the binding of RANKL to RANK – thus OPG inhibits bone resorption. RANK and RANKL have been found to be strongly expressed in the synovium with the expression of OPG to be dramatically reduced in patients with hemophilic arthropathy. This important finding demonstrates how the synovial tissue of

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**Fig. 1.** Preoperative radiographic imaging. Anteroposterior and lateral views of the left lower ankle and foot with visualized significant end stage ankle arthritis with joint space narrowing and anterior tibiotalar joint osteophyte formation.
hemophilic patients favor a shift towards osteoclastic activity and thus bone resorption.

In a recent study comparing the differences between Hemophilia type A and B found that patients with type A may have more severe arthropathy than type B. Melchiorre and colleagues in 2016 found that patients with hemophilia type A had significantly decreased World Federation of Hemophilia (WFH) orthopedic joint and ultrasound scores compared with hemophilia type B patients. The authors found that most of the ultrasound findings (joint effusion, synovial hypertrophy, hemosiderin deposition, bone and cartilage remodeling) were observed in the hemophilia type A cohort. They also found that a greater percentage of hemarthroses occurred in the hemophilia type A group compared to type B.12

In cases of severe ankle hemophilic arthritis, total ankle replacement or arthrodesis may be considered. Barg et al in 2010 reported the outcomes of 10 total ankle replacements in 8 patients. Their patients had a mean age of 43 years and all had type A hemophilia. Utilizing the HINTEGRA prosthesis, they reported significant improvements in SF-36 and AOFAS scores at a mean 5.6-year follow-up. Four patients became pain free and all patients were satisfied with the results.18 Asencio and colleagues then followed up this study in 2014 reporting on 32 total ankle replacements in 21 patients. They also found that total ankle replacements in patients with arthropathy leads to significant improvements in AOFAS and function scores.19

**Conclusion**

This case report details the long-term sequela Hemophilia type A and its deleterious effect on the tibiotalar joint. In patients with hemophilia, regular replacement therapy with clotting factor concentrates (prophylaxis) is effective in preventing recurrent bleeding episodes into joints and muscles. However, despite this success, intra-articular and intramuscular bleeding is still a major clinical manifestation of the disease. Bleeding most commonly occurs in the knees, elbows, and ankles, and is often evident from early childhood. The pathogenesis of hemophilic arthropathy is multifactorial, with changes occurring in the synovium, bone, cartilage, and blood vessels. Our case report presents a unique pathology and the successful outcome that can be accomplished through surgical intervention and a multidisciplinary treatment approach.
Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Patient Informed Consent Statement

Per HIPAA guidelines, patient authorization prior to submission is not needed, as the case had been de-identified and stripped of the 18 HIPAA identifiers.

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