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Complicated Total Knee Arthroplasty in a Patient with Klippel-Trenaunay-Weber Syndrome:

A Case Report

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Abstract

Case—A 66-year-old Caucasian man with Klippel-Trenaunay-Weber syndrome (KTWS) presented with chronic changes related to the KTWS, along with worsening pain and motion associated with residual damage from an episode of spontaneous septic arthritis that occurred 1 year prior. He underwent total joint arthroplasty with a rotating hinged knee implant.

Conclusion—Arthroplasty is a treatment option for patients with KTWS; however, there are risks that must be considered. This case report outlines the management of a patient with KTWS and a history of septic arthritis.

Klippel-Trenaunay-Weber syndrome (KTWS) is a rare congenital vascular disease associated with soft-tissue and osseous hypertrophy, cutaneous hemangiomas, and vascular malformations involving the lymphatic, venous, and arterial vessels^{1–11}. Commonly used diagnostic criteria include varicose veins, cutaneous hemangiomas, and hypertrophy of soft tissue and bone^{1–4,6–10}. At least 2 of these features must be present for the diagnosis of KTWS¹. Our patient met all 3 of these criteria. KTWS most often presents at birth or in early childhood, and affects the lower extremities in approximately 95% of patients^{1,3}. Limb hypertrophy generally develops unilaterally but can manifest bilaterally^{4,5}. Vascular irregularities present not only on the limbs, but also in the oral cavity and on the head, the neck, the trunk, and/or the buttocks⁴. There is no sex or racial predilection for this syndrome^{3,7}.

Vascular malformations can lead to secondary orthopaedic symptoms, such as bone and soft-tissue hypertrophy⁸. Soft tissue swells because of blood-flow stasis; the swollen soft tissue stimulates the periosteum, leading to the compression of periosteal blood vessels and lymphatic vessels, causing blood-supply disorders in the bones. Hyperactivity of periosteal osteoblasts results from chronic stimulation, and ultimately, bone hypertrophy occurs^{3,6}. Bone hypertrophy can lead to numerous other symptoms, such as limb-length inequality, secondary pelvic tilt, scoliosis, gait abnormalities, and joint contractures. Because of its effect on bone and soft tissue, other orthopaedic anomalies are seen in patients with KTWS, including macrodactylia, polydactyly, syndactyly, and hip dislocation³. Additionally,

degenerative arthropathy can be a consequence of bone and soft-tissue hypertrophy, chronic inflammation, vascular malformations, and, as in our patient, a history of underlying prior septic arthritis^{2,6}.

KTWS does not have a cure; therefore, treatment consists of managing the symptoms of pain and immobility secondary to the disease process. There are surgical and nonsurgical interventions to treat symptoms. Nonsurgical options include sclerotherapy, laser therapy, and compression therapy, as well as medical management, which includes anticoagulation, pain medications, and anti-inflammatory drugs^{2,6,7}. Surgical options include, but are not limited to, surgical vein ligation, vein stripping, vein resection, epiphysiodesis, tibial osteotomy, arthroplasty, and debulking. Additionally, affected limbs occasionally are amputated to lower cardiac output and myocardial stress in order to prevent heart failure^{5,8}.

Knee arthrosis and disparity in leg length are common orthopaedic effects of KTWS. KTWS-associated arthropathy is commonly monoarticular because the disease is usually localized to 1 extremity. Our patient presented with degenerative arthropathy of the knee, which is the most commonly affected joint². A literature review revealed limited reports of arthroplasty in the setting of KTWS; 2 reports involved the knee and 1 involved the hip^{2,9,10}. This case report highlights the challenges in treating a patient with KTWS and end-stage knee arthritis, and provides insight into managing a prior history of septic arthritis.

The patient was informed that data concerning the case would be submitted for publication, and he provided consent.

Case Report

A 66-year-old Caucasian man with KTWS presented with a history of worsening pain and limited range of motion in the right knee. The patient had chronic changes related to KTWS as well as worsening pain and motion related to spontaneous septic arthritis that had occurred 1 year prior. The episode was attributed to *Enterobacter*, an unusual microbial etiology, and led to the subsequent use of trimethoprim-sulfamethoxazole for suppression. The patient's height was 170.2 cm, his weight was 74.6 kg, and he had a body mass index of 25.75 kg/m². The knee had a 23° arc of motion (58° to 81°). He walked with the assistance of Canadian crutches, with the toes of the affected extremity barely touching the floor and a flexed knee. He rated the pain as 10 of 10, which worsened with weight-bearing. The patient also had a remote history of recurrent infections in the foot and ankle, but specifics, including culture results and details surrounding an irrigation and debridement of the ankle, were not available; therefore, it was unclear if the knee and ankle infections were related. The medical history was noteworthy for 2 episodes of deep vein thrombosis of the lower extremities and chronic obstructive pulmonary disease; the patient was a former smoker. The surgical history included multiple vascular procedures, including iliac vein stenting for May-Thurner syndrome and a bone debulking surgery of the right knee when he was a child. He had undergone an open irrigation and debridement of the right knee via a medial parapatellar approach following the episode of spontaneous septic arthritis described above. He was on chronic anticoagulation therapy with warfarin, and anti-platelet therapy with clopidogrel.

The right leg was visibly larger in diameter than the left. Several vascular malformations were palpable, and skin changes were consistent with venous stasis (Fig. 1). The range of motion was limited, and the patient had substantial instability with valgus stress, which was concerning for an incompetent medial collateral ligament. Although difficult to assess because of flexion contracture, the right leg was shorter by about 4 cm. While the affected extremity typically is longer, the history of debulking surgery in childhood may have contributed to the leg-length discrepancy. The patient had normal sensation and +2 palpable dorsalis pedis and posterior tibial pulses. Radiographs revealed extensive loss of joint space with destruction of the articular surfaces (Figs. 2-A and 2-B). Additionally, marked osseous hypertrophy and soft-tissue changes were present.

After multiple visits and failed nonoperative treatments (including bracing, crutches, and medications), we discussed treatment options with the patient; possible considerations included knee fusion, resection arthroplasty (Girdlestone procedure), elective amputation, and total knee arthroplasty. The patient wished to undergo arthroplasty with the understanding that he was at high risk for failed total knee arthroplasty and could require an amputation. Considering the history of septic arthritis, ruling out infection was important. Suppressant antibiotics were discontinued for 6 weeks before testing so that an ongoing infection could be detected. The C-reactive protein level was 0.30 mg/dL (normal, <0.80 mg/dL), and the erythrocyte sedimentation rate was 18 mm/hr (normal, 0 to 20 mm/hr). A joint aspiration was attempted, but failed to produce adequate fluid for testing, so the patient was sent for a computed tomography (CT)-guided biopsy and aspiration. These culture samples were negative for infection. The synovial fluid analysis demonstrated 3,333 white blood cells/ μ L with 71% neutrophils.

A hinged total knee arthroplasty was planned. In addition to medical and cardiac clearance, we obtained a preoperative consultation with the vascular surgery and plastic surgery departments. This workup included a CT angiogram, which demonstrated patent iliac veins and 2 areas of high-flow arteriovenous malformations (AVMs) (Fig. 2-C).

General endotracheal anesthesia, an injection of intrathecal duramorph, and a tourniquet were used. A standard medial parapatellar approach was utilized; however, given the preoperative stiffness and the substantial scarring, a quadriceps snip was performed to allow better visualization. A wide synovectomy was performed to reestablish both the medial and lateral gutters. The medial collateral ligament was found to be incompetent, consistent with preoperative examinations; therefore, we decided to utilize a hinged knee implant. Several small vascular malformations that were encountered during dissection were cauterized using bipolar radiofrequency and saline solution without complication. Multiple vascular malformations were encountered within the joint space, the distal aspect of the femur, and the proximal aspect of the tibia. The tissues had a hemosiderin-laden appearance (Fig. 3), as might be seen in a patient with hemophilia. Three synovial samples were taken intraoperatively for frozen section histology; none showed signs of acute inflammation. Permanent pathology specimens were also taken; they demonstrated fibrous tissue with focal hemosiderin deposition and no evidence of malignancy. In total, 5 tissue culture samples were taken from the synovium, the femur, and the tibia. A rotating hinged knee implant was secured utilizing manufacturer-prepared antibiotic cement with gentamicin. Given the

success of hybrid fixation stems in the revision setting and the high risk of failure because of infection, the femoral and tibial stems were press-fit, although fully cemented stems could have been considered. The tourniquet was then released, and hemostasis was achieved with cautery prior to closure. Two deep drains were used along with topical tranexamic acid. The drains were clamped for a period of time and then released in the postoperative period per our standard protocol and those previously outlined in the literature¹².

The intraoperative cultures of the femur and the tibia were positive for *Propionibacterium*. This bacterium, different from the prior *Enterobacter* infection, were addressed with a 6-week course of intravenous vancomycin, which was based on consultation with the infectious disease department. The postoperative course was complicated by a non-ST elevated myocardial infarction. The patient required a transfusion of 2 units of packed red blood cells postoperatively; he was discharged on postoperative day 9.

At 2 months postoperatively, the patient presented with bloody drainage from the inferior aspect of the incision, which had developed 3 weeks after the cessation of intravenous vancomycin therapy. There was no erythema around the wound or pain with motion, which would suggest infection. The drainage was related to a superficial AVM, but considering the complicated history and high risk for recurrent infection, the patient underwent irrigation and debridement and polyethylene exchange as a precautionary measure. Preoperative synovial fluid analysis demonstrated 2,000 white blood cells/ μ L with 69% neutrophils. The intraoperative culture specimens remained negative, and no evidence of infection was identified. During the postoperative course, the knee was immobilized in extension, and an incisional negative-pressure system was used. He was restarted on anticoagulation therapy and a suppressive trimethoprim-sulfamethoxazole regimen, as prescribed by the infectious disease physician.

At 26 months after the total knee arthroplasty, radiographs demonstrated a well-fixed hinged implant (Figs. 4-A and 4-B). The range of motion was 0° to 90° (Video 1). He utilized crutches only when walking long distances and did not need to use them routinely. He also had begun using a shoe lift for the leg-length discrepancy since the flexion contracture had been corrected. He was taking amoxicillin as a suppressive antibiotic (directed primarily against the *Propionibacterium*). He had experienced no additional wound issues, and the pain and function were dramatically improved.

Discussion

Although not to be undertaken lightly, arthroplasty is an option to treat patients with KTWS who are substantially disabled because of orthopaedic manifestations of the syndrome. As with our patient, they are at high risk for wound complications, postoperative anemia, cardiovascular complications, and infections. These patients also have increased risk for deep vein thrombosis and pulmonary embolism^{10,11}. Because of the potential for intraoperative and postoperative complications, we feel that an extensive preoperative workup with the involvement of other subspecialists (vascular surgeons, infectious disease specialists, and cardiac specialists) is paramount, as outlined in prior reports^{2,10,11}.

In our patient, the preoperative CT angiogram identified 2 AVMs with high arterial flow. Although smaller AVMs were easily controlled using bipolar radiofrequency cauterization, we ensured that intraoperative consultations with the vascular surgeons were available. Given the patient's prior stenting and high risk for venous thromboembolism, he was bridged off warfarin with an injectable anticoagulant 5 days preoperatively.

Postoperatively, the patient had wound drainage over a superficial AVM. Aggressive treatment of this drainage with irrigation and debridement and polyethylene exchange was performed as a precautionary measure against the possibility of infection. The wound was subsequently managed with immobilization and negative-pressure therapy for 6 days. This prolonged immobilization did not affect the long-term range of motion; at the latest follow-up, the patient had a 90° arc of motion and was able to navigate most activities of daily living without difficulty.

After discussions with the infectious disease team, our patient was maintained on suppressive antibiotics, initially trimethoprim-sulfamethoxazole and then amoxicillin because he tolerated it better. While there are drawbacks to chronic antibiotic therapy, an improvement in infection-free survival has been demonstrated and was felt to benefit this patient^{13,14}.

In comparison with 2 similar cases in the literature of patients with congenital vascular syndromes undergoing total joint arthroplasty, our case demonstrates several notable differences and similarities^{2,9}. The differences between these 2 cases and our case include age, comorbidities, and implant type. At 66 years old, our patient was substantially older compared with the 35-year-old and 38-year-old patients in the literature. Our patient also had a complicated history of infective arthritis, a history of deep vein thrombosis, and chronic obstructive pulmonary disease, while the other cases did not report any noteworthy comorbidities. Lastly, our patient demonstrated substantial ligament incompetence that required a total hinged knee implant rather than a posterior stabilized implant with a modular tibial component, which had been used in the other 2 cases. The cases were similar with regard to exhausting non-surgical options, using a cemented prosthesis, and limb-length discrepancies that were noted on physical examination.

Although the comparison between patients with congenital vascular syndromes undergoing total knee arthroplasty and those undergoing total hip arthroplasty is limited, hemostasis is similarly thought to be of great importance. In a published case of total hip arthroplasty, hemostasis was managed with the use of preoperative embolization¹⁰, while we relied upon the use of a tourniquet and bipolar radiofrequency cauterization in our patient.

The treatment of patients with KTWS bears some resemblance to those with arthropathy due to hemophilia, but patients with KTWS present challenges that are unique to this syndrome. While the surgical treatment in our patient involved extensive debridement and a hinged knee arthroplasty, the true challenge in treating these patients occurs during the preoperative preparation and the short-term postoperative follow-up. Although follow-up will determine the long-term success of this procedure, with careful preoperative planning and

consideration of potential complications, we believe that these patients can have a safe and functional arthroplasty experience.

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Fig. 1. Clinical photograph depicting chronic changes to the knee from KTWS, including hypertrophic tissue, arteriovenous malformations (AVMs), and skin changes related to venous insufficiency.

Fig. 2-A

Fig. 2-B

Fig. 2-C

Fig 2.

Figs. 2-A and 2-B Preoperative anteroposterior (Fig. 2-A) and lateral (Fig. 2-B) radiographs of the affected extremity. There are destructive, erosive-appearing changes at the joint interface as well as a relatively elevated joint line compared with the contralateral extremity. Soft-tissue shadows are evident throughout, demonstrating chronic hypertrophied soft tissue and AVMs. **Fig. 2-C** Axial cross-sectional CT angiogram of the lower extremities. The areas marked with an asterisk (*) indicate areas of high-flow AVMs.

Fig. 3.
Intraoperative image demonstrating the chronic changes to the joint surfaces, including the hemosiderin-laden tissues.

Fig 4.
Figs. 4-A and 4-B Postoperative anteroposterior (Fig. 4-A) and lateral (Fig. 4-B) radiographs of the knee at 26 months. These images demonstrate a well-fixed hinged knee implant.