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Case study of idiopathic degeneration of the talonavicular joint

by Ryan Allen, DPM[1*]; William Arthur, DPM[1]; Christina Ma, BS[2]; Charles Parks, DPM, FACFAS[3]; Monara Dini, DPM, FACFAS[3]

The differential diagnosis for chronic pain out of proportion is broad, and a final diagnosis of Mueller-Weiss syndrome is often a diagnosis of exclusion. We present a patient who experienced pain out of proportion following minor trauma. This progressed into worsening pain that affected his day-to-day activities and ability to perform work. Eventually, there was destruction of his talonavicular joint and early stages of idiopathic fusion. Multiple specialties were involved in this case including infectious disease, rheumatology, and neurology. Excluded diagnoses were septic joint, osteomyelitis, complex regional pain syndrome, Charcot arthropathy, and rheumatoid arthritis. We present a rare case of a patient who experienced idiopathic destruction and fusion of his talonavicular joint following minor trauma, with Mueller-Weiss syndrome suspected. The patient would make significant recovery following arthrodesis of the talonavicular joint.

Keywords: Mueller-Weiss-syndrome, complex regional pain syndrome, idiopathic fusion, talonavicular joint

Müller-Weiss syndrome is a rare disease that is described as a spontaneous adult-onset tarsal navicular osteonecrosis [1]. Symptoms include chronic midfoot pain, swelling, and tenderness on the dorsal and medial midfoot. It is commonly found bilaterally and is found more frequently in women [1]. Its pathogenesis remains controversial. Some believe it to be caused by secondary compressive forces acting on the tarsus or possibly a congenital defect, while others believe it to be an ischemic process[2]. The characteristic findings for Müller-Weiss syndrome include a dorsomedial dislocation along with the collapse of the lateral navicular bone, resulting in a comma-shaped.

The first description of this condition was in the early twentieth century in Europe. In 1925, Schmidt reported on a patient with pluriglandular endocrine failure with deformities at the tarsal navicular. Walther Müller described this condition in 1927, where he suggested that the disease developed from a forceful compression of the lesser tarsus. Müller later suggested a congenital defect was the cause of the disease the following year [3]. Also, in 1927, an Austrian radiologist, Konrad Weiss, described similar findings in two patients suggesting osteonecrosis as the cause for the condition [4]. Although Schmidt was the first to describe this condition, the disease is named after Müller and Weiss.

The exact prevalence and incidence of this disease is currently unknown. However, there are isolated case reports throughout the literature. It is commonly present in the fourth to sixth decade of life. It is also frequently bilateral and usually found in patients with a higher body mass index. There is limited data

1 - Podiatric Surgery Resident, Department of Veteran Affairs at San Francisco
2 - Podiatric Medical Student, California School of Podiatric Medicine
3 - Assistant Professor, Department of Orthopaedic Surgery, University of California at San Francisco
* - Corresponding author: ryandylanallen@gmail.com

suggesting an environmental and nutritional component [5]

The pathogenesis of this condition is poorly understood. However, the literature suggests that there are two contributing factors; a delay in ossification of the navicular and atypical compressive forces on the midfoot.

There are multiple factors that can contribute to a delay in ossification including poor nutritional status, endocrinopathies, metabolic disease, or malabsorption disease [5]. When there is a delay in the ossification of the navicular, the weak outer chondral surface is susceptible to abnormal development from excessive compressive forces. Current literature suggests a large compressive force would contribute to plastic deformation of the navicular during ossification [5]. However, it is possible that persistent low compressive forces on the pliable chondral surface puts the navicular at risk of ossifying in an irregular orientation.

The second contributing factor is a result of the biomechanical insult on the navicular. As mentioned previously, excessive compressive forces on the navicular is suggested to contribute to this condition. This is particularly true when these forces are applied to the lateral half of the navicular between the talar head and the cuneiforms. There are several conditions which can lead to compressive forces to the lateral aspect of the navicular including: primary subtalar joint varus, first ray brachymetatarsia (both congenital or acquired), and clubfoot deformities [5]. Other biomechanical factors such as a short hallux metatarsal, shortening of the entire medial column due to internal rotation of the navicular in the transverse plane, or retroposition of the first tarsometatarsal joint in relation to the second tarsometatarsal joint can also contribute to lateralization forces [5]. When the first ray is hypermobile, loading forces transfer to the second ray which may also lead to compressive forces into the intermediate cuneiform and lateral navicular [5].

Case Report

A 41-year-old male with a past medical history of right foot plantar fasciitis, depression, and insomnia was brought in by ambulance to the Emergency Department of San Francisco General Hospital with a chief complaint of 10/10 right foot pain accompanied by swelling. Two days prior, the patient had been exercising at his gym. He did not recall any particular injury apart from his right foot slipping off exercise equipment onto the floor, without any immediate pain. Several hours after exercising, he noticed a gradual increase in right foot pain. In the Emergency Department, plain films did not demonstrate fracture, there was diffuse soft tissue swelling on the dorsum of the foot (Figure 1), labs were not drawn, however, the patient's vital signs were all within normal limits. He was discharged from the ED with an ankle brace, crutches, and Tylenol for pain relief.

He again visited the ED 3 days later, with a further increase in pain and edema; he was not found to have any underlying fluctuance or erythema. He remained unable to bear weight on his right foot. He remained in an ankle brace and had been using ibuprofen without any significant pain relief, and he continued to not have any constitutional symptoms. Radiographs demonstrated increased soft tissue swelling from prior films, however, no evidence of fractures, soft tissue calcifications, or joint effusions were seen. He was recommended to continue use of the ankle brace, to stop ibuprofen, and trial Tylenol.

Nearly one month later he presented to the ED with continued pain, however now able to bear weight. He had plain films which demonstrated even further increase in soft tissue swelling as well as a CT (Figure 2) with findings of prominent osteopenia involving the midfoot. Orthotics and Prosthetics was consulted and he was dispensed a CAM boot. The podiatry service was notified, and an appointment was made for the following day in the clinic. He was dispensed opioid pain medication, as well as ibuprofen, at discharge.

The next day, he presented to the Podiatry Clinic. His right dorsal foot was edematous and erythematous from the metatarsal heads to the level of the ankle joint. Fluctuance was not appreciated. The right dorsal foot was tender to palpation.

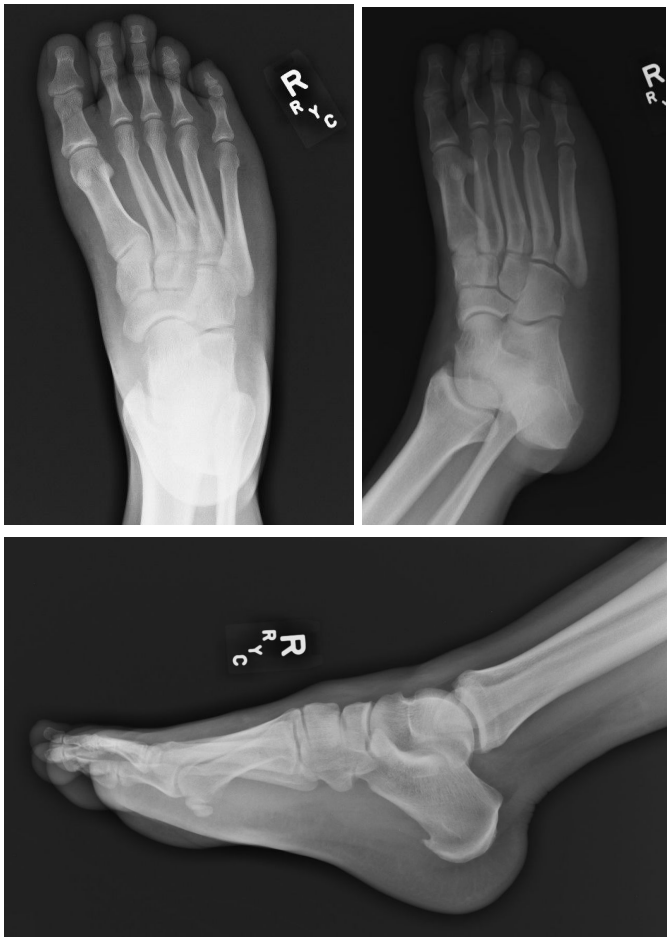

Figure 1 Radiographs of patient's foot. Diffuse swelling on the dorsum of the foot without evidence of fracture or dislocation.

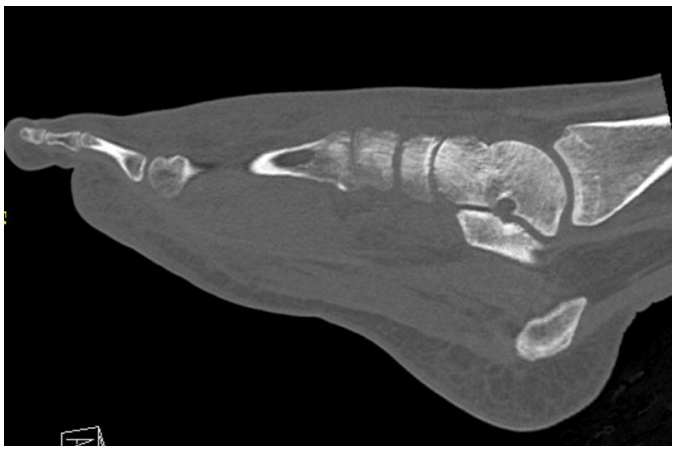

Figure 2 Sagittal CT demonstrating further soft tissue swelling with prominent osteopenia of the midfoot.

Allodynia was present. Pain increased with plantarflexion of the right ankle joint. Pain was also present with eversion and inversion of the right subtalar joint. He was unable to wear the CAM boot, dispensed to him the prior day, as this increased his pain. A CBC, ESR, CRP, BMP, and HgA1c, and a MRI with contrast were ordered to assess for abscess or osteomyelitis. His ESR and CRP were >130 mm/h and 198.0 mg/L, respectively. His HgA1C was 5.9%, and his uric acid level was 3.7 mg/dL. He was without leukocytosis and his remaining labs were unremarkable. An ACE wrap was applied from the foot to the tibial tuberosity to compress the foot. The patient was given strict instructions to elevate, do ROM exercises, and wear the CAM walker boot at a 90 degree angle to prevent worsening flexion contracture. He was given explicit instructions to return to the ED if he developed any fever, chills, nausea, and vomiting or any worsening pain. He was dispensed gabapentin to be taken at bedtime for pain.

The previously ordered MRI was taken and reviewed. Findings were reviewed with radiology, and they were suggestive of broad diagnoses. Clinically and radiographically there was support of complex regional pain syndrome. It showed destruction of joint spaces at the TN joint, subtalar joint, as well as deep marrow edema within the midfoot outside of the talonavicular joint (Figure 3). The navicular bone was almost entirely replaced with edema and enhancement.The MRI findings were also consistent to support Charcot arthropathy and osteomyelitis as well. Septic joint was also on the differential due to minimal fluid within joint spaces. Interventional Radiology was consulted for urgent bone biopsy. There would be no growth from the cultures of the specimen acquired.

The patient then underwent a right foot irrigation and debridement with bone biopsy approximately one week later with the presumption of infection versus complex regional pain syndrome. At this time the symptoms began 3 months prior. Gout was lowest on the differential due to a prior uric acid level of 3.7 mg/dL. In the operating room, a 4 cm incision was made dorsally to gain access to the talonavicular joint. A Rongeur forceps was used to collect a sample of synovial tissue which was noted to appear inflamed and thickened; this was sent to microbiology. The talus and navicular were assessed and noted to be hard and viable. A sample of cartilage and bone was taken from both the talus and the navicular and sent to microbiology. Bone specimens were also sent to pathology from both talus and navicular. The patient would subsequently be admitted to the hospital.

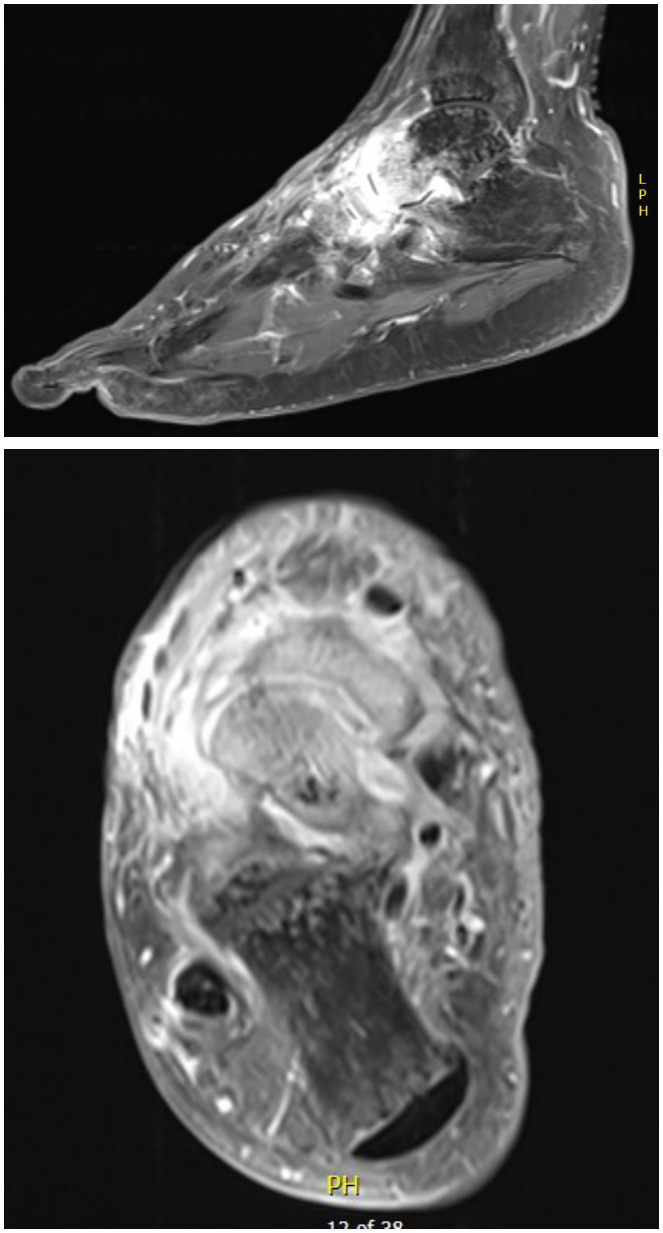

Figure 3 Sagittal and transverse T2 MR imaging demonstrating destruction of the talonavicular joint with extensive marrow edema that was also present to the calcaneus and cuneiforms.

During his hospital stay, neurology was consulted to rule out CRPS. He was noted to not have any evidence of neuropathy with "intact afferent and efferent limbs of reflex arc." Neurology assessed a low suspicion of neuropathic etiology of symptoms. In terms of the acquired specimens from the OR, there was no growth noted from any of the specimens (C&S, AFB, and fungal). The synovial tissue final pathologic diagnosis was noted to be "lymphoplasmacytic inflammation." Excised bone of both the talus and the navicular were noted to be without any evidence of acute osteomyelitis.

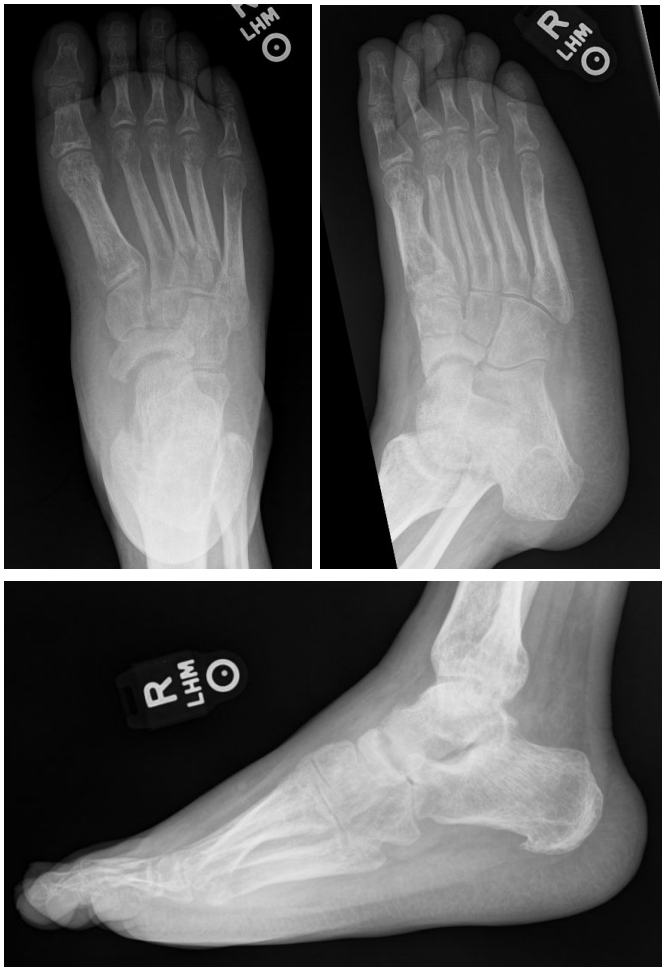

Figure 4 Radiographs demonstrating focal osteopenia and erosive changes of the talonavicular joint.

The patient remained in-house for 5 days; he was then discharged home. Two days after discharge he returned to the clinic; he was now one week status post biopsy. He remained without constitutional symptoms. The differential diagnosis remained the same at this point, with complex regional pain syndrome as most likely diagnosis despite low suspicion from the Neurology team. New labs were ordered. Rheumatoid factor was noted to be within normal limits <3.5 IU/mL and antinuclear antibody was noted to be negative as well. His ESR had decreased to 68 mm/h and his CRP had decreased to 6.4 mg/L. The patient was encouraged to continue ambulating with a CAM boot and to practice ROM exercises. He would later follow up with the infectious disease clinic who noted that "osteopenia and marrow enhancement on imaging, elevated inflammatory markers, and absence of evidence for neuropathy on exam (making Charcot arthropathy unlikely) are concerning for infectious etiology."

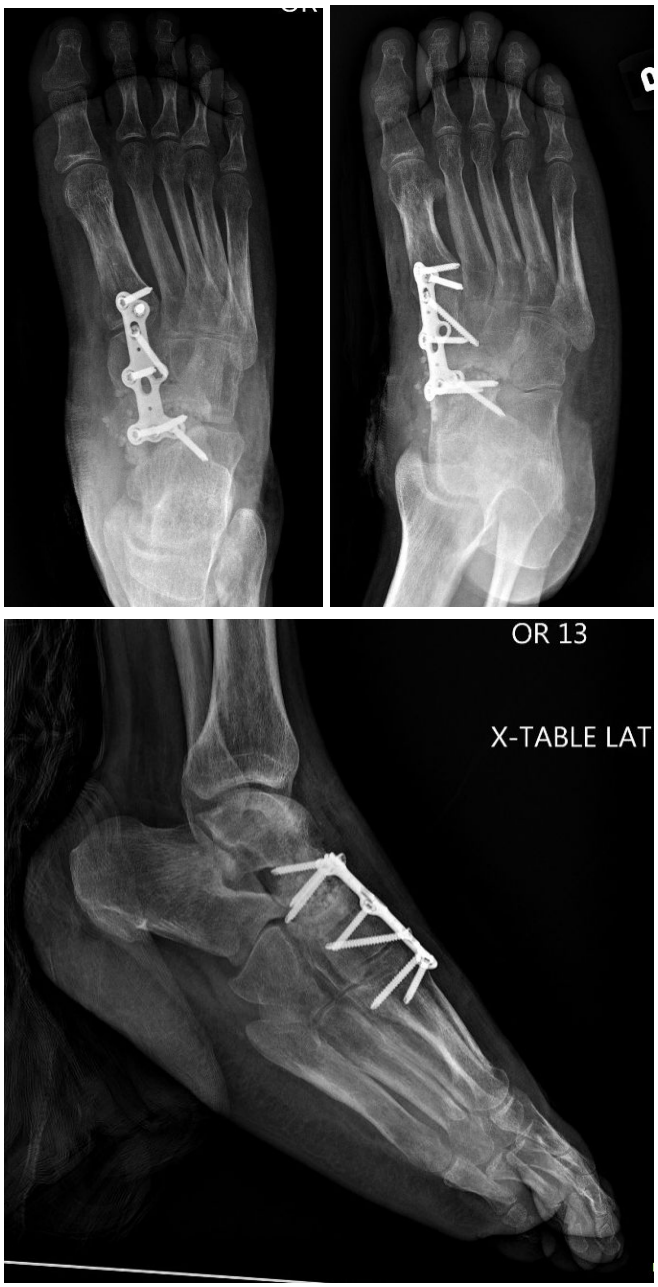

Figure 5 Immediate post-operative films of the talonavicular fusion.

However, both cultures and pathology were without evidence of osteomyelitis and he did not have a systemic illness which could have led to hematologic seeding or skin breaks down allowing for direct inoculation. Serologies were sent to rule out less common infectious agents, such as TB and coccidioides. The serologies were noted to be negative for both TB and coccidioides. The patient returned to the Podiatry Clinic two weeks later, now 3 weeks status postbiopsy and almost four months from the time of onset of symptoms. He attempted to ambulate in his CAM boot but was unable to do due 5/10 throbbing pain along his medial arch. He continued to not have any constitutional symptoms. He was encouraged to transition to regular shoes and discontinued the CAM boot. His sutures were removed at this visit. Since he had been discharged from the hospital, he had begun taking a new pain management regimen of gabapentin 300 three times daily which he noted to help alleviate some pain.

The patient returned to the clinic 5 weeks status post the open biopsy. He was still unable to walk in regular shoe wear and remained in his CAM boot utilizing a knee scooter. It had been two months since he last tried physical therapy and he wished to restart therapy. New plain film imaging was taken, and noted to be without any interval changes. He was encouraged to ambulate in regular shoe wear and to mobilize the foot as much as possible. It was stressed that weight bearing was imperative to make a recovery.

Two weeks later the patient returned to the clinic with new plain films. He was now 7 weeks status postbiopsy. He attempted to ambulate only utilizing the CAM boot but remained unable to bear weight on his right foot in regular shoe wear. On plain film imaging he was noted to have unchanged severe osteopenia from prior films, however with more focal osteopenia/erosion involving the talonavicular joint (Figure 4). X-ray findings correlated with TNJ degenerative joint disease.

At this point in summary, neurology had evaluated the patient and had very low suspicion for complex regional pain syndrome. Medical workup had also been negative for inflammatory arthritis. Of note, the patient had no risk factors for Charcot arthropathy. The case was discussed on multiple accounts with the orthopaedic department and the patient was then placed in a short leg cast, made non-weight bearing, with a plan of serial casting with imaging to promote autofusion of the TNJ. If the patient was to continue to have pain, surgical fusion of the TNJ would be considered.

The patient returned to the clinic 2 weeks, now 6 months status-post the initial onset of pain, and now with new assessment of TNJ degenerative joint disease secondary to unknown etiology with differential of CRPS, Müller-Weiss syndrome, septic joint, or osteomyelitis.

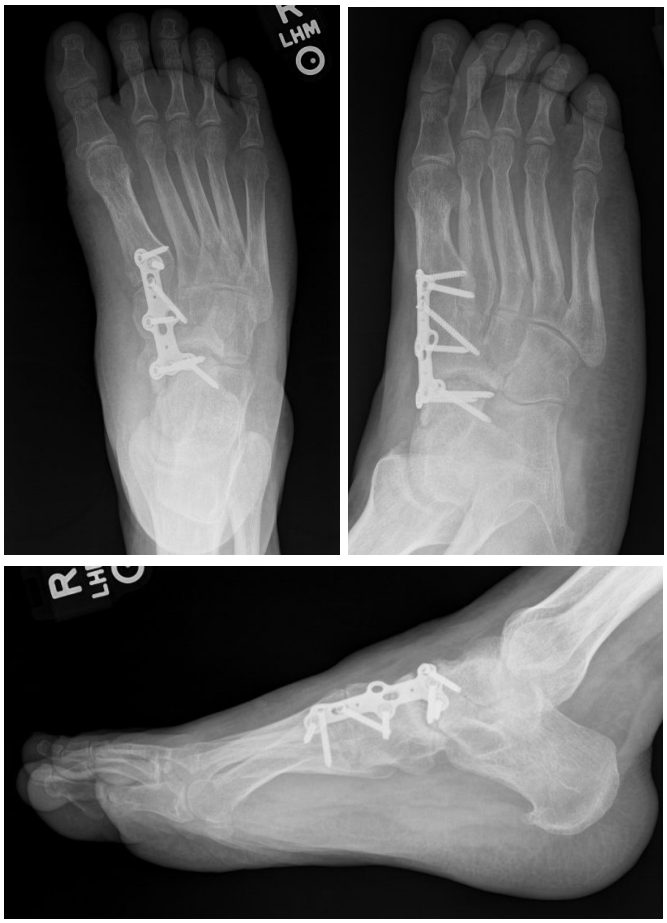

Figure 6 Eight weeks post-operative films demonstrating trabeculation across the arthrodesis site.

He had remained in a short leg cast and NWB as instructed, and ambulating with a knee scooter. He remained without constitutional symptoms. At this time, surgery for fusion was discussed, as well as conservative treatment of serial casting. Patient opted to go with the latter. It was discussed that 6-8 weeks of serial casting may be adequate for fusion. Patient was again casted and he followed up again 2 weeks later. Again, surgery was discussed as the patient had no change in his symptoms, and was placed in a CAM boot with a plan for incisional biopsy.

The patient then opted to undergo a right foot incisional biopsy of the talus and a TN joint preparation for fusion. The patient returned to the operating room now just over 8 months from the initial onset of pain and symptoms. An incision was made through the prior incision site and deepend down to the talonavicular joint and a capsulotomy was performed. A pseudoarthrosis was noted. The articular surface of the navicular and talus were exposed and the surfaces were noted to have patchy areas of hyaline and fibrocartilage. A curette was then used to resect three separate specimens consisting of cortical bone from the articular surfaces of the navicular and the talus. This procedure's goal was to prepare the joint for fusion and for a final biopsy. Fungal, bacterial, and acid fast bacilli would all be negative for growth.

The patient returned to the operating room for a third and final time, to undergo a right foot talonavicular arthrodesis with iliac crest bone graft at 9 months from onset of symptoms. An incision was made approximately 10 cm from the talar neck to the base of the first metatarsal. Dissection was carried down to the talonavicular joint was prepared utilizing curettes, fish scaled with osteotomes, and fenestrated using 0.062 K-wire. Attention was then directed to the right anterior superior iliac spine to acquire autograft. The talonavicular site was temporarily fixated and a combination of cancellous autograft, cancellous allograft bone chips, and 10 cc of Stimulan Biocomposite calcium sulfate were placed into the talonavicular arthrodesis site around the structural iliac crest autograft. Next, the talonavicular arthrodesis site was then fixated with a small Wright Medical medial column fusion plate and 3.5 mm fully threaded locking and nonlocking screws (Figure 5). The patient was then subsequently admitted for 48 hours for observation. Intraoperative bacterial cultures from bone were acquired but would be negative for growth.

The patient remained non-weight bearing for a total of 12 weeks, both with a short leg cast for the first 8 weeks followed by a CAM boot with ROM exercises for an additional 4 weeks. Serial radiographs demonstrated uneventful healing of the arthrodesis site (Figure 6).

He continued to be followed weekly in the Podiatry Clinic. He continued to have interval changes of healing of the arthrodesis site on serial x-rays. The patient continued to work with physical therapy, primarily range of motion exercises. At 11 months he was noted to have significant improvement in pain during ambulation. He was able to transition to regular shoe wear at this time. At 13 months, he was noted to have 1/10 level of pain. At this point, he felt that he could return to day to day activities without restriction. There were no complications from any of

the surgical interventions at his last follow up visit of this review, 13 months from the onset of symptoms.

Discussion

This is an atypical presentation of Müller-Weiss. The inciting event was minor trauma, however, this is per the patient's report, and therefore is subjective. The first ray mobility was not evaluated prior to the initial presentation. The patient had swelling and pain of the midfoot, however, this was unilateral. Although the patient did not have a higher than average BMI, he was athletic and he had exceedingly high midfoot torque on his foot during exercise routines. The appropriate steps in the management of other diseases including complex regional pain syndrome were met. Biopsies were taken to rule out infectious causes, and multiple serologies were taken, even to exclude TB. Multiple imaging modalities were also used. The patient was directed to pain management, and he was treated by physical therapy before and prior to the talonavicular fusion; he improved following the procedure of a talonavicular joint arthrodesis.

Apart from minor trauma, the unilateral destruction of the talonavicular joint is what makes this case unique. Müller-Weiss syndrome is often bilateral and not following any incidence of trauma. It is possible in this case that the initial inflammatory phase of this disease process leads to the eventual destruction of this patient's talonavicular joint, resulting in the need for an arthrodesis. This would explain why he would make a recovery following this procedure in conjunction with continuing physical therapy. We therefore believe that this is an atypical case of complex Müller-Weiss Syndrome, although atypical.

Conflict of Interest Declaration

The corresponding and contributing authors have no relevant financial interest in this manuscript.

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