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CASE REPORT

Mueller Weiss Syndrome: A Missed Cause of Mid Foot Pain

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Abstract

Mueller-Weiss syndrome, or spontaneous osteonecrosis of the tarsal navicular bone in adults, is an idiopathic foot condition that causes chronic mid- and hind-foot pain with progressive deformity of the midfoot in adults. In this report, we describe the radiographic and magnetic resonance imaging of a rare case of Mueller-Weiss syndrome associated with spontaneous osteonecrosis of tarsal navicular bone resembling a cystic lesion in a 62-year-old female.

Keywords

Ankle pain, Cystic lesion, Navicular avascular necrosis, Spontaneous osteonecrosis

Introduction

The Mueller-Weiss Syndrome (MWS), which was first described in 1927, refers historically to spontaneous adult-onset tarsal navicular osteonecrosis, originates after the surgeon Walther Mueller who described patients with this condition, and the radiologist Konrad Weiss, who subsequent postulated osteonecrosis as the underlying etiology [1]. Unlike Kohler's disease, which is a childhood osteochondrosis of the navicular bone where asymptomatic and symptomatic patients alike may demonstrate similar radiographic findings, most patients with MWS are symptomatic at the time of radiographic diagnosis [2]. In this case report, we present a rare case of Mueller-Weiss syndrome characterized by cystic lesion to increase awareness of this important and under-recognized cause of foot pain in adults.

Case Report

A 57-year-old woman with no relevant medical history presented with left midfoot pain with intermittent swelling for 4 months. Her symptoms worsened with weight-bearing activities and improved with rest and elevation of the affected leg. On physical examination, she showed tenderness on palpation and manipulation of the medial midfoot and swelling limited to the dorsomedial aspect of the right foot, with no other focal abnormalities. Range of motion in the subtalar joint and midfoot joint was painful but not limited. Neurovascular status was normal in both feet.

Radiographs showed a well-demarcated cystic lesion as well as increased radio density and a slight decrease in bone height at the lateral aspect of the navicular bone. Dorsolateral protruding fragmentation was noted. Arthritic changes were also noted in the metatarsal region (Figure 1).

Because of the persistent pain, an MRI was performed, which showed bone marrow edema with low-signal areas on T1- and T2-weighted images in the marrow of the tarsal navicular bone, suggesting persistent osteonecrosis at the site of maximal pain (Figure 2).

Discussion

MWS is characterized by a progressive painful clinical course with progressive navicular fragmentation and talo-navicular destruction leading to midfoot and hindfoot deformity [3,4]. To our knowledge, 277 cases of Mueller-Weiss syndrome have been described in the literature [2].



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Figure 1: Radiography of the foot showing a well-demarcated cystic lesion as well as increased radiodensity and a slight decrease in bone height at the lateral aspect of the navicular bone.

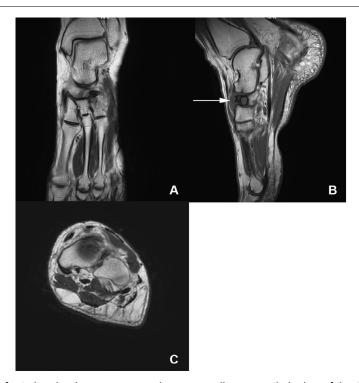


Figure 2: MRI of the foot showing bone marrow edema as well as a cystic lesion of the tarsal navicular bone.

MWS manifests clinically with chronic pain in the midfoot that occurs during periods of weight-bearing activity. It is suggested that chronic stress, previous injury, and disruption of blood supply may be the cause of this syndrome [5]. MWS is more common in women. Risk factors include smoking, alcohol consumption, and corticosteroid use, as well as rheumatologic, hematologic, and metabolic disorders [6].

MWS is thought to be a result of excessive compressive mechanical loading, along with the alteration of foot biomechanics characterized by pes planus, which disrupts the microvascular structure of the bone, leading to bony osteonecrosis [1,7,8]. Additionally, this compressive displacement leads to flattening and fragmentation of the dorsolateral

aspect of the navicular bone with subsequent hindfoot varus, which is a common finding in MWS [1]. The medial plantar artery supplies the plantar aspect of the navicular bone, while the dorsal pedal artery supplies the dorsal and lateral aspects. The central zone of the navicular bone is therefore less well supplied with blood and is also the area of maximum shear forces [1].

Due to internal rotation of the medial half and compression of the lateral half, the navicular in MWS shows a "comma-shaped" configuration instead of the normal "boat-shaped" configuration, with decreased size, increased radiodensity and fragmentation [7]. Most patients present with chronic midfoot and hindfoot pain in the fourth or fifth decade, when secondary osteoarthritis develops [2].

Weight-bearing radiographs in the frontal and lateral projections remain the test of choice in the evaluation of MWS [3]. Cystic lesions are not common in MWS. Only one case in a 25-year-old woman has been reported, which was localized in the talus head [4]. In the present case, imaging showed a well-demarcated cystic lesion on the lateral portion of the navicular bone, a finding that, to our knowledge, has not been reported in the literature.

Initial treatment options are nonsurgical, including anti-inflammatory analgesic medications and possible immobilization without weight bearing. If pain persists after conservative treatment, surgical interventions such as core decompression and autologous bone grafting may be considered [9]. In our case, the patient was offered navicular core decompression surgery after several unsuccessful attempts with conservative treatment, but she declined surgery. She is treated conservatively with a satisfactory result.

Conclusion

MWS is a rare condition that can be misdiagnosed and improperly treated. It is a rare cause of chronic midfoot and hindfoot pain of the foot. MWS should not be confused with normal bone cysts, and MRI is always recommended to rule out osteonecrosis. Navicular decompression surgery is the best treatment for a definitive result. A better understanding of this disease and its radiographic manifestations may allow earlier diagnosis and improved future treatment.

Authors Declaration

The authors declare that they have no competing interests. The authors also declare that the patient gave informed consent before our report was performed.

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