# Answer to Case of the Month #129 Mueller-Weiss Syndrome

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## History

A 57-year-old female developed a 1-year history of progressive midfoot pain, made worse with activity. There was no history of significant trauma or corticosteroid use, and no clinical features of infection. Computed tomography (CT) imaging was performed to assess for a navicular stress fracture.

## Diagnosis

Mueller-Weiss Syndrome.

## CT Findings

Helical axial and reconstructed sagittal imaging demonstrate collapse and sclerosis of the lateral aspect of the tarsal navicular (Figure 1), with sparing of the medial aspect of the navicular (Figure 2), resulting in a comma-shape deformity (Figure 3). No stress fracture is identified. The talonavicular joint demonstrates mild degenerative change.

## Discussion

Mueller-Weiss syndrome is a rare entity related to adult-onset spontaneous osteonecrosis of the navicular. It is a painful and debilitating illness, which is more frequent in women and can be either unilateral or bilateral.[1,2] It was first described by Walther Mueller in 1927, who believed this condition was due to chronic compression of the navicular from the adjacent bones.[3] In 1929, Konrad Weiss further described a similar pathology; however, he ascribed osteonecrosis as the primary etiology.[4]

Mueller-Weiss syndrome is not to be confused with Kohler disease, a well-recognized, separate entity characterized by spontaneous osteochondrosis of the navicular in children. These disorders differ not only in age of onset but also in etiology, radiological findings, and clinical outcome.

The classic radiological finding of spontaneous osteonecrosis of the navicular, as first illustrated by Mueller, is collapse of the lateral aspect of the navicular with dorsal protrusion.[6] Current literature describes both early and late radiological changes. Initially, there is loss of volume and increased radiodensity observed in the lateral aspect of the navicular, resulting in the classic comma-shape deformity. This is followed by dorsal protrusion and fragmentation.

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Received January 31, 2006

Revision requested April 13, 2006

Resubmitted June 5, 2006

Accepted June 16, 2006

Can J Assoc Radiol 2008;59(1):39–40

**Key Words: avascular necrosis; navicular**

**Figure 1:** Sagittal reconstructed computed tomography (CT) image shows collapse and sclerosis of the lateral aspect of the navicular.

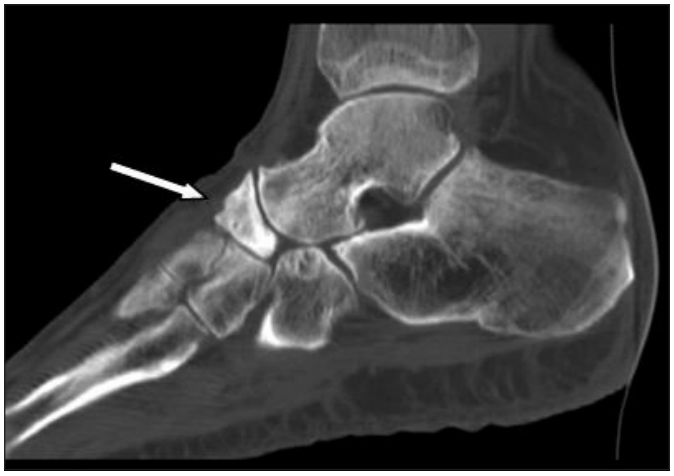


**Figure 2:** In an image more medial to Figure 1, there is relative sparing of the medial aspect of the navicular.

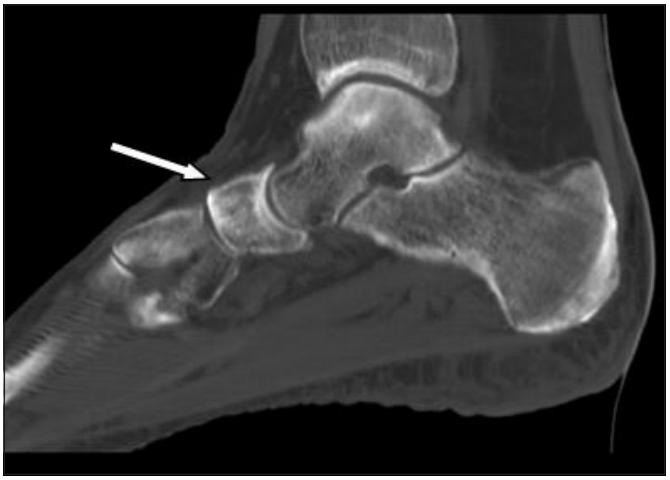
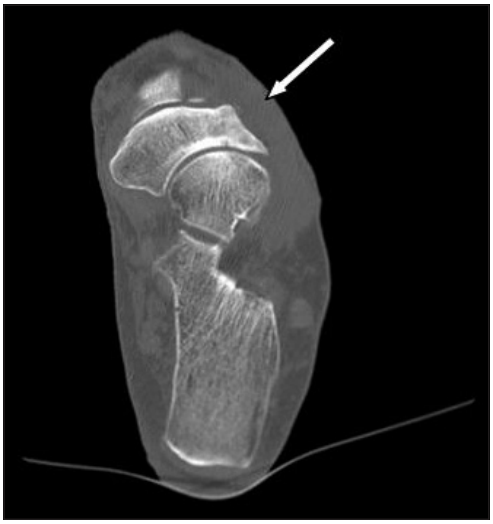


**Figure 3:** Helical axial CT image demonstrates the classic comma-shaped deformity of the navicular seen in Mueller-Weiss syndrome, with mild degenerative change in the talonavicular joint.



The etiology of Mueller-Weiss syndrome is still unclear; there are several proposed mechanisms relating to previous injury or chronic stress.[2] Increased mechanical strain, and altered tension forces may lead to diminished blood supply to the lateral portion of the navicular.[2,7,8] Subsequent structural changes in the bone cause abnormal concentration of loading forces that, along with ligamentous laxity, may account for dorsal protrusion, and further impingement of the vascular supply.[2,5,7]

Patients with Mueller-Weiss syndrome follow a chronic and severe clinical course, often associated with pain and progressive deformity. Initial conservative management, such as immobilization with orthoses and antiinflammatory medications, is often unsuccessful.[5,9] In such cases, patients may require surgical intervention, and talonavicular +/– naviculocuneiform arthrodesis may be indicated.[5,9,10] Mueller-Weiss syndrome left untreated can result in advanced midtarsal osteoarthritis and subsequent permanent disability.[10]

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