An Uncommon and Under recognised Contributor of Foot Pain: A Case Study of Mueller Weiss syndrome

Rachel Wei Lian Loh*, Eelin Tan, Chern Yue Glen Ong

Department of Diagnostic Radiology, Tan Tock Seng Hospital, Singapore

***Correspondence:** Rachel Wei Lian Loh, Department of Diagnostic Radiology, Tan Tock Seng Hospital, 11 Jln Tan Tock Seng, Singapore 308433

racheloh.wl@gmail.com

Radiology Case. 2024 October; 18(10):14-21 :: DOI: 10.3941/jrcr.5410

ABSTRACT

Mueller-Weiss syndrome refers to spontaneous osteonecrosis of the navicular bone in adults. It is often bilateral, affecting women aged between 40 and 60 years. Herein, we present a case demonstrating characteristic imaging features on radiograph, magnetic resonance imaging and sonography.

CASE REPORT

CASE REPORT

A 57-year-old female presented with a 5-month history of pain and gradually enlarging swelling over the plantar aspect of her right foot. She had no related history of trauma and was otherwise clinically well. On physical examination, there was palpable fullness and tenderness over the medial midfoot. The overlying skin and neurovascular examination were unremarkable.

Imaging findings

Radiographs showed sclerosis and collapse of the lateral aspect of the navicular bone, resulting in comma-shaped configuration and dorsomedial protrusion of the navicular bone, with associated collapse of the longitudinal arch and perinavicular osteoarthritis (Figure 1). Radiographs from 11 years prior, obtained for a different complaint, showed no navicular bone abnormality (Figure 2).

Further imaging with magnetic resonance imaging (MRI) of the foot showed a severely collapsed and fragmented navicular bone, with marrow edema and cystic changes, compatible with osteonecrosis (Figure 3). There was re-demonstration of longitudinal arch collapse and secondary osteoarthritis in the talonavicular and naviculo-cuneiform joints. Ancillary findings included subcutaneous adventitial bursitis at the lateral plantar aspect of the midfoot, as well as severe fatty replacement of the abductor digiti minimi suggestive of chronic Baxter neuropathy (Figure 4). These changes likely reflect altered mechanics and loading caused by the structural alterations of Mueller-Weiss syndrome (MWS).

Attempted aspiration of adventitial bursa under ultrasound guidance for symptomatic relief yielded minimal thick serous fluid. During pre-procedure sonography, the navicular was observed to be deformed and fragmented with surrounding synovitis (Figure 5).

Management/Follow-up

The patient declined surgical management and was referred to podiatry for footwear modification. She continues to be followed up by the Foot and Ankle service.

DISCUSSION

Etiology & Demographics

MWS refers to the occurrence of spontaneous osteonecrosis of the navicular bone in adults. Typically, this syndrome affects individuals aged between 40 and 60 years. It often manifests bilaterally, with a higher incidence among women, comprising up to 70% of cases [1-3]. The precise prevalence of MWS remains uncertain and is likely underestimated, as it can be misdiagnosed for other conditions such as osteoarthritis of the perinavicular joints [4].

This condition presents as persistent mid and hindfoot pain, particularly exacerbated during weightbearing activities [1,2]. Individuals may also exhibit deformity of the tarsal navicular, with swelling and tenderness over the dorso-medial aspect of the mid foot. Furthermore, there may be hindfoot varus with a normal or reduced medial longitudinal arch, depending on disease severity [1]. While some patients may tolerate the deformity and remain mostly asymptomatic for an extended period, others may experience symptomatic discomfort [5]. The pain and discomfort associated with MWS may be attributed to abnormal pressure distribution, with increased plantar pressures at the midfoot and reduced toe pressures, along with degenerative osteoarthritis affecting the perinavicular joints [5,6].

The etiology of MWS remains a subject of debate and uncertainty. Various theories have been proposed, with Maceira

and Rochera presenting one of the largest series of MWS cases. They suggested that a combination of delayed ossification of the tarsal navicular and abnormal force distribution could contribute to the development of MWS [5]. Delayed ossification may stem from localized or generalized developmental disturbances. Generalized developmental disorders could result from nutritional deficiencies, which may be extrinsic or intrinsic (related to endocrinopathies, metabolic or malabsorption diseases) [1,5].

The tarsal navicular serves as a critical component of the medial column of the foot, contributing to the integrity of both the medial longitudinal and transverse arches. Vascular supply to the navicular is provided by the medial plantar artery on the plantar aspect, and the dorsalis pedis artery on the dorsal and lateral aspects, forming a circumferential and centripetal vascular network. Consequently, the central zone of the navicular experiences reduced blood supply and is also the area subjected to maximal shear forces, potentially explaining the predilection for stress fractures and osteonecrosis in this region [1,2,4].

Clinical & Imaging Findings

Plain weightbearing radiographs form the mainstay for diagnosing MWS. Key radiologic findings include the presence of sclerosis, fragmentation, and comma-shaped deformity of the navicular bone, with associated dorsomedial protrusion. Additional observations may include peri-navicular degenerative changes, widened sinus tarsi, medial shift of the cuboid relative to the calcaneus, and hypertrophy of the second metatarsal secondary to lateral shift of compressive forces [1,2].

Maceira delineated five stages of the condition on plain weight-bearing lateral radiographs. Stage 1 exhibits minimal to no radiographic changes. Stages 2 to 4 demonstrate progressively worsening collapse and fragmentation of the navicular, accompanied by loss of the longitudinal arch. Stage 5 represents the final stage, characterised by complete extrusion of the navicular fragment and talocuneiform neo-articulation [5].

However, the severity of symptoms may not necessarily align with radiologic deformity or disease stage. Degenerative osteoarthritis at the perinavicular joints is a common finding and likely contributes to pain, often representing the final stage of the disease [5].

Computed tomography (CT) scans offer more precise determination of navicular bone stock and density, measurement of medial column shortening, evaluation of fracture lines and deformity, assessment of surrounding arthritis, as well as planning for fixation size and surface [7,8].

MRI aids in early detection of the condition, demonstrating bone marrow edema, loss of signal intensity in dorsolateral bone marrow, and joint effusion. It also helps detect early perinavicular osteoarthritic changes, assess soft tissue structures, and rule out other conditions like stress fractures or infection [8,9].

Treatment & Prognosis

Currently, there is no consensus on the most effective treatment approach for patients with MWS. Most studies advocate for an initial trial of non-surgical management, which can be attempted over 2 to 60 months. Non-surgical treatment options include nonsteroidal anti-inflammatory drugs (NSAIDs) for pain relief, activity modification, healthy weight management, adjusting footwear, utilizing ankle-foot orthotics, bracing, and sometimes immobilization with non-weight-bearing casts [1,7,9,10].

Surgical intervention should be considered if non-operative treatments have been attempted without success and symptoms persist. The decision for surgery should be based on the severity of symptoms rather than the extent of deformity [1,10].

The fundamental principles of surgical intervention involve addressing symptomatic degenerative joints through arthrodesis to alleviate pain, as well as to restore the plantar vault and medial longitudinal arch (Meary–Tomeno axis) [1,10]. Various surgical options exist, including core decompression, internal fixation of the navicular, talonavicular arthrodesis, triple arthrodesis, talonavicular-cuneiform arthrodesis, and navicular excision with reconstruction of the medial column [1,10,11]. Additionally, procedures like Achilles tendon lengthening or calcaneal osteotomy may be considered if necessary [10].

TEACHING POINT

Mueller-Weiss syndrome refers to spontaneous osteonecrosis of the navicular bone in adults. The hallmark radiographic sign is a “comma-shaped” navicular with lateral collapse and sclerosis.

QUESTIONS

Question 1: Which of the following best describes Mueller-Weiss syndrome?

A) A type of inflammatory arthritis affecting the small joints of the hands and feet

B) Uncommon condition characterized by aseptic necrosis and collapse of the navicular bone

C) Degenerative joint disease primarily affecting the midfoot joints

D) Congenital deformity of the foot involving abnormal development of the plantar arch

E) Chronic infection involving the midfoot

Answer: B

Explanation:

Mueller-Weiss syndrome (MWS) refers to the occurrence of spontaneous osteonecrosis of the navicular bone in adults. The etiology of MWS remains a subject of debate and uncertainty. Various theories have been proposed, with the suggestion that a combination of delayed ossification of the tarsal navicular and abnormal force distribution could contribute to the development of MWS.

Question 2: Which of the following characteristics on imaging may help differentiate Mueller-Weiss syndrome from primary osteoarthritis?

A) Presence of bone marrow edema and joint effusion on magnetic resonance imaging
B) Sclerosis of the navicular
C) Collapse and fragmentation of the navicular
D) Joint space narrowing
E) Osteophyte formation
Answer: C
Explanation:

Joint space narrowing, osteophyte formation, subchondral sclerosis and subchondral cysts are characteristic features of osteoarthritis. However, bone collapse and fragmentation are not typically observed. The key radiological finding in Mueller-Weiss syndrome includes increased sclerosis and collapse of the lateral aspect of the navicular bone, resulting in a comma-shaped configuration and dorsomedial protrusion of the navicular. Secondary degenerative osteoarthritis at the perinavicular joints is a common finding and often represents the final stage of the disease.

Question 3: Which of the following is true regarding Mueller-Weiss syndrome?
A) It is more common in females than males
B) Typically presents with acute onset of symptoms such as joint pain and swelling
C) Primarily affects the ankle joint
D) Commonly affects children and adolescents
E) Always occurs unilaterally
Answer: A
Explanation:

Mueller-Weiss syndrome typically affects middle-aged individuals between 40 and 60 years, with higher incidence among women. It often manifests bilaterally, presenting as persistent mid and hind foot pain, particularly exacerbated during weight-bearing activities.

Question 4: What is a potential treatment option for Mueller-Weiss syndrome?
A) Immunosuppressive therapy
B) Arthroscopic surgery
C) Arthrodesis of the perinavicular joints
D) Topical corticosteroid application
E) Antibiotic treatment
Answer: C
Explanation:

Non-operative treatment is usually trialed first – options include nonsteroidal anti-inflammatory drugs (NSAIDs) for pain relief, activity modification, maintaining a healthy weight or weight reduction, adjusting footwear, utilizing ankle-foot orthotics, bracing, and sometimes immobilization with non-weight-bearing casts. Surgical intervention should be considered if non-operative treatments have been attempted without success and symptoms persist. Various surgical options exist, including core decompression, internal fixation of the navicular, talonavicular arthrodesis, triple arthrodesis, talonavicular-cuneiform arthrodesis, and navicular excision with reconstruction of the medial column.

Question 5: What is the primary goal of surgical treatment in Müller-Weiss syndrome?
A) Improve joint stability
B) Restore normal foot function
C) Promote bone fusion
D) Prevent recurrence of deformity
E) To eliminate pain
Answer: E
Explanation:

The decision for surgery should be based on the severity of symptoms rather than the extent of deformity. The fundamental principles of surgical intervention involve addressing symptomatic degenerative joints through arthrodesis to alleviate pain, as well as to restore the plantar vault and medial longitudinal arch.

REFERENCES

1. Mohiuddin T, Jennison T, Damany D. Müller–Weiss disease. - review of current knowledge. *Foot Ankle Surg.* 2014; 20(2): 79–84. PMID: 24796823.
2. Bartolotta RJ, McCullion JC, Belfi LM, Hentel KD. Mueller–Weiss Syndrome: Imaging and Implications. *Clin Imaging.* 2014; 38(6): 895–898. PMID: 25064253.
3. Doyle T, Napier RJ, Wong-Chung J. Recognition and management of Müller-Weiss disease. *Foot Ankle Int.* 2012; 33(4): 275–281. PMID: 22735199.
4. Samim M, Moukaddam HA, Smitaman E. Imaging of Mueller-Weiss Syndrome: A review of clinical presentations and imaging spectrum. *AJR Am J Roentgenol*.2016; 207(2): W8-W18. PMID: 27145453.
5. Maceira E, Rochera R. Müller-Weiss Disease: Clinical and biomechanical features. *Foot Ankle Clin.* 2004; 9(1): 105–125. PMID: 15062217.
6. Hetsroni I, Nyska M, Ayalon M. Plantar pressure distribution in patients with Muller Weiss disease. *Foot Ankle Int.* 2007; 28(2): 237-241. PMID: 17296146.
7. Mayich D. The treatment of Mueller-Weiss Disease: A Systematic Approach. *Techniques in Foot & Ankle Surgery.* 2016; 15(2): 59–73.
8. Welck M, Kaplan J, Myerson M. Müller-Weiss syndrome. *Foot & Ankle Specialist.* 2016; 9(3): 245–251. PMID: 26847192.
9. Ahmed AA, Kandil MI, Tabl EA, Elgazzar AS. Müller-Weiss Disease: A topical review. *Foot Ankle Int.* 2019; 40(12): 1447–1457. PMID: 31538823.
10. Volpe A, Monestier L, Malara T, Riva G, Barbera G, Surace MF. Müller-Weiss Disease: Four case reports. *World J Orthop*. 2020; 11(11): 507–515. PMID: 33269217.
11. Fernández de Retana P, Maceira E, Fernández-Valencia J, Suso S. Arthrodesis of the talonavicular-cuneiform joints in Müller-Weiss disease. *Foot Ankle Clin.* 2004; 9(1): 65–72. PMID: 15062214.

FIGURES

Figure 1: Navicular bone osteonecrosis and associated findings on radiographs.
Most recent (a) weightbearing dorsoplantar, (b) oblique and (c) weightbearing lateral radiographs of the right foot show sclerosis and comma-shaped configuration of the navicular bone, suggestive of osteonecrosis, as well as longitudinal arch collapse and perinavicular osteoarthritis.

Figure 2: Normal navicular bone on radiographs.
Non weightbearing right foot radiographs of the same patient acquired 11 years ago show normal configuration and density of the navicular bone.

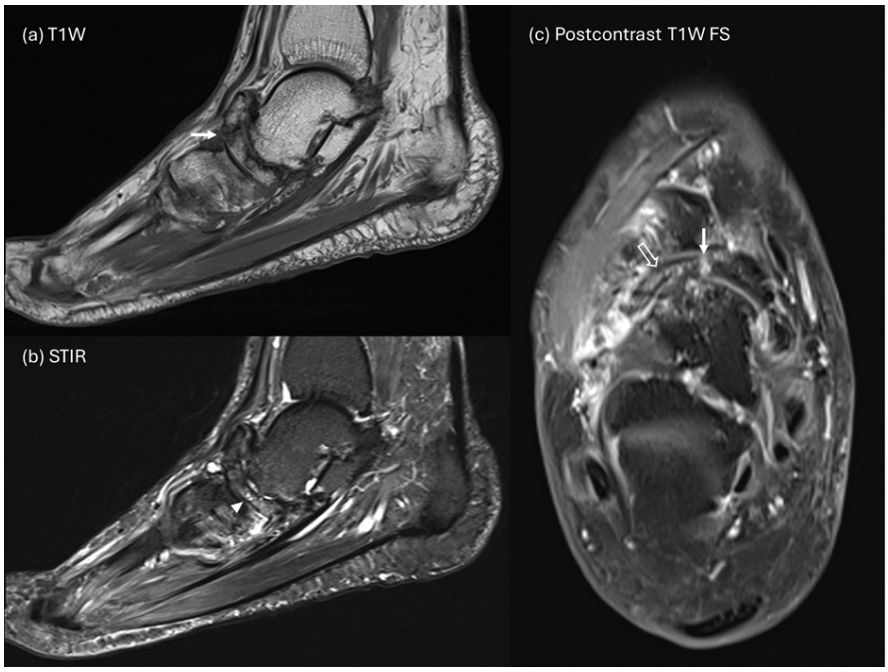

Figure 3: Navicular bone osteonecrosis on MRI.
(a) Sagittal T1-weighted (T1W), (b) sagittal short tau inversion recovery (STIR) and (c) coronal postcontrast T1W fat suppressed (T1W FS) sequences through the navicular bone show collapse and fragmentation (arrows in (a) and (c)). Marrow oedema-like signal (arrowhead in (b)) and enhancement (open arrow in (c)) in the lateral aspect of the navicular bone are probably related to fibrovascular tissue. Talo-navicular joint osteoarthritis is also noted.

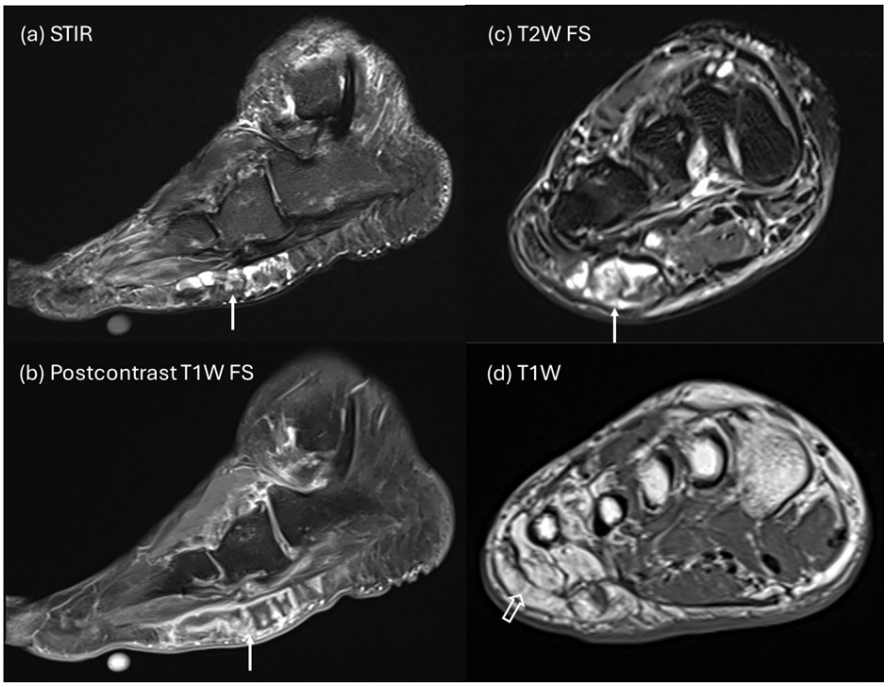

Figure 4: Ancillary findings on MRI related to altered mechanics.
(a) Sagittal STIR, (b) postcontrast T1W FS and (c) T2-weighted fat suppressed (T2W FS) sequences through the cuboid bone show a lobulated fluid collection with thin peripheral enhancement in the subcutaneous plane at the lateral plantar aspect of the midfoot, suggestive of adventitious bursitis (arrows).
(d) Short axis T1W image at the level of the proximal metatarsal shafts shows severe fatty replacement of the abductor digiti minimi (open arrow), as well as the adjacent flexor digiti minimi brevis, dorsal and plantar interossei muscles, which are probably related to chronic Baxter and lateral plantar nerve neuropathy.

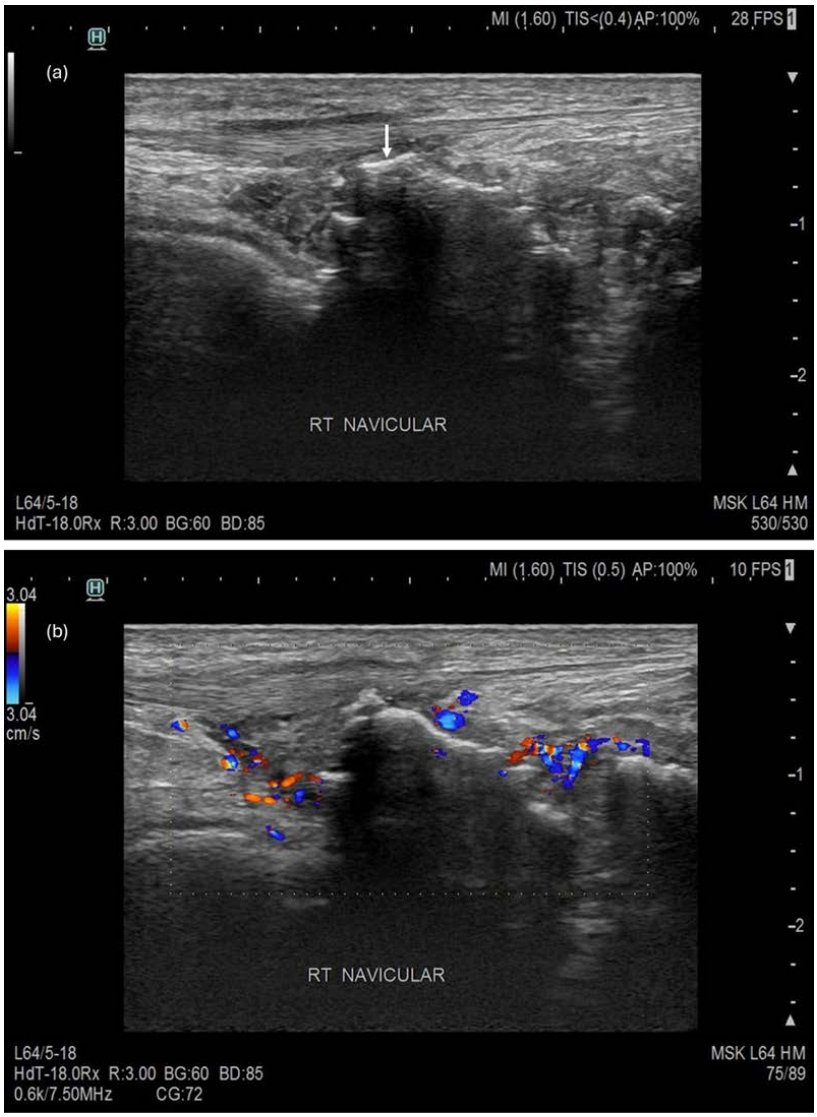

Figure 5: Navicular bone osteonecrosis on ultrasound.
(a) Greyscale ultrasound of the navicular bone shows cortical irregularity and fragmentation (arrow).
(b) Ultrasound Doppler shows hypervascularity surrounding the navicular, suggestive of synovitis.

SUMMARY TABLE

Etiology	Idiopathic
Incidence	Uncertain, likely underestimated as it can be misdiagnosed for other conditions such as perinavicular joint osteoarthritis
Gender ratio	Predominantly women
Age predilection	Between 40 and 60 years
Risk factors	Nutritional deficiencies, endocrinopathies, metabolic and malabsorption diseases
Treatment	Non-operative (analgesia, activity modification, footwear adjustment, orthotics, bracing, casting), surgical intervention (core decompression, internal fixation, arthrodesis)
Prognosis	Unknown
Findings on imaging	Collapse and sclerosis of the navicular with comma-shaped configuration and dorsomedial protrusion. Secondary peri-navicular degenerative changes.

DIFFERENTIAL TABLE

Differential diagnosis	Clinical features	Imaging findings
Kohler syndrome	• Occurs in children with male preponderance • Benign and self-limiting	• Collapse and fragmentation of the navicular, with patchy sclerosis
Primary osteoarthritis	• Prevalence increases with age • More common in older individuals while MWS often affects middle-aged adults	• Joint space narrowing, osteophyte formation, subchondral sclerosis and subchondral cysts • Bone collapse and fragmentation are not typical features
Neuropathic arthropathy	• Most common cause is diabetes mellitus • Usually painless	• Sclerosis and fragmentation of midfoot bones with joint disorganization and deformity • Vascular calcifications are an adjunctive finding
Fracture	• History of trauma or chronic overuse	• Acute traumatic fractures may involve the dorsum (avulsion type), body or tuberosity of the navicular bone, and can be classified using the Sangeorzan classification • Stress fractures tend to be linear and oriented in the sagittal plane
Osteomyelitis	• Clinical symptoms and signs of infection	• Cortical bone erosion and destruction, periosteal reaction • May have associated soft tissue collections

KEYWORDS

Mueller-Weiss syndrome; Spontaneous osteonecrosis; Comma-shaped navicular; Navicular collapse; Perinavicular osteoarthritis