Hong Kong J Radiol. 2016;19:57-62 | DOI: 10.12809/hkjr1615337

CASE REPORT

Mueller-Weiss Syndrome: an Important but Under-recognised Cause of Foot Pain and Deformity

CH Chan[1], KK Cheng[1], NY Pan[2], KC Wong[1], HY Lai[2], HF Chan[2], HL Chow[1]

[1]Department of Diagnostic and Interventional Radiology, Kwong Wah Hospital, Yaumatei, Hong Kong; [2]Department of Radiology, Princess Margaret Hospital, Laichikok, Hong Kong

ABSTRACT

Mueller-Weiss syndrome, or spontaneous osteonecrosis of the tarsal navicular bone in adults, is an important but under-recognised idiopathic foot condition that causes chronic mid- and hind-foot pain. Progressive deformity leading to advanced mid-tarsal osteoarthritis occurs when the disease is not recognised in its early stages. We present a case series of Mueller-Weiss syndrome from two regional hospitals in Hong Kong to illustrate the key radiographic features as well as computed tomography and magnetic resonance imaging findings accompanied by a review of the current literature, with the aim of increasing awareness of this important but under-recognised cause of foot pain in adults.

Key Words: *Foot bones; Magnetic resonance imaging; Osteonecrosis; Pain; Tarsal bones*

中文摘要

Mueller-Weiss綜合徵：引致足部疼痛和畸形的一個重要但未被廣泛認識的病因

陳志軒、鄭加勁、潘寧遠、黃國俊、黎曉欣、陳可鋒、仇鴻烈

Mueller-Weiss綜合徵是指成人足舟骨自發性壞死而導致中足和後足部慢性疼痛的病變。此症雖然是一個重要的足部病因，卻未被廣泛認識。如果未能早期發現，足部進行性畸形會導致中跗骨的嚴重骨性關節炎。本病例系列報告分別來自香港兩間分區醫院，探討Mueller-Weiss綜合徵的主要X綫特徵，以及電腦斷層掃描和磁力共振成像的表現；同時複習文獻，以提高對此成人足部疼痛病因的認識。

INTRODUCTION

Spontaneous osteonecrosis of the tarsal navicular bone in adults, also known as Mueller-Weiss syndrome, is a complex idiopathic foot condition. It causes chronic mid- and hind-foot pain and progressive deformity leading to advanced mid-tarsal osteoarthritis and subsequent permanent disability. It was first described by surgeon Walther Mueller in 1927 as chronic deformation of the tarsal navicular bone in adults[1] and was proposed by radiologist Konrad Weiss in the same

Correspondence: *Dr CH Chan, Department of Diagnostic and Interventional Radiology, 1/F, Kwong Wah Hospital, 25 Waterloo Road, Hong Kong.*
Tel: (852) 3517 7953; Fax: (852) 3517 5454; Email: cch626@ha.org.hk

Submitted: 15 Jun 2015; Accepted: 13 Aug 2015.

year to be secondary to spontaneous osteonecrosis.[2] This condition should not be confused with Köhler disease, which refers to osteochondrosis of the tarsal navicular bone in children and is a self-limiting condition with good outcome. We present a case series of Mueller-Weiss syndrome from two regional hospitals, in an attempt to increase awareness of this important but under-recognised cause of foot pain in adults.

CASE REPORTS

Case 1

A 74-year-old woman with previous bilateral total knee replacements for severe knee osteoarthritis presented with a 1-year history of left ankle pain in October 2006. Physical examination showed bilateral pes planus that was managed conservatively. Over the next several years, the patient developed bilateral medial mid-foot pain. Radiographs of both feet revealed comma-shaped tarsal navicular bones with collapse of the lateral portion, which was more severe in the left foot. Medial navicular protrusion and marked talonavicular osteoarthritis were also observed. Medial shift of the cuboid in relation to the calcaneus bone was noted in the left foot (Figure 1).

Figure 1. Dorsoplantar radiograph of both feet shows comma-shaped navicular bones with collapse of the lateral portion (white arrows). Medial shift of the cuboid in relation to calcaneus is observed in the left foot (black arrows).

Case 2

A 78-year-old woman with hypertension was admitted after experiencing bilateral lower limb swelling and pain for 1 month in January 2015. She was clinically diagnosed with cellulitis and subsequently responded to a course of antibiotic treatment. Incidentally, radiographs of both feet showed lateral collapse and medial protrusion of the navicular bone, with marked talonavicular joint osteoarthrosis (Figure 2).

Case 3

A 45-year-old man with type II diabetes mellitus and prior debridement and sequestrectomy for right fifth metatarsal osteomyelitis underwent radiographic examination for suspected Charcot arthropathy in June 2013. Collapse, sclerosis, and fragmentation were seen in the right tarsal navicular bone with dorsal and medial protrusion of the fragments. Mild sclerosis was also seen in the adjacent talar head and cuneiform bones (Figure 3a). Left foot radiographs were reported as normal but subsequent review of the oblique view showed subtle collapse of the lateral portion of the tarsal navicular bone (Figure 3b).

Case 4

A 72-year-old man with type II diabetes mellitus

Figure 2. Dorsoplantar radiograph of both feet shows symmetrical collapsed lateral portion of both navicular bones with medial protrusion (black arrows) and talonavicular joint osteoarthrosis.

presented 10 years ago with pain and swelling over the right mid-foot in May 2005. At that time, radiographs and magnetic resonance imaging (MRI) scans (no longer available) were suspicious of septic arthritis of the talonavicular joint and bone biopsy taken from the tarsal navicular bone showed bone necrosis during operation. More recent radiographs showed comma-shaped tarsal navicular bone with medial protrusion as well as peri-navicular osteoarthritic changes (Figure 4a). Computed tomography for preoperative assessment demonstrated fragmentation of the tarsal navicular bone, particularly of the lateral portion (Figure 4b), and pes planus.

Figure 3. Oblique radiographs of both feet show (a) fragmentation of the right tarsal navicular bone with dorsal and medial protrusion of the fragments (black arrows) as well as mild sclerosis in the adjacent talar head and cuneiform bones, and (b) subtle collapse of the lateral aspect of the left tarsal navicular bone (black arrow).

Case 5

A 57-year-old woman with severe osteoarthritis of both knees awaiting total knee replacement presented with bilateral severe flat feet without pain in April 2013. Radiographs of both feet showed a comma-shaped tarsal navicular bones. Fragmentation was also seen in the left navicular bone (Figure 5a). Computed tomography subsequently performed for preoperative assessment of pes planus showed similar findings (Figure 5b).

Case 6

A 57-year-old woman presented with medial foot pain and physical examination revealed rigid flat foot as well as localised tenderness and swelling at the region of the posterior tibialis tendon in January 2014. Initial radiographs were reported as normal. MRI for further assessment of posterior tibialis tendon dysfunction in the right ankle revealed a flattened lateral portion of the tarsal navicular bone with associated bone marrow oedema and subchondral cysts as well as extensive

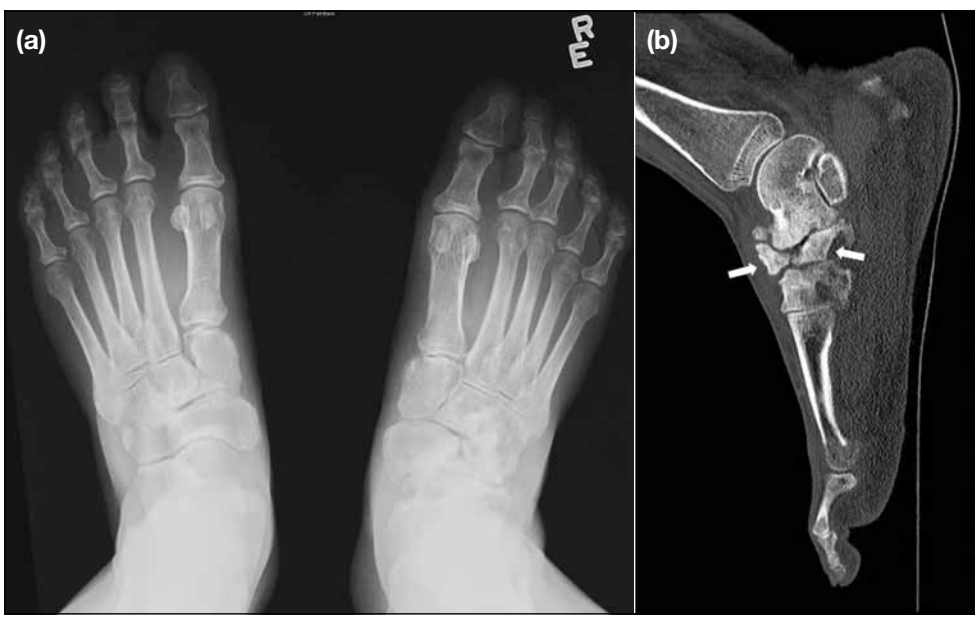

Figure 4. (a) Dorsoplantar radiograph of both feet shows comma-shaped right tarsal navicular bone with sclerosis of its lateral portion. A normal left navicular bone is shown for comparison. (b) Sagittal computed tomography image demonstrates fragmentation of right tarsal navicular bone (white arrows). Prior bone biopsy of the right tarsal navicular bone showed necrosis.

peri-navicular degenerative changes (Figures 6a to 6c). Subsequent review of previous radiographs showed mild lateral collapse and mild medial protrusion of both tarsal navicular bones (Figure 6d).

Figure 5. (a) Oblique radiograph of left foot shows fragmentation of the tarsal navicular bone (black arrows). (b) Sagittal computed tomography image demonstrates fragmentation of the lateral portion of the left tarsal navicular bone between the tarsal head (white arrow) and lateral cuneiform bone (black arrow).

A summary of our six cases is shown in the Table.

DISCUSSION

The tarsal navicular bone is a key contributor to the structural stability of both medial longitudinal and transverse arches of the foot. The plantar aspect of the navicular bone is supplied by the medial plantar artery, while its dorsal aspect and lateral third are supplied by the dorsalis pedis artery. Anastomosis between these two arteries supplies the medial tuberosity of the navicular bone. As a result, the central zone has a relatively poor blood supply but at the same time, it is the area of maximal shear force.[3,4] This combination of factors renders the navicular bone especially susceptible to repetitive injury and ischaemia.

The exact aetiology of Mueller-Weiss syndrome remains controversial. Excessive mechanical strain, both compressive and tensile, that disrupts the navicular blood supply is proposed as a cause for spontaneous osteonecrosis of the tarsal navicular bone.[3,5] A large series of this disease conducted by Maceira and Rochera[6] proposed a combination of delayed ossification of tarsal navicular and lateral shift of compressive force distribution on the navicular bone as a causative mechanism. More recently, Mohiuddin et al[4] proposed that the syndrome occurs secondarily to undiagnosed navicular stress fractures, but this requires verification with further research. Nevertheless, further study is needed to determine the exact aetiology of the disease.

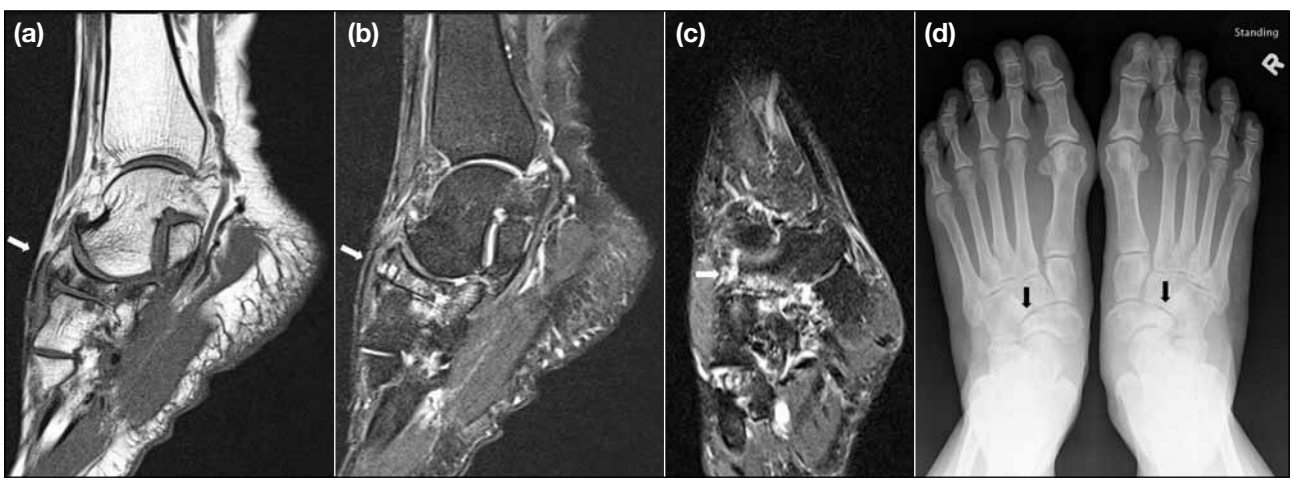

Figure 6. (a) Sagittal T1-weighted, (b) sagittal T2-weighted, and (c) coronal T2-weighted magnetic resonance images show flattened lateral portion of the right tarsal navicular bone with associated bone marrow oedema and subchondral cysts, as well as extensive peri-navicular degenerative changes (white arrows). (d) Dorsoplantar radiograph of both feet demonstrates mild collapse of the lateral portion of bilateral tarsal navicular bones (black arrows) with mild medial protrusion of the bones.

Table. Summary of patient data: demographics, radiological features, and associated clinical findings.

Case No.	Age (years)	Gender	Bilateral involvement	Comma-shaped navicular	Fragmentation of navicular	Medial protrusion of navicular	Peri-navicular osteoarthrosis	Presence of pes planus
1	74	F	Yes, asymmetrical	+	-	+	+	+
2	78	F	Yes, symmetrical	+	-	+	+	-
3	45	M	Yes, asymmetrical	+	+	+	+	-
4	72	M	No, only on right side	+	+	+	+	+
5	57	F	Yes, asymmetrical	+	+	+	+	+
6	57	F	Yes, symmetrical	+	-	+	+	+

Abbreviations: F = female; M = male; + = presence; - = absence.

The precise prevalence of Mueller-Weiss syndrome is unknown, but the condition is probably under-recognised owing to a low clinical index of suspicion and radiographic features that overlap with degeneration, infection, and tarsal coalition (that also produces prominent peri-navicular osteoarthrosis). In general, women are more commonly affected (73% of cases) than men.[6] Most patients develop insidious pain in the fourth and fifth decade, with a mean age of 47 years.[6] On physical examination, hindfoot varus is a hallmark that occurs secondary to lateral displacement of the talus relative to the calcaneus.[7] As a result of concurrent medial protrusion of the navicular bone, however, navicular tuberosity is also prominent on the medial side of the foot and may produce the false impression of hindfoot valgus.[7] This combination of apparent hindfoot valgus and true hindfoot varus is also known as paradoxical pes planovarus.[7]

To diagnose Mueller-Weiss syndrome, clinical examination and weight-bearing radiographs are usually sufficient.[4] Computed tomography is helpful for assessment of bone stock and preoperative planning. MRI is mainly reserved to assess the status of soft tissue structures, especially the tibialis posterior tendon and spring ligament in associated pes planus. MRI also allows exclusion of differential causes of mid- and hind-foot pain such as stress fracture and infection.[4]

While the initial radiographic findings in Mueller-Weiss syndrome are subtle loss of volume and increased radiodensity in the lateral portion of the tarsal navicular bone,[5] most of the patients presented at the time of diagnosis with comma- or hourglass-shaped tarsal navicular bone due to collapse of its lateral portion and internal rotation of its medial portion. This distinctive radiographic finding was present in all patients in our case series. Other described common findings include fragmentation, dorsal and / or medial protrusion of navicular bone with secondary lateral shift of the talar head and hindfoot varus,[4,5] and early or significant peri-navicular osteoarthritis.[4,6] Medial protrusion of the navicular bone and peri-navicular degenerative changes were evident in all patients in our series. Fragmentation of the navicular bone was present in three (50%) patients. The 'cuboid sign', which refers to a medial shift of the cuboid in relation to the calcaneus bone, is another frequently associated finding, occurring in five of our six cases (except case 3) in varying degrees of severity. Other associated imaging findings include a widened appearance of the talar head (due to osteoarthritis) and hypertrophied second metatarsal (due to lateralisation of compressive forces from the first to second metatarsal).[6] Large open sinus tarsi reflecting a supinated hindfoot may also be seen.[4] As disease progresses, fragmentation of the dorsolateral navicular bone creates enough space for the talar head to acquire a plantar flexed position, leading to pes planus varus[4,6]—a finding that was evident in four of our six cases.

Maceira and Rochera[6] has described five stages of Mueller-Weiss syndrome based on weight-bearing lateral radiographs but admitted that these stages are for descriptive purposes and may not correspond to clinical severity.

No specific computed tomographic findings have been described for Mueller-Weiss syndrome in addition to those visible in radiographs. MRI findings have not been frequently described in the literature. It may show non-specific marrow oedema, which is more prominent in the lateral portion of the tarsal navicular bone. This can be accompanied by small effusions within the peri-navicular joints.[5]

Similar radiographic findings of comma-shaped configuration, fragmentation, and medial and / or dorsal protrusion of the tarsal navicular bone can

also be seen in secondary osteonecrosis of the tarsal navicular bone in patients with rheumatoid arthritis, chronic renal failure, systemic lupus erythematous, and trauma. Discrimination of primary (spontaneous) from secondary osteonecrosis is not possible by radiologic means alone, although a bilateral occurrence, particularly in women, favours the diagnosis of spontaneous disease.[5] Exclusion of possible underlying causes of osteonecrosis, including steroid use and vasculitis, is needed before diagnosing Mueller-Weiss syndrome. In late stage, radiographic features of Mueller-Weiss syndrome and stress fracture of the tarsal navicular bone are almost indistinguishable from each other and some authors have proposed undiagnosed tarsal navicular stress fracture as the aetiology of Mueller-Weiss syndrome.[4] At an early stage, stress fracture of the tarsal navicular bone classically involves the central third of the proximal dorsal margin of the tarsal navicular bone[8] in young athletes.

Most histopathological studies cannot provide conclusive evidence of osteonecrosis, even though Mueller-Weiss syndrome has been widely described as osteonecrosis or avascular necrosis of the navicular bone.[4] A single case report by Tan et al[9] provided histologic proof of avascular osteonecrosis. In one of our cases (case 4), we also had histologic proof of bone necrosis in the tarsal navicular bone with subsequent imaging features compatible with Mueller-Weiss syndrome.

Currently, there is no gold standard treatment for Mueller-Weiss syndrome. Most authors support initial non-surgical treatment including anti-inflammatory / analgesic medication and possible non–weight-bearing cast immobilisation. Surgical interventions are considered if conservative management fails. Core depression can be attempted in early stages of the disease.[10] Some authors have also reported excision of diseased navicular bone with reconstruction by bone grafting.[9,11] Arthrodesis of various tarsal joints has been used for cases associated with significant painful peri-navicular osteoarthritis.[4]

CONCLUSION

Mueller-Weiss syndrome is an important but under-recognised cause of mid- and hind-foot pain, and a differential cause of pes planus. Clinical examination and weight-bearing radiographs are usually sufficient to make a diagnosis. Key radiographic features of this condition include collapsed lateral aspect of the tarsal navicular bone with consequent comma-shaped appearance. Subsequent medial and / or dorsal protrusion and even fragmentation of the tarsal navicular bone leading to progressive deformity in the mid- and hind-foot including pes planus and hindfoot varus are also common findings. Attention to possible underlying causes of secondary osteonecrosis of the navicular bone is needed before making the diagnosis of Mueller-Weiss syndrome.

REFERENCES

1. Mueller W. Ueber eine eigenartige doppelseitige Veraenderung des os naviculare pedis beim Erwachsenen. Dtsch Z Chir Leipzig. 1927;201:84-9. cross ref
2. Weiss K. Uber die “malazie” des os naviculare pedis. Fortschr Geb Rontgenstr. 1927;45:63-7.
3. Scott-Moncrieff A, Forster BB, Andrews G, Khan K. The adult tarsal navicular: why it matters. Can Assoc Radiol J. 2007;58:279-85.
4. Mohiuddin T, Jennison T, Damany D. Müller-Weiss disease — review of current knowledge. Foot Ankle Surg. 2014;20:79-84. cross ref
5. Haller J, Sartoris DJ, Resnick D, Pathria MN, Berthoty D, Howard B, et al. Spontaneous osteonecrosis of the tarsal navicular in adults: imaging findings. AJR Am J Roentgenol. 1988;151:355-8. cross ref
6. Maceira E, Rochera R. Müeller-Weiss disease: clinical and biomechanical features. Foot Ankle Clin. 2004;9:105-25. cross ref
7. Arranz JC, Maceira ES, Monteagudo de la Rosa M, Herrera LH, Gomez Herrera JJ, Recio M et al. Müeller-Weiss disease. Greatly unknown, great pretender. European Society of Radiology. Education Exhibit, ECR 2011. DOI: 10.1594/ecr2011/C-1797.
8. Kiss ZS, Khan KM, Fuller PJ. Stress fractures of the tarsal navicular bone: CT findings in 55 cases. AJR Am J Roentgenol. 1993;160:111-5. cross ref
9. Tan A, Smulders YC, Zöphel OT. Use of remodeled femoral head allograft for tarsal reconstruction in the treatment of Mueller-Weiss disease. J Foot Ankle Surg. 2011;50:721-6. cross ref
10. Janositz G, Sisák K, Tóth K. Percutaneous decompression for the treatment of Mueller-Weiss syndrome. Knee Surg Sports Traumatol Arthrosc. 2011;19:688-90. cross ref
11. Tosun B, Al F, Tosun A. Spontaneous osteonecrosis of the tarsal navicular in an adult: Mueller-Weiss syndrome. J Foot Ankle Surg. 2011;50:221-4. cross ref