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Mueller-Weiss Syndrome: A Radiological Diagnosis

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ABSTRACT

Mueller-Weiss syndrome is an uncommon condition characterized by the spontaneous development of osteonecrosis in the navicular bone of adults, with no known cause. It predominantly affects women, either on one side or both sides, and can result in significant functional impairment. The diagnosis is typically made based on clinical observations of mechanical-type pain on the upper surface of the middle foot and is confirmed through radiological examinations.

Keywords: Mueller-Weiss syndrome; Navicular bone; Standard X-ray, CT-scan

Introduction

Osteonecrosis of the navicular bone, also known as Muller-Weiss syndrome, is a rare condition described in the literature in 1927 by Walther Mueller. It occurs in adults between 40 and 60 years of age and is more frequent in women¹. Its varied clinical presentation is dominated by midfoot and ankle pain. The typical radiological image is a comma-shaped aspect of the navicular bone².

The aim of this case is to present the radiological findings of this disease.

Discussion

Mueller-Weiss syndrome is a rare condition that often affects females. It is characterized by osteonecrosis of the navicular bone and its compression by the talus and lateral cuneiform¹.

First described by Muller and Weiss in 1927 yet its pathophysiology is still a matter of debate. According to some authors, the necrosis may be back up to the mechanical compression of the navicular bone caused by trauma or increased pressures on the midfoot arch in flat feet because of being overweight. Others assume that the unstable vascularization of the navicular bone is the cause of osteonecrosis¹.

Most patients report experiencing a chronic mechanical pain of gradual onset accompanied by a deformation of the dorsal part of the midfoot. Their physical examination recovers tenderness and dorsomedial foot swelling and varus deformity of the heel.

Standard X-ray is the first line radiological exploration, including weight bearing anteroposterior and lateral views, which reveals a characteristic appearance:

- Ankle and hindfoot: the collapse of the lateral half of the navicular bone engenders a medial subluxation of the talus head which provokes the hindfoot varus (**Figure 1**).
- Midfoot: when the navicular bone's lateral half falls down, it becomes sclerotic and takes the aspect of a comma or hourglass (Figure 2). This along with the aforementioned medial subluxation of the talus head, creates a talo-navicular articulation.
- Forefoot: as the arch of the foot collapses, the metatarsals align in parallele and hypertrophic changes occur in the second metatarsal due to the force of compression exercised by the second metatarsal instead of the first one and the tarsometatarsal articulations.



Figure 1. Oblique radiograph of the forefoot showing the collapse of the lateral part of the navicular bone and the medial subluxation. (Red arrow).



Figure 2. Lateral radiograph of the forefoot revealing a sclerosis and densification of navicular bone (Red arrow).

Maceira and Rochera had also provided a descriptive staging system with 5 degrees of deformity for the Mueller Weiss syndrome as assessed on weight bearing lateral radiographs. The degree of deformity is defined by the navicular bone aspect and the Meary-Tomeno angle (the angle formed between the longitudinal axes of the talus and the first metatarsal). In a normal foot, these axes are aligned. A Meary-Tomeno angle superior to 4° convex downwards indicates a flat foot².

CT scan is also useful for diagnosis and plays an important role in the preoperative assessment by evaluating the bone's structure and their mineralization (**Figures 3,4**).

MRI can show oedema on STIR / DP FS (short T1 inversion recovery / density proton with fat saturation) images and is more precise in detecting early changes because of its ability to detect signal change in the bone marrow, thereby excluding differential diagnosis as fatigue fracture (overuse fracture) and infections^{2,3}.

The therapeutic strategy is not unified among authors. However, the majority of publications advocate for an initial phase of medical treatment, weight loss, oral anti-inflammatory drugs and immobilization with orthoses or plaster for a few weeks. The chirurgical treatment depends on the severity of symptoms and is reserved for cases where medical treatment fails or for stage 3 and above³.



Figure 3. Sagittal CT scan of the forefoot demonstrating the comma aspect of the navicular bone (Red arrow).



Figure 4. Coronal CT scan illustrating the collapse of the lateral part of the navicular bone (Red arrow) associated with medial subluxation.

Conclusion

Mueller-Weiss syndrome is a rare, under-diagnosed and multifactorial condition. It can rapidly lead to deformities and disability. However, a better understanding of this disease and its radiological manifestations could allow for earlier diagnosis and better future management.

Conflicts of Interest

This study does not have any conflict of interest.

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