

Muller Weiss syndrome –Demystifying the conundrum

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Abstract

Background: Muller-Weiss disease (MWD) is a rare entity that refers to spontaneous avascular necrosis of tarsal navicular bone in adults [1, 2]. This is usually underrecognised and hence treatment guidelines are unclear. Here we report a case of Muller Weiss syndrome that presented in its early stages and underwent open drilling and decompression and cancellous bone grafting, with a two year follow up.

A 32 year old gentleman presented with pain over the dorsal aspect of left foot of four months duration. The pain got aggravated on weight bearing walking. There was no history of any trauma or constitutional symptoms. On clinical examination, there was tenderness over the navicular bone. Radiographs showed sclerosis of the navicular bone with a subtle lucency (Figure 1). CT confirmed a fracture in the navicular bone (Fig 2 and 3). A possibility of spontaneous avascular necrosis of navicular bone is considered.



Figure 1: Radiographs showed sclerosis of the navicular bone with a subtle lucency

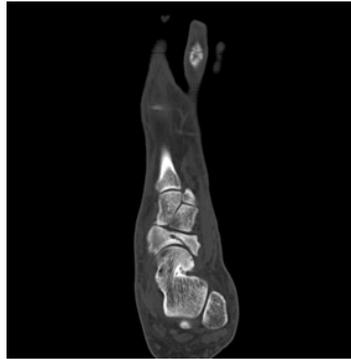


Figure 2: CT confirmed a fracture in the navicular bone.



Figure 3: CT confirmed a fracture in the navicular bone.

Treatment

It was decided to start on conservative treatment in the form of immobilisation in a below knee cast for two months. Evaluation after two months showed persistent pain over navicular bone. Immobilisation was given for one more month. However his symptoms did not improve. Hence it was decided to proceed with surgical intervention.

Navicular bone was exposed through a 5 cm long lazy 'S' incision. The oblique fracture in navicular was an incomplete one. Multiple drill holes were made on either side of the fracture with a 2 mm drill bit. Drill holes were made through the fracture interface also. The navicular bone appeared hard and sclerotic. Freshened up the fractured surface of navicular bone. Packed the gap with fine cancellous bone graft harvested from ipsilateral iliac crest. No internal fixation was considered as the navicular bone was stable after packing with bone graft. No periarticular bony changes were noticed intraoperatively. The wound was closed and below knee cast was applied with the advice of non-weight bearing for two months. Cast was removed after two months and weight bearing walking started. He had mild pain for next two months. On further follow up, it was noticed that he could resume his normal work and he became fully asymptomatic. Follow up radiographs after two years showed a relatively normal appearance of navicular bone (Figure 4).



Figure 4: Follow up radiographs after two years showed a relatively normal appearance of navicular bone

Discussion

Spontaneous avascular necrosis of navicular bone in adults is relatively uncommon and is termed as Muller Weiss syndrome. In 1927, Mueller recognised this condition of navicular bone and attributed it to compression from adjacent bones. In 1929, Weiss proposed osteonecrosis as the primary cause of this presentation. It is characterised by collapse of the lateral portion of the navicular bone with a medial protrusion of the talar head and perinavicular osteoarthritic changes [3]. It is usually bilateral and occurs more commonly in women. The counterpart of similar presentation in children is called Kohler's disease. But it is usually unilateral and males are more commonly affected.

Various theories have been proposed regarding the etiology of Muller Weiss syndrome. The blood supply of the navicular is precarious. The dorsal and volar parts of the bone are separately supplied by the branches of the dorsalis pedis artery and medial plantar artery. Thus, the central portion of navicular is hypovascular. A compromised peripheral blood flow coupled with repetitive stresses can lead to osteonecrosis of this bone.

Patients usually present with a dull aching pain over the dorsum of foot without any history of significant trauma. Heel varus is typical in MWS [4]. The medial prominence of navicular gives an impression of hindfoot valgus, which is an apparent one. Radiological findings include medial or dorsal deviation of part or the entire navicular bone, and a comma-shaped deformity due to the collapse of the lateral portion of navicular; increased radiodensity, fragmentation and fractures [5]. CT scan gives a better assessment of morphology of navicular bone and adjoining joints. It helps to delineate the fracture which may not be obvious in routine radiographs. Maceira and Rochera [4] reported five stages of MWS based on the Meary-Tomeno angle (intersection of the talar axis and first metatarsal axes) in the lateral weight bearing view of the foot. The downsides of this staging system are poor prognostic value and poor interobserver reliability.

Surgical intervention needs to be considered when there are persisting symptoms even after a fair trial of conservative treatment. There is no gold standard treatment for MWS. Various treatment options have been reported. Simple excision of the dorsolateral fragment of navicular with bone graft, excision of entire navicular with autogenous cortical bone graft, percutaneous drilling and decompression [6], talonavicular arthrodesis,

talonavicular-cuneiform arthrodesis [7] and triple arthrodesis and were reported with varying outcomes. For cases detected in the early stages, drilling and decompression or simple excision with bone grafting will suffice. For cases identified in the later stages, isolated talonavicular arthrodesis can give gratifying results. More advanced stages with significant arthritic changes in the adjoining joints, poses a great challenge for even the most experienced surgeon. Talonavicular-cuneiform arthrodesis or triple arthrodesis is warranted in such situations.

In our case, patient presented well before perinavicular arthritic changes set in. The changes were confined to tarsal navicular. Hence drilling, decompression and cancellous bone graft packing of the fracture site yielded a favourable outcome. Early detection and timely surgical intervention gives good results in MWS.

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