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Pigmented Villonodular Synovitis of the Ankle and MRI: A Case Report

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Abtract: Although most common in the hip and knee joints, pigmented villonodular synovitis (PVNS) has also been reported to occur in the ankle. The distinctive appearance of PVNS on magnetic resonance imaging (MRI) has proven to be useful in both the diagnosis and treatment of this pathologic process. The following is the report of a case that illustrates the utility of MRI in determining the extent of soft tissue and osseous involvement of PVNS about the ankle. This information is especially helpful in planning subsequent surgical intervention.

Introduction

Pigmented villonodular synovitis (PVNS) is a locally aggressive disease process of uncertain etiology affecting the linings of joints, tendon sheathes, and bursae. Originally described in 1941 by Jaffe et al., this phenomenon affects 1.8 patients per million population, and presents most commonly in the knee and hip joints followed by the ankle and shoulder [7,10]. Whereas the onset of this process is insidious in nature, if left untreated, it can become expansive involving adjacent osseous tissue. Although diagnosis in the past relied on combined clinical, radiographic, gross, and histological analyses, magnetic resonance imaging (MRI) has provided a useful means of diagnosis based on the relatively unique appearance of this lesion on T1- and T2-weighted images [3,8,9]. The following case is unique in comparison to previous reports of PVNS involving the ankle joint in that the extent of its soft tissue involvement with relative sparing of surrounding osseous anatomy. MRI in this instance is especially helpful, not only in the diagnosis of this clinical condition, but also in the preparation for surgical treatment .

Case Report

This 29-year-old male presented with complaints of pain, swelling, and instability of his right ankle that had progressed in both intensity and frequency over the last year. The patient's past medical history is significant for a right ankle fracture 11 years ago that was treated by closed reduction and immobilization. Subsequently, the patient developed instability of this ankle, and successfully underwent a lateral stabilization procedure 1 year later. At this procedure a nidus of PVNS was found and excised. Post-operatively, the patient had recalled intermittent swelling for 1 year's duration, but denied any episodes of recurrent instability or pain until 12 months ago when his present symptoms developed. The patient described the pain as insidious, but more intense with weight-bearing and activity. He denied any recent trauma and stated that with immobilization the pain resolved, but that the swelling had become persistent. The patient subsequently underwent an MRI scan after routine X-rays of his ankle appeared normal. T-1 weighted images performed in the sagittal and axial planes demonstrated a diffuse hypodense infiltrative lesion involving the soft tissue structures about both sides of the ankle. The osseous anatomy appeared relatively spared from this pathologic process despite its intra-articular involvement. This decreased signal intensity appeared most predominant on the lateral aspect of the ankle; however, medial involvement was also noted at this level and more distally involving the hindfoot.

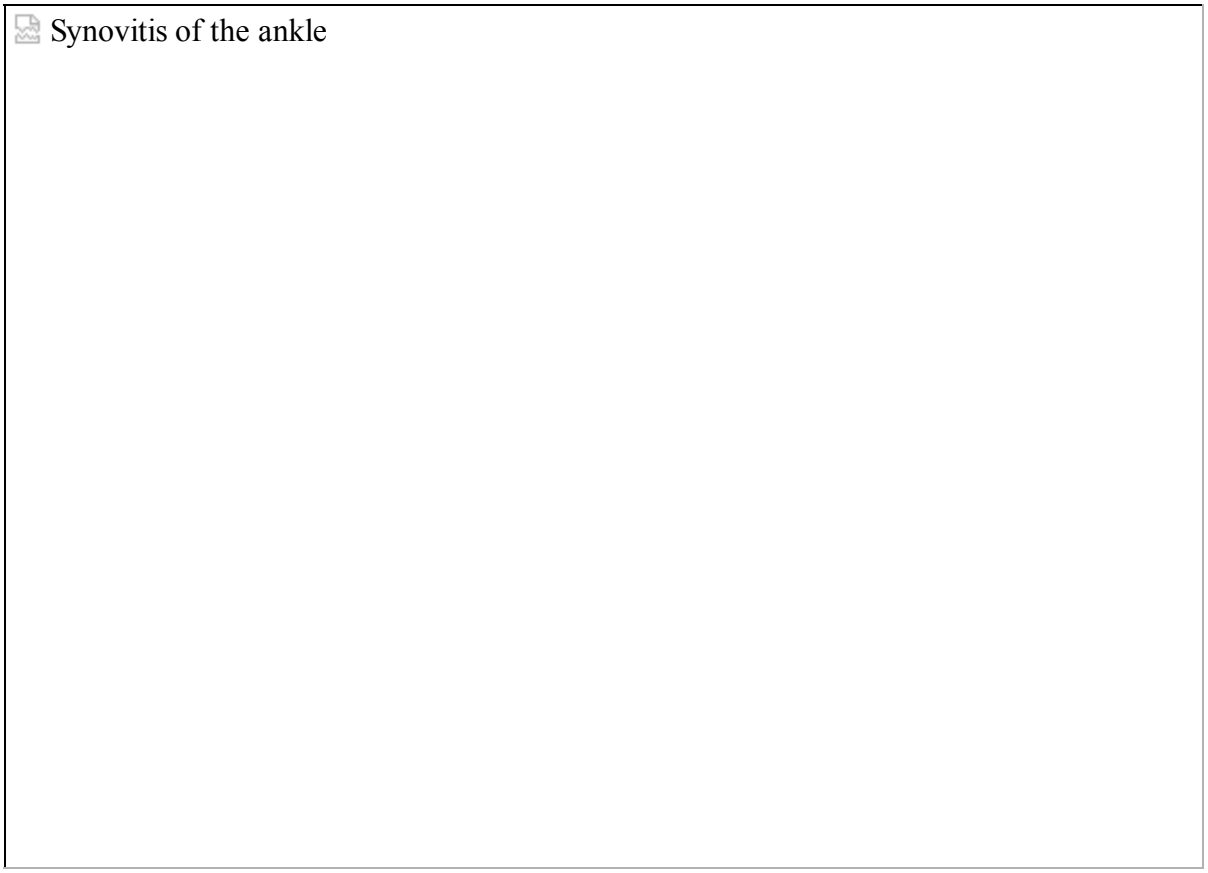

Fig. 1. (a) Sagittal T-1 weighted image demonstrating diffuse areas of decreased signal intensity involving the medial and lateral aspects of the ankle. **(b)** Axial T-1 weighted image displaying hindfoot involvement in the regions of the sustentaculum tali and sinus tarsi.

(Figure 1 a,b) These findings were consistent with PVNS. The patient then underwent

open synovectomy and resection of this mass (at an outside institution) through a lateral approach. Upon dissection through the subcutaneous layer, an infiltrative mass of brownish-yellow tissue was noted in both peroneal tendons. This process appeared to infiltrate extensively both proximally and distally. The abnormalities tracked proximally within the course of the peroneal tendons to 10 cm above the ankle at the junction of the mid and distal third of the tibia. There was also destruction of the peroneal brevis tendon. Additionally, the mass appeared to involve both the ankle and subtalar joints and extended distally into the hindfoot, to the area of the sinus tarsi. This process infiltrated and destroyed the capsular attachments at the tibiotalar joint. Lateral excision was performed until no further dystrophic tissue was visualized. Medial exploration was then performed. Involvement was found in both the posterior tibial and the flexor digitorum longus tendons. Medial synovectomy with mass excision was additionally performed to a grossly clear margin. There did not appear to be any osseous involvement of the tibia or talus, either medially or laterally. Frozen section obtained at the time of surgical exploration was consistent with giant cell tumor. Pathology specimens of tendon with their associated sheathes and synovium were sent for analysis. At gross examination, multiple fragments of yellow, red and tan, dense, mottled tissue was received measuring 11.5 × 8.5 × 3.0 cm in aggregate. The cut surfaces displayed areas of soft, rusty-red, friable, granular material admixed with white, dense tissue. Microscopic analysis revealed synovial tissue with several areas displaying a focal villous architecture. The predominant cellular constituents were large mononuclear cells with abundant, eosinophilic cytoplasm, and ovoid nuclei consistent with epithelioid histiocytes. Interspersed amongst these histiocytes were multiple multinucleated giant cells and scattered small lymphocytes (Figure 2).

Fig. 2. High powered magnification (100×) histologic section demonstrating multi-nucleated giant cell with coarse hemosiderin uptake both intra- and extra-cellularly.

Coarse, granular hemosiderin pigment was present both intra- and extra-cellularly. Varying amounts of collagen bands were also identified dividing the sheets of epithelioid histiocytes. Focally, sheets of foamy histiocytes, were also readily identified. These features were consistent with those described for pigmented villonodular synovitis [2].

Post-operatively, this patient continues to have mild discomfort at 6 months. Clinical examination reveals 10 degrees of dorsiflexion and 40 degrees of plantar flexion with mild discomfort in this range, and associated mild instability in the anteroposterior plane to an anterior drawer test. The patient is presently bearing full weight in conventional shoes and may require a stabilization procedure in the future for his instability.

Discussion

PVNS is most commonly a unilateral, monoarticular arthritis that affects adults in the third or fourth decade. Its onset is insidious in presentation, and clinically the patients present with symptoms of local discomfort and swelling. Occasionally, areas of erythema, stiffness, and a palpable mass have been identified. Less than 50% of patients recall any previous trauma to the affected region [3,10]. Whereas the knee, flexor tendon sheaths of the hand, and hip are the most common anatomic sites of involvement, Rao et al. estimated its incidence in the ankle joint at 2.5% [11]. Reports of this process also occurring in the subtalar joint, tarsal bones, and toes are also present in the literature [11,12,14,15].

Granowitz et al. subclassified PVNS into three categories based on clinical presentation [6]. The first type is an isolated, discrete lesion occurring within a tendon sheath, most often seen in the hand. A second type, termed localized pigmented villonodular synovitis (LPVS), is a localized discrete intra-articular lesion occurring most commonly in the knee and presenting most commonly with mechanical symptoms. Lastly, a diffuse entity is described that presents with chronic edema and pain, most commonly in the knee, hip, and ankle. This is termed diffuse pigmented villonodular synovitis (DPVS). All three categories display similar histology including villous synovial proliferation with microscopic villi, histiocytes, foam cells, and multi-nucleated giant cells [1]. However, in the localized presentation, the synovial tissue does not display a reactive hyperplasia distinct from the central disease process, as is seen in the diffuse variant [10,11].

A single testing modality that is both sensitive and specific for diagnosing PVNS presently does not exist. A combination of clinical, radiographic, and histologic findings is necessary to make the diagnosis with certainty. Plain radiographs alone are not confirmatory of this diagnosis despite the proper clinical presentation. Smith et al. reported that only 33% and 25% of those patients with diffuse and localized disease, respectively, displayed radiographic cysts or erosions secondary to osseous involvement [13]. Additionally, the patients with radiographic changes were those with more extensive disease involving bone. Recently, MRI has provided a very valuable addition to non-invasively identifying these lesions [1,5,8,9,14]. Findings on MRI, pathognomonic to PVNS, are likely attributable to the hemosiderin deposition in the affected tissues. Hemosiderin causes a decrease in signal on both T1- and T2-weighted images. Although very sensitive in diagnosing these lesions, MRI is non-specific, and is often confused with rheumatoid arthritis or soft tissue sarcoma [1]. Clinical correlation in these cases is utilized to differentiate between these different pathologic disease processes.

This particular case demonstrates the role of MRI in both the diagnosis and treatment of PVNS. This case is unique in the extent of the pathologic process described, and clearly illustrates the utility of MRI in the subsequent surgical decision making process. MRI in this case allowed precise definition of this lesion's extent into the posterior and lateral compartments of the leg proximally, more specifically, its involvement within the posterior tibialis and flexor digitorum longus posteriorly, and the peroneal tendons and muscle laterally. Such extensive involvement necessitated an open procedure on both sides of the joint to ensure complete excision of the mass and the affected surrounding tissues. This information has implications on the operative success of synovectomy, as complete removal with synovectomy is the preferred treatment for PVNS. Incomplete removal will most likely result in recurrence of this process [3,4]. Based on the MRI findings in this case, arthroscopy, or a more limited surgical approach, were both excluded as a treatment plans, secondary to the extensive nature of the underlying process. Additionally, MRI helped confirm the absence of osseous infiltration into the

distal tibia or talus.

The patient currently appears improved over his preoperative functional status with a diminished pain intensity and character at 6 months after operative intervention. His persisting instability may, however, require future intervention.

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