Diffuse Pigmented Villonodular Synovitis of the Ankle With Severe Bony Destruction: Treatment of a Case by Surgical Excision With Limited Arthrodesis

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Pigmented villonodular synovitis (PVNS) is a relatively rare disease affecting the synovial-lined joints. It was first fully described in 1941, by Jaffe and colleagues,[1] as a benign inflammatory state of the synovium of unclear etiology and as a tumorlike aggression of synovial tissue involving joints. The disease can be diffuse or localized and either intra-articular or extra-articular.[2] Diffuse PVNS involves the entire joint synovium; localized PVNS involves a discrete nodular, lobulated mass.[3] The disease commonly occurs in the knee joint, whereas involvement of the foot and ankle is rare[4] (reported incidence, ~1.8/million[5]). Severity of bony involvement in PVNS of the ankle may be high, possibly because the pressure erosion easily occurs in the narrow joint space of the ankle joint.[6] The recurrence rate for the diffuse type may be as high as 50%, whereas the rate for the localized type is considered low.[2,5,7] So, to avoid tumor spread, complete excision and careful tissue handling are essential. However, a more curative approach causes more structural morbidity to the joint, which may necessitate a more invasive procedure, such as talocrural arthrodesis, depending on location of the lesion. Thus, the surgeon faces a difficult choice between aggressive surgery and more conservative treatment.

Here we report a case of diffuse PVNS of the ankle with severe bony destruction, most of which originated in the distal tibiofibular joint (DTFJ). As this rare location was accessible, complete resection and arthrodesis only of the DTFJ were sufficient for curative operation. The authors have obtained the patient's written informed consent for print and electronic publication of the case report.

Case Report

A woman in her early 40s was referred to us with a 1-year history of slowly increasing right posterolateral ankle swelling and pain. Ankle pain increased with walking. The patient had no history of trauma. Examination revealed swelling over the posterior ankle, but the skin overlying the mass appeared normal. Range of motion and stability of the ankle were normal.

Plain radiographs and computed tomography showed severe bony destruction of the DTFJ in the right ankle (Figure 1). Magnetic resonance imaging (MRI) showed not only a large multinodular mass in the posterolateral aspect of the ankle but also a few small masses in the posteromedial aspect. The lateral mass connected with the DTFJ, and the medial mass connected with the medial talocrural joint cavity. There were low signals on

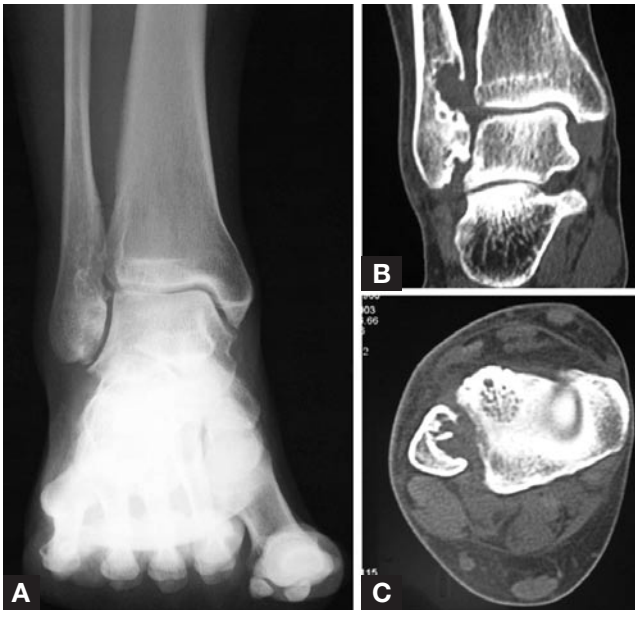

Figure 1. (A) Anteroposterior radiograph shows osteolytic change in distal tibiofibular joint (DTFJ). (B,C) Computed tomography of DTFJ shows highly eroded bone.

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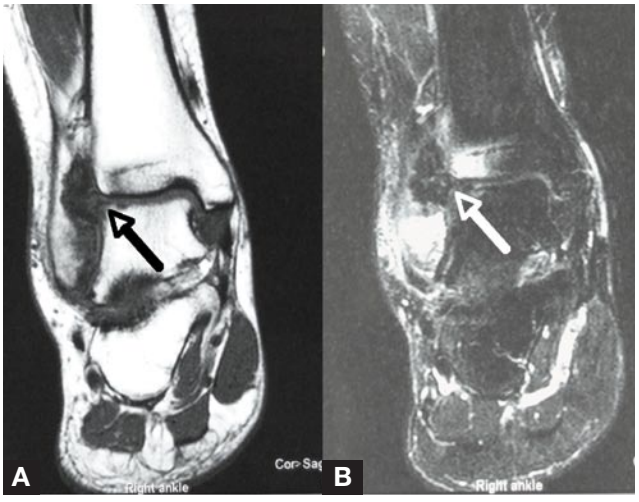
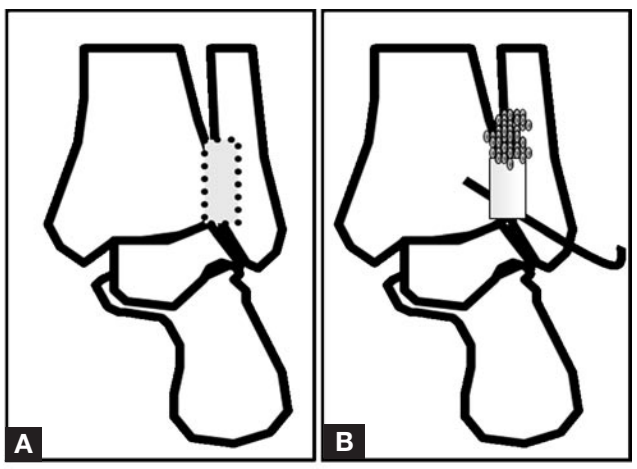


Figure 2. T_1-weighted (A) and T_2-weighted (B) magnetic resonance imaging of right ankle shows large multinodular mass in posterolateral aspect of ankle.



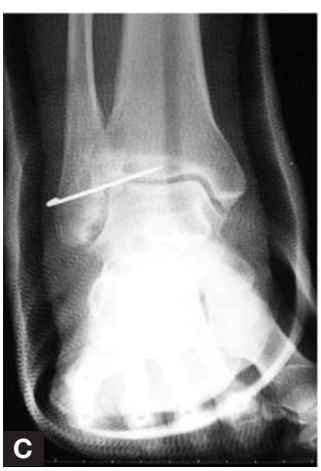
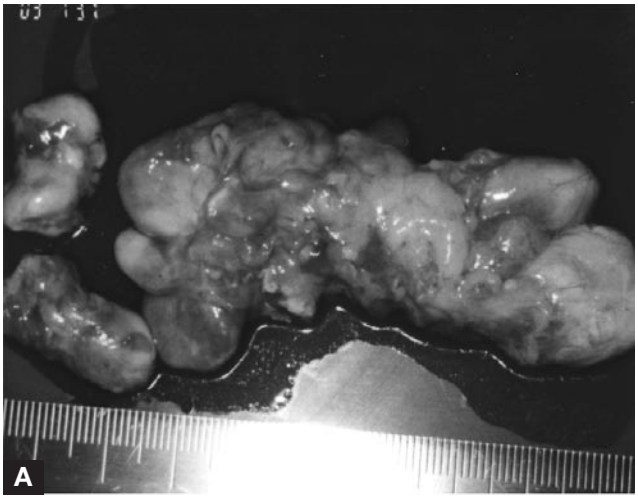


Figure 3. (A) Posterior view of distal tibiofibular joint (DTFJ) of right ankle. For complete visualization of DTFJ, rectangular cortical window was created in posterior aspect of DTFJ. (B) After thorough resection of lesions, arthrodesis of DTFJ was performed. Corticocancellous autograft was transplanted in denuded DTFJ. Kirschner wire was used to secure graft. (C) Anteroposterior radiograph of ankle after surgery.

T_1- and T_2-weighted images in these masses (Figure 2). Arthroscopic biopsy was performed to rule out malignancy, as the lesion was diffuse and large with distinct bony invasion. Microscopic examination of the biopsy specimen revealed mononuclear histiocytes, multinuclear giant cells, foaming histiocytes, and hemosiderin deposits—consistent with a diagnosis of PVNS.



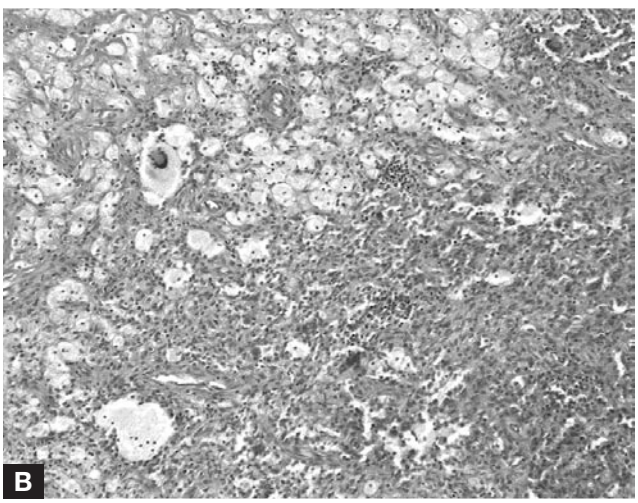


Figure 4. (A) Postoperative photograph shows large brown and yellow mass. (B) Microscopic photograph of histologic specimen of pigmented villonodular synovitis shows mononuclear histiocytes, multinuclear giant cells, foaming histiocytes, and hemosiderin deposits (hematoxylin and eosin stain, original magnification ×100).

Thorough resection of the masses and synovitis in the ankle was carried out with 2 separate longitudinal incisions. In the posterolateral incision, a rectangular cortical window was created in the posterior aspect of the DTFJ after resection of the extra-articular masses. The lesions in the DTFJ, lateral tibiotalar joint, and talofibular joint were completely removed from this window. In contrast, the lesions in the medial tibiotalar joint as well as the extra-articular masses were removed through the posteromedial incision. After resection, a corticocancellous autologous bone graft harvested from the iliac bone was shaped and snugly impacted in the DTFJ and fixed with a Kirschner wire (Figure 3).

Findings from the histologic examination of the excised mass were identical to those for the biopsy specimen (Figure 4). A short leg cast was applied for 6 weeks after surgery, and then the patient started a gradual regimen of ankle range-of-motion exercises and weight-bearing. Five years after surgery, a follow-up evaluation revealed no significant pain, no limitation of ankle motion, and complete

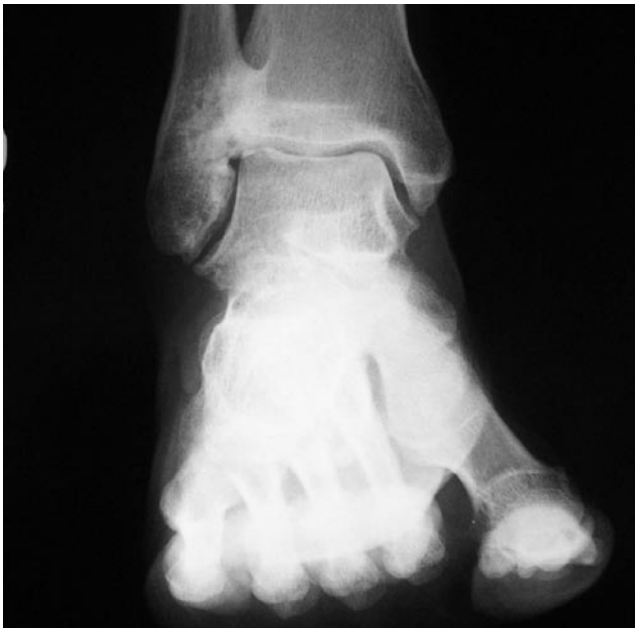


Figure 5. Anteroposterior radiograph of ankle at final follow-up shows completely united distal tibiofibular joint.

osseous fusion of the DTFJ (Figure 5). No local recurrence was noted on the follow-up MRI (data not shown).

Discussion

PVNS is a relatively rare disease affecting the synovial-lined joints. It was first fully described in 1941 by Jaffe and colleagues,[1] who considered it a benign inflammatory state of the synovium of unclear etiology. PVNS in the foot and ankle is even rarer but is also characterized by extensive lesion spread and frequent bony destruction.[8,9] Although Pandey and Pandey[8] stated that controversy still exists about the bony involvement of PVNS, the pressure from the lesion in the narrow or less expansile joint is considered the dominant etiology in joint destruction.[6,10]

The challenges in diagnosing PVNS were solved by MRI. For PVNS lesions, MRI findings are relatively unique—lobulated or multinodular masses extending from the joint with low signal intensity on both T_1- and T_2-weighted images, indicating hemosiderin deposits.[10,11] Other diseases that can have similar MRI findings—including rheumatoid arthritis, osteoarthritis with chronic hemorrhagic effusion, and hemophilia—are to be differentiated from PVNS.[10] Moreover, in the cases accompanied by extensive bony destruction, a differential diagnosis that excludes malignant tumor is essential. Thus, we performed an arthroscopic biopsy, which allowed for accurate diagnosis of PVNS before surgery; however, a needle or an open biopsy may also be useful.[12]

The optimal treatment for PVNS is surgery.[13] In the majority of cases, recovery is complete with thorough excision. However, particularly when PVNS is diffuse and shows extensive spread both inside and outside the joint, complete mass resection and total synovectomy are necessary. Total resection of the diseased tissue is extremely difficult because the ankle joint is not easily accessible for exposure of the entire joint without some invasion of healthy tissue. Therefore, when a curative operation leads to structural morbidity of the ankle joint, a bone graft to fill in the defect or arthrodesis is mandatory.[14]

Our patient's lesions were confined to the posterior aspect of the ankle joint; most emerged from the posterolateral quadrant. At first, we thought that 2 separate posterior approaches, posterolateral and posteromedial, would be optimum for excision of the lesions. As the DTFJ was highly eroded, however, we predicted that thorough curettage of it might lead to painful instability of the joint or to a pathologic fibular fracture necessitating an additional DTFJ-stabilizing procedure after excision. We concluded that sacrificing the DTFJ would be acceptable in achieving easier curettage and in obtaining better visualization of the posterolateral quadrant of the ankle joint. Given that our patient had a successful result, this limited arthrodesis technique may be useful in PVNS originating mainly in the DTFJ.

We successfully treated a rare case of PVNS of the ankle with severe bony destruction. Arthroscopic biopsy was useful in making an accurate diagnosis before surgical intervention. Arthrodesis of the DTFJ after thorough synovectomy should be considered an option in patients with ankle PVNS mainly originating in the DTFJ.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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