

# Pigmented Villonodular Synovitis

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**P**igmented villonodular synovitis (PVNS) is an uncommon, predominately benign neoplastic disorder affecting joints, bursae, and tendon sheaths. PVNS is characterized, grossly, by friable villous, nodular, or villonodular synovial proliferations. These proliferations project into the joint and may bleed, resulting in hemosiderin pigmentation and bloody or xanthochromic synovial fluid. Histologically, in addition to intracellular and extracellular hemosiderin deposition, there is a lymphocytic infiltrate with lipid-laden macrophages, isomorphic proliferating mononuclear synovial cells, and osteoclast-like giant cells.<sup>1</sup> The disorder is most common in the third to fifth decades of life. Men and women are equally affected.

Clinically, PVNS can be classified by the area of involvement. When a synovial joint is affected, either in a focal or diffuse manner, the disease is known as PVNS. Extra-articular manifestations of the disease occur when a bursa (pigmented villonodular bursitis [PVNB]) or a tendon sheath (pigmented villonodular tenosynovitis [PVNT]) is involved. Disease affecting the tendon sheath is also known as giant cell tumor of the tendon sheath (GCTTS). For the extra-articular and intra-articular types of the disease, incidence is estimated at 9.2 and 1.8 cases, respectively, per 1 million people.<sup>2</sup>

The extra-articular form of the disease most commonly presents as a solitary mass (50%-70%), often with associated joint pain and mechanical locking symptoms. Radiographs are normal in 20% of cases; however, extrinsic bone erosions (9%-25%), periosteal reaction (8%), and calcification (6%) may be present.<sup>2</sup> The ultrasound and magnetic resonance imaging (MRI) show a well-circumscribed mass intimately associated with a tendon sheath or bursa. MRI demonstrates a mass of low-to-intermediate signal intensity on both T<sub>1</sub>- and T<sub>2</sub>-weighted images.

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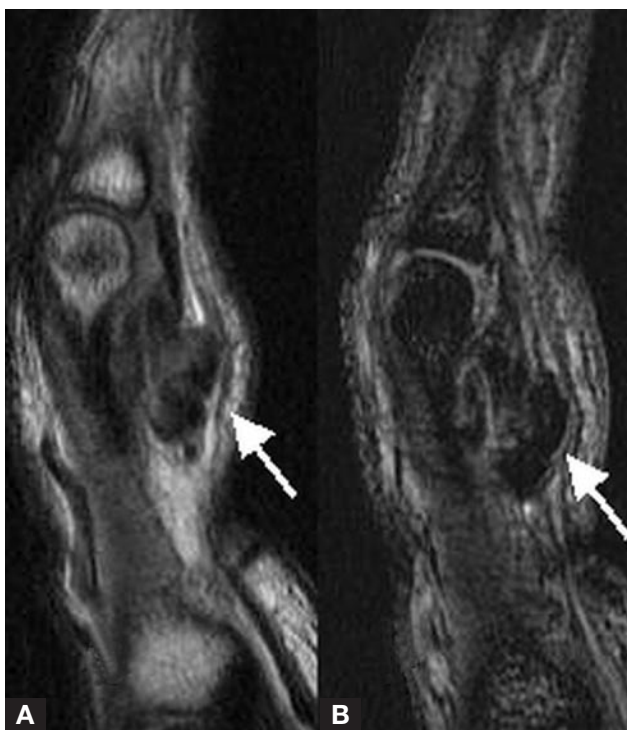
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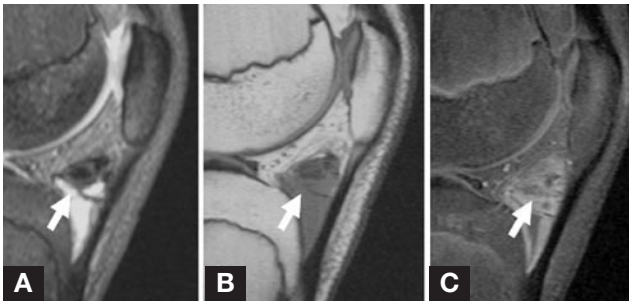
GCTTS is the second most common cause of soft-tissue mass in the hand and wrist after ganglion cysts (Figure 1). Up to 89% of GCTTS occurs in the hand and wrist, with the next most common site being the foot and ankle. The most common site of PVNB is within bursae around the knee and hip. Surgical resection is the preferred method of treatment for all localized forms of the disease. Recurrence rates of PVNB, PVNT, and GCTTS are low and return of function is generally favorable.<sup>3</sup>

Intra-articular forms of the disease characteristically involve a single joint. The knee is affected in up to 80% of cases followed in order by the hip, shoulder, elbow, and ankle.<sup>4</sup> Localized intra-articular disease occurs almost exclusively in the knee. In those cases, plain films are usually normal; however, a soft-tissue density mass may be seen, most commonly associated with Hoffa's fat pad (Figure 2). Localized intra- or extra-articular forms of the disease treated by simple excision have shown low recurrence rates.

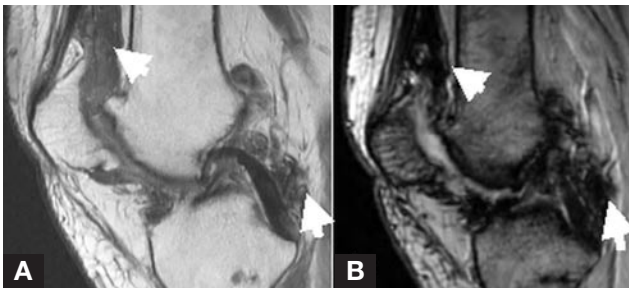
Diffuse intra-articular PVNS typically presents with slowly progressive pain, swelling, and decreased range of motion at the involved joint. Effusion is common and the joint may feel warm to the touch. Aspirated joint fluid typ-



**Figure 1.** Sagittal proton density (A) and sagittal gradient echo (B) images demonstrate a volar soft-tissue mass (arrows). Note blooming of the low signal intensity mass within the flexor tendon sheath on the gradient echo image reflecting the paramagnetic effect from hemosiderin degradation products.



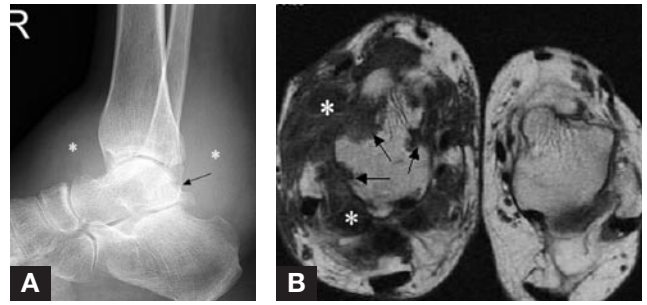
**Figure 2.** Sagittal fat-saturated proton density (A), T<sub>1</sub>-weighted (B), and fat-saturated T<sub>1</sub>-weighted postcontrast (C) images of knee demonstrate a complex focal enhancing predominately low-signal mass (arrows) within Hoffa's fat pad.



**Figure 3.** Proton density (A) and gradient echo sequences (B) of knee demonstrate diffuse intra-articular synovial proliferation with areas of nodularity, which are more pronounced on gradient echo sequences (arrows) as a result of blooming secondary to hemosiderin deposit.

ically is hemorrhagic or xanthochromic. Radiographs are normal in up to 21% of cases; however, common findings include dense effusion, nodular synovial-based soft-tissue masses, and bone erosions over both sides of the joint. There is usually normal bone mineralization and preservation of joint space.<sup>2</sup> Bone erosions are uncommon in the knee but occur more frequently in the shoulder, ankle, and hip where a constrained joint capsule is unable to expand in response to the enlarging synovial masses. Bone erosions are more readily identified with computed tomography and MRI than they are with x-ray (Figure 3).

The preferred imaging modality for assessing PVNS is MRI. Synovial-based masses demonstrate low signal intensity on all pulse sequences, a finding amplified at higher field strengths. The disease is characterized by the presence of increased prominence, or “blooming,” of PVNS lesions on gradient echo sequences, a paramagnetic effect (magnetic susceptibility) due to the presence of the blood breakdown product hemosiderin (Figure 4).<sup>2,5</sup> However, as in all forms of the disease, typical MRI findings are not always present, and on T<sub>2</sub>-weighted images, lesions may exhibit heterogeneous signal depending upon the amount



**Figure 4.** (A) Lateral radiograph of the right ankle shows a large, dense ankle effusion (asterisks) with suggestion of articular erosions (arrow). (B) Axial proton density images of both ankles in the same patient show prominent diffuse intra-articular soft-tissue masses (asterisks) with multiple erosions (arrows) in the right ankle. Normal left ankle is shown for comparison.

of hemosiderin deposit.<sup>6</sup> When present, low signal on T<sub>1</sub>- and T<sub>2</sub>-weighted sequences is helpful in making the correct diagnosis. MRI provides detailed information regarding intra-articular extent of disease. Postcontrast studies show prominent enhancement of the proliferative synovial masses, but, generally, contrast provides little additional diagnostic information.

The treatment of diffuse intra-articular disease is complete synovectomy, but recurrence rates have been reported as high as 50%. Surgery has been combined with adjuvant radiation therapy and postoperative external beam radiation therapy. Intra-articular administration of beta emitters (eg, yttrium-90) has been utilized in an attempt to combat recurrence of PVNS, with varying success.<sup>2,7</sup> Total joint arthroplasty or arthrodesis is recommended in patients with joint destruction and secondary arthritis.<sup>3</sup>

### AUTHORS' DISCLOSURE STATEMENT

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

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