MRI features of pigmented villonodular synovitis: A pictorial essay

Poster No.: C-2252
Congress: ECR 2010
Type: Educational Exhibit
Topic: Musculoskeletal
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Keywords: Pigmented Villonodular Synovitis, Magnetic resonance imaging, Hemosiderin
DOI: 10.1594/ecr2010/C-2252

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Learning objectives

To illustrate the MRI features of pigmented villonodular synovitis (PVNS), with special focus on the diffuse intraarticular form.

Background

Pigmented villonodular Synovitis (PVNS) belongs to a family of uncommon benign neoplastic diseases of uncertain etiology that involve the synovium of the joint diffusely or focally (PVNS), or it can occur in the extraarticular synovium of a bursa (pigmented villonodular bursitis - PVNB) or a tendon sheath (pigmented villonodular tenosynovitis - PVNTS). The current World Health Organization nomenclature describes these lesions as giant cell tumor of the tendon sheath (GCTTS) for PVNB or PVNTS and diffuse-type giant cell tumor for the diffuse intraarticular form of PVNS [1].

It typically affects adults 20-50 years of age (mean age, late 30s), but it might occur at any age group [1,3].

Clinical symptoms vary greatly and depend on lesion location. Extraarticular localized PVNB or PVNTS most frequently manifests with soft-tissue mass (83%-99% of cases) and pain (22%-71%). The intraarticular type of PVNS presents more commonly with pain (79%-90%) and swelling (72%-79%). In most cases the onset of the disease is chronic with duration of symptoms for months to years [1].

Lesions are frequently monoarticular and solitary, with polyarticular or multifocal involvement being very unusual.

The lesion location vary with the lesion subtype, either localized or diffuse, and on wether the lesion is intraarticular or extraarticular.
• PVNTS most commonly involves the hand or wrist, representing the second most common soft-tissue mass in this location (exceeded in frequency only by ganglia).
• PVNB most frequently involves the hip or knee.
• The localized intraarticular form of PVNS almost exclusively occurs in the knee.
• The diffuse intraarticular form of PVNS most frequently affects the large joints with a predilection for the lower extremities, particularly the knee and hip. In decreasing order of frequency it can also involve the ankle, shoulder and elbow [1, 4]. However, it should be noted that any articular site may be involved.

The pathologic specimens show a hypertrophyc synovium typically villous, nodular, or villonodular which contains variable amounts of hemosiderin. Hemosiderin deposition occurs in the majority of cases, but it is most prominent in the diffuse intraarticular form of the disease [1].

The treatment of choice for PVNS is surgical resection with total synovectomy. Recurrence is more frequent with diffuse intraarticular PVNS, and adjuvant radiation therapy may be employed for treatment in this cases [1].

**Imaging findings OR Procedure details**

Imaging plays an important role in the diagnosis of PVNS. MRI is superior to conventional radiography for depicting abnormalities of the joint, and is able to determine the true extent of the disease.

• MRI is especially important evaluating diffuse intraarticular PVNS, as it presents on radiography as a monoarticular arthropathy with nonspecific clinical symptoms [1].

The characteristic MRI findings include a heterogeneous, diffuse, synovial-based thickening, often associated with nodularity, that affects most or the entire joint.
The synovial thickening presents with:

1. a intermediate-to-low signal on T1-weighed images (Fig.1 on page 5, Fig.3 on page 7, Fig.5 on page 9, Fig.6 on page 10, Fig.7 on page 11, Fig.8 on page 12, Fig.9 on page 13);
2. low signal intensity on T2-weighed images (Fig.7 on page 11, Fig.9 on page 13);
3. there is a characteristic enlargement of the low signal areas ("blooming") at T2* images (gradient-echo), due to the magnetic susceptibility effects from the hemosiderin deposits; these deposits vary markedly in severity, from tiny low signal particles to large areas of low signal (Fig.1 on page 5, Fig.2 on page 6, Fig.6 on page 10);
4. intermediate-to-low signal intensity on proton density-weighted images (Fig.2 on page 6, Fig.3 on page 7, Fig.4 on page 8, Fig.10 on page 13);
5. intermediate-to-high signal intensity on short inversion time inversion recovery images (STIR) (Fig.5 on page 9, Fig.8 on page 12);
6. enhancement after intravenous administration of gadolinium is also present, in relation to the rich vascularisation of the diseased synovial tissue, but is not required for the diagnosis.

Additional findings include bone erosion (Fig.1 on page 5) caused by pressure atrophy, invasion of bone, or both (most commonly seen in the hip, ankle and elbow, owing to the tight joint spaces) bone marrow edema (Fig.8 on page 12) and joint effusion (Fig.4 on page 8).

- Localized intraarticular PVNS, which almost exclusively involves the knee, presents as soft-tissue masses with well defined nodular or lobular margins. The signal intensity on T1 and T2 - weighted images is similar to that of the diffuse form. The hemosiderin deposition is much less extensive than that seen in diffuse intraarticular disease, and it usually presents as focal circular areas of low signal intensity on T2-weighted or gradient-echo images.

- Localized extraarticular disease (PVNB and PVNT) is less commonly evaluated with MRI because the diagnosis is often suggested clinically. MRI typically demonstrates a well circumscribed, soft tissue mass adjacent to the affected tendon (PVNT) or involving the bursa (PVNB). The bursal form of extraarticular disease is more similar to diffuse intraarticular PVNS, as it demonstrates prominent MR imaging effects of hemosiderin.

Differential diagnosis
Although hemosiderin deposition in the soft tissue with its "blooming effect" is highly characteristic of PVNS, it’s not exclusive of this disease and we have to consider other differential diagnosis.

Synovial hemangioma, haemophiliac arthropathy and hematomas may demonstrate similar MR imaging findings to PVNS, but the combination of synovial proliferation, villonodular soft tissue masses, hemosiderin deposits, and subchondral bone erosion is highly diagnostic for PVNS. Also, PVNS is not associated with either serpentine vascular channels (hemangioma) or a clinical history of haemophilia [1, 4].

Within the differential diagnosis for PVNS is also synovial osteochondromatosis, which usually presents with multiple loose bodies within the joint (there is "blooming" of the cortical portions of the loose bodies caused by susceptibility artifact on gradient-echo images) but occasionally appears like a confluent mass of tissue with high signal intensity on T2-weighted images.

Images for this section:
Fig. 1: Figure 1. Intraarticular PVNS of the elbow in a 48-year-old woman. Sagittal (a) and axial (b) T1-weighted images reveal the diffuse intraarticular low signal intensity mass around the right elbow, with bone erosions (small arrow in a). (c) Sagittal short inversion time inversion recovery (STIR) shows the intra osseous extension of the lesion (white arrow). (d) Coronal gradient-echo image shows very low signal intensity of hemosiderin ("blooming artifact").
Fig. 2: Figure 2. Intraarticular PVNS of the knee in a 42-year-old woman. (a, b, c) Axial (a), coronal (b) and sagittal (c) proton-density-weighted fat-suppressed images, reveal a large synovial based mass with intermediate signal intensity, involving the anterior knee but extending into the posterior compartment. The sagittal gradient-echo image (d) shows focal hypointense areas (white arrow), representing the "blooming artifact" of hemosiderin.
Fig. 3: Figure 3. PVNS of the knee in a 54-year-old woman. Sagittal T1-weighted (a, b) and sagittal (c) and coronal (d) proton-density-weighted fat supressed images show synovial proliferation with hypointense masses around the knee.
**Fig. 4:** Figure 4. Same patient as in figure 3. Sagittal (a and b), coronal (c) and axial (d) proton-density-weighted fat suppressed images demonstrating joint effusion (arrows in b and d), large erosions along the posteromedial aspect of the medial femoral condyle (arrows in a) and full thickness medial compartment articular cartilage loss with adjacent subarticular stress change (arrow in c).
Fig. 5: Figure 5. Intraarticular PVNS of the ankle in a 39-year-old woman. Sagittal STIR (a, b) and T1-weighted images (c, d) show involvement of the anterior and posterior recesses of the ankle. The mass has intermediate signal intensity on STIR (arrows in a and b) and on T1-weighted images (arrows in c and d).
**Fig. 6:** Figure 6. Intraarticular PVNS of the ankle in a 34-year-old man. (a, b) Coronal (a) and sagittal (b) T1-weighted images show low signal intensity mass involving the posterior recess of the ankle. This mass has intermediate signal intensity in proton-density-weighted images (c) and there are focal hypointense areas in gradient-echo images in keeping with hemosiderin deposits (arrow in d).
Fig. 7: Figure 7. Same patient as in figure 6. (a, b) Axial T1-weighted (a) and T2-weighted images (b) show the posterior involvement of the ankle joint. The mass presents with intermediate signal intensity in T1-weighted images (a) and low signal intensity in T2-weighted (b) images.
**Fig. 8:** Figure 8. PVNS of the ankle in a 39-year-old man. (a, b) Sagittal T1-weighted (a) and STIR images (b) show prominent nodular thickening of the anterior joint line synovium of the ankle. This synovial thickening has intermediate signal intensity with T1-weighting (a) and with STIR images (b). There are advanced degenerative changes within the anterior ankle joint adjacent to the synovial disease, with associated bone marrow edema (white arrow in b).

![Figure 8](image)

**Fig. 9:** Figure 9. Same patient as in figure 8. Axial T1-weighted (a) and T2-weighted images (b) reveal the anterior synovial thickening with intermediate signal intensity in T1-weighted images (a) and low signal intensity in T2-weighted images (b).

![Figure 9](image)
Fig. 10: Figure 10. Same patient as in figures 8 and 9. (a, b) Coronal oblique proton-density-weighted images show advanced degenerative changes within the anterior ankle joint, with small lateral and anterior talar erosions (arrows).
Conclusion

MR imaging is the preferred modality for diagnosing PVNS. The MRI appearance of PVNS is not pathognomonic but is strongly suggestive of the diagnosis when the typical features are present. MRI is also useful for preoperative planning and in post-operative monitoring for recurrence.

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References


