Radiological features of pigmented villonodular synovitis and giant cell tumor of the tendon sheath

P. Schvartzman\textsuperscript{a,b,}\textsuperscript{*}, V. Carroz\textsuperscript{a}, T. Pascual\textsuperscript{b}, L. Mazza\textsuperscript{b}, M. Odesser\textsuperscript{b} and J.L. San Rom\textsuperscript{\textdagang{a}}

\textsuperscript{a} Centro Médico Deragopyan, CABA, Buenos Aires, Argentina
\textsuperscript{b} TCba Fundación Jaime Roca, CABA, Buenos Aires, Argentina

Abstract

\textbf{Purpose:} To show the resonance magnetic imaging (MRI) findings of pigmented villonodular synovitis (PVNS) and giant cell tumor of the tendon sheath (PVNTS), entities with similar histology but differences in clinical and some radiological manifestations.

\textbf{Materials and methods:} We studied 25 cases with histologically benign synovial proliferation in intra and extra-articular location of the extremities. We highlighted the different types of imaging manifestations with a 1.5T MRI unit. The results were analyzed and compared with the literature.

\textbf{Results:} MRI displayed very specific imaging features in all patients. However, we were able to distinguish 4 main patterns of presentation depending on the morphology, location of the lesion and differential radiological characteristics. These were: as dominant presentation, giant cell tumor of the tendon sheath (n = 10), all of them extra-articular in location; bursal form of pigmented villonodular synovitis (n = 2); focal pigmented villonodular synovitis (n = 5); and diffuse pigmented villonodular synovitis (n = 8).

\textbf{Conclusion:} Both pigmented villonodular synovitis as well as giant cell tumor of the tendon sheath are considered similar from the point of view of histological findings. MRI was useful to objectify both the similar and differential radiological features of these entities, which along with the location, enabled us to distinguish 4 patterns of presentation. Recognition of these patterns allows for an adequate follow-up of disease and an optimal therapeutic management.

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\textbf{Keywords:} Pigmented villonodular synovitis; Giant cell tumor of the tendon sheath; Synovial proliferation

Introduction

Pigmented vellonodular synovitis (PVS) and the giant cell tumor of the tendon sheath (GCTTS) --though usually considered as independent entities-- represent a diverse group of proliferative lesions of the synovium\textsuperscript{1}. Although an inflammatory origin has been suggested, their etiology remains unknown. Recent studies have highlighted a potential autonomous growth, and they are therefore considered as benign neoplastic processes\textsuperscript{1-3}. However, rare cases of malignant PVS have been also reported\textsuperscript{1,2}.

PVS may occur as a localized intraarticular focal form (FPVS), a diffuse intraarticular form (DPVS) or an extraarticular form involving the bursa (BPVS); the GCTTS is considered as a localized extraarticular form, being --according to the literature—the most common entity\textsuperscript{1,4,5}. These 4 patterns have similar histological findings and certain radiological patterns in common. However, some imaging characteristics (as well as other differences in clinical symptoms, treatment and the course of disease) help us to differentiate them.

The aim of this study is to evaluate the imaging characteristics and differential aspects of this group of lesions.

Materials and methods

Between May 2011 and June 2013, we retrospectively studied 25 patients (16 men and 9 women) with an age range between 37 and 86 years (mean: 45.5 years) at our institution. Cases with histologically confirmed diagnosis of PVS were selected. All patients were immunocompetent.

The evaluation was performed using a 1.5 Tesla MRI scanner (General Electric Gyroscan Intera, Best, The Netherlands) and another 1.5 Tesla for small joints (General Electric Optima, Medical System, Wilmington, MA, USA). Following the standard protocol at our institution, T1- and T2-weighted images, fat-suppressed images and those specific to the disease were obtained: gradient echo and contrast-enhanced imaging. In
all cases, the contrast administered was intravenous gadolinium Optimark® 0.5 mmol/ml (Mallinckrodt Inc.) at a dose of 1 ml/10 kg body weight. Kidney disorders had been previously ruled out. Catheters were inserted before the patients went into the MRI scanner and none of the patients experienced subsequent study-related symptoms. After the MRI exam, post-processing was performed with measurement of lesions. Results were analyzed and compared with the literature published to date.

![Figure 1](image)

**Figure 1** Giant cell tumor of the tendon sheath. (a) Sagittal proton-density-weighted fat-suppressed image with microcoil shows a lesion with well-defined margins, adjacent to the flexor tendon in the index finger. Predominantly low and heterogeneous signal, with strong enhancement after intravenous contrast administration. (b) On sagittal T1-weighted image, the lesion shows a hypointense signal on T1-weighted image, the lesion shows a hypointense signal, in intimate relationship with the flexor tendon.

<table>
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<tr>
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<th>GCTTS</th>
<th>BPVS</th>
<th>FPVS</th>
<th>DPVS</th>
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<tr>
<td>Percentage of cases</td>
<td>40%</td>
<td>8%</td>
<td>20%</td>
<td>32%</td>
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<tr>
<td>Predominant location</td>
<td>Hand (volar aspect of the 2nd and 3rd finger)</td>
<td>Foot (adjacent to the 3rd metatarsus)</td>
<td>Knee</td>
<td>Knee</td>
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<tr>
<td>Clinical symptoms</td>
<td>Palpable mass and localized pain</td>
<td>Palpable mass and localized pain</td>
<td>Numbing and localized pain</td>
<td>Intense pain, numbing and, in some cases, joint dysfunction</td>
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GCTTS: giant cell tumor of the tendon sheath; BPVS: pigmented villonodular synovitis of the bursa; FPVS: focal pigmented villonodular synovitis; DPVS: diffuse pigmented villonodular synovitis.
Results

Patients experienced variable symptoms, depending on lesion location (table 1). Extraarticular lesions (BPVS or GCTTS), clinically manifested with a soft tissue lesion and pain; whereas when there was joint involvement (FPVS or diffuse PVS), intense pain and reddening was noted and, in 2 cases, joint dysfunction.

Four types of clinical presentations were detected:

Giant cell tumor of the tendon sheath

GCTTS represents the localized extraarticular form of PVS. In agreement with the literature, in our study this entity was the dominant presentation, with 10 cases (40%). In all patients, the lesion, of approximately 1.3 cm, was located in the hand, predominantly involving the index or long fingers adjacent to flexor tendons. Three of the cases had previous ultrasounds scans that reported this pathology as suspected diagnosis and revealed a well-defined hypoechoic solid mass, intimately related to the involved tendon. Doppler imaging showed blood flow inside.

Figure 2 Giant cell tumor of the tendon sheath. (a) Coronal proton-density-weighted fat-suppressed image shows a lobulated mass completely enveloping the flexor tendon. Predominantly low and heterogeneous signal with mild underlying soft tissue swelling. (b) Coronal T1-weighted image shows two hypointense lesions of similar size, enveloping the flexor tendon of the 5th finger in the middle and distal phalanx anatomy (rare presentation).

Figure 3 (a and b) Bursal pigmented villonodular synovitis in the foot. Coronal fat-suppressed T1-weighted images show a small hypointense nodular mass at the level of the 3rd intermetatarsal space, intimately related to the bursa, extending towards the plantar region.
On MRI, GTTSs had an encapsulated appearance, were hypointense on T1-weighted images and heterogeneous, predominantly hypointense on T2-weighted images, with strong enhancement after intravenous contrast (fig. 1). In some cases the lesion was noted to completely envelop the tendon sheath in a diffuse manner (fig. 2a). We also detected a very infrequent case of two lesions in one finger (the 5th), with extension to the flexor tendon. The primary lesion was located at the level of the middle phalanx and the satellite nodule in the distal phalanx (fig. 2b).

**Bursal pigmented vellonodular synovitis**

Bursal pigmented vellonodular synovitis was observed in 2 cases (8%). Both patients had a palpable mass in the foot (one on the plantar aspect and the other on the dorsal aspect) adjacent to the 3rd intermetatarsal space. Lesions were approximately 3 cm in diameter and had defined margins, being iso- or slightly hypointense to muscle. After contrast administration, lesions showed homogeneous enhancement (fig. 3).

**Focal pigmented vellonodular synovitis**

This pattern was found in 5 patients (20%). Four of them had knee involvement at the level of the infrapatellar fat pad and one had subcoracoid involvement in the shoulder. In all cases, lesions were well-defined and had an approximate diameter of 2.5-3 cm. MRI showed a well-defined or lobulated-margin nodular mass, which appeared hypointense on T1-weighted images, while T2-weighted images revealed a variable and heterogeneous, predominantly low, signal intensity with respect to adjacent joint effusion (present in 40% of cases). Gradient-echo sequences revealed areas of lower signal intensity in the periphery, corresponding to hemosiderin deposition (blooming effect) (figs. 4 and 5).

**Diffuse pigmented vellonodular synovitis**

Eight cases (32%) were detected: 6 occurring in the knee, one in the hip and the other in the ankle. Most patients were young (20-40 years old). Multiple areas of diffuse and heterogeneous synovial thickening were seen. In 3 of the cases this thickening was associated with bone erosions and sclerotic margins, findings that are related to more aggressive involvement of the lesion. Synovial thickening areas showed a hypointense to intermediate nodular pattern, both on T1- and proton-density-weighted images.

It is important to highlight that in this disease, the hypointense pattern on T2-weighted images is caused by the mag-

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![Figure 4](image_url)

**Figure 4** (a) Focal pigmented vellonodular synovitis in the knee. Coronal gradient-echo image shows a small hypointense mass with areas of lower signal intensity in the periphery, corresponding to hemosiderin deposition (blooming effect). (b) Proton-density-weighted image shows a lobulated, iso- to hypointense and heterogeneous mass posteriorly to the posterior cruciate ligament, associated with mild joint effusion.
netic susceptibility artifact from hemosiderin. In our study, this finding was particularly visible on gradient-echo sequences. It is commonly detected in the periphery of the lesion (blooming effect) (fig. 6).

Occasionally, areas of high-signal intensity were noted within the lesion due to the presence of fluid, edema or fat. After intravenous administration of gadolinium, there is strong enhancement, which may be homogeneous or septated (fig. 7).

Discussion

Imaging characterization of PVS and GCTTS varies depending on the subtype of disease, localized or diffuse, and on the joint involved.

According to Hughes et al.⁴, plain radiograph (X-ray), in cases of GCTTS or BPVS, shows a soft tissue mass in 60% of cases, whereas in 18% of cases there may be bone erosions, generally with well-defined margins, and very rarely a periosteal reaction (8%) and calcifications (6%). BPVS appears normal on the x-ray and only in some cases soft-tissue opacity that replaces the infrapatellar Hoffa fat pad may be seen. Bone erosion is extremely rare. Radiographs of DPVS usually demonstrate joint effusion, soft-tissue swelling, and subchondral lesions with absence of calcification. Overall, erosive changes are seen in 50% of cases, although this depends on the site of involvement, being more frequently seen when they occur in the ankle, hip and elbow (i.e., all the joints with less space and higher intracapsular pressure)⁵. Anyway, the X-ray may appear normal in 25% of patients. On CT, in cases of DPVS, shows synovial thickening, with slightly decreased attenuation relative to that of the muscle (a finding associated with hemosiderin deposition). This method is optimal to demonstrate the presence of bone erosions and subchondral cysts ⁶.

GCTTS represents the second most common cause of soft tissue mass in the wrist, with ganglion being the most common ⁴,⁷. The volar aspect is affected approximately twice as often as the dorsal aspect. In addition, differential diagnosis may include fibrous tumor, of similar features but far less frequent. In our study, we found lesions involving only the 3rd, 4th or 5th finger. Although this is the most common presentation of this disease (85%)⁸, it may also be located at the level of the ankle, the foot and, very rarely, in the knee, hip, elbow and shoulder ⁶.

Diagnosis may be suggested by clinical symptoms or made by ultrasound. Anyway, MRI shows a well-defined mass, adjacent to the tendon involved. Lesions are usually small and encapsulated, with signal intensity similar to or lower than that of muscle on T1-weighted images and predominantly low and heterogeneous on T2-weighted images. After intravenous contrast, strong and homogeneous enhancement is observed.

Our findings were similar to those reported in the literature. The case with multifocal involvement, where there was a pri-
mary lesion and a satellite nodule, is uncommon\textsuperscript{1, 5}. Surgery is usually curative, although in 7-27% of patients there may be recurrence\textsuperscript{4}.

BPVS, unlike our two cases of foot location, more frequently occurs in the hip or knee, as a soft tissue mass with hypointense to intermediate signal on T1- and T2-weighted images\textsuperscript{7}. As it occurs in the diffuse and localized form, the low signal intensity may be due to the presence of hemosiderin. Small intrallesional foci of high signal intensity are usually seen that represent fluid entrapped within the (a common finding in the FBPS form). Occurrence in the foot, as reported in our study, is uncommon\textsuperscript{2, 7}.

Localized PVS, also referred to as focal or nodular synovitis, is usually an intraarticular lesion that almost exclusively involves the knee, being located in the infrapatellar fat pad in 70% of cases, in the suprapatellar pad in 20% and in the posterior intercondylar area in 10%\textsuperscript{1}. The form detected in the shoulder in our study, though unusual, represents the second most frequent. In the localized form, PVS manifests as a well-defined solitary mass, which appears hypointense on T1-weighted images and heterogeneous on T2-weighted and gradient-echo images. In addition, focal areas of low signal intensity are usually seen (75%), predominantly in the periphery of the lesion, corresponding to hemosiderin deposition, particularly visible on T2-weighted images and gradient-echo images. However, these areas are much less extensive than those seen in diffuse intraarticular disease. Moderate enhancement of FPVS is seen after contrast administration.

For DPVS, the typical location is the knee (80%), although in decreasing order of frequency, the hip, the ankle, shoulder, and elbow may be affected. DPVS usually occurs in young patients (in the 3rd to 4th decades of life), with equal frequency in both sexes, and unlike other entities, its presentation with pain and soft tissue mass is not unusual.

MRI is typically used after X-ray because of the nonspecific clinical symptoms of a monoarticular arthropathy. MRI shows a heterogeneous, diffuse synovial thickening that often is associated with nodularity. Joint effusion is common, particularly in large joints such as the knee and the ankle. The signal intensity is intermediate to low on T1-weighted images, while low signal intensity predominates on T2-weighted and gradient-echo images because of the magnetic susceptibility artifact from hemosiderin. This finding is nearly pathognomonic of the intraarticular form of the disease and it enables us to differentiate it from other conditions such as chondromatosis. The effect is produced by the local magnetic field created by iron present in hemoglobin, which causes proton dephasing and, consequently, a signal void\textsuperscript{9}. After intravenous contrast, a strong enhancement of homogeneous or peripheral appearance is seen.

When there is great involvement, it may extend outside the joint towards periarticular tissues, such as the proximal tibia.

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{fig6.png}
\caption{(a and b) Diffuse pigmented vellonodular synovitis in the knee. Sagittal proton-density and T2-weighted images show multiple areas of synovial thickening with diffuse pattern, with small hypointense foci of lower signal intensity inside, corresponding to hemosiderin deposition (blooming effect).}
\end{figure}
Other common findings of diffuse PVNS include erosions or subchondral cysts (62%), septations (67%), articular cartilaginous defects (31%), diffuse osteitis and edema in the soft tissue (23%)\(^{10}\).

The major conditions in the differential diagnosis of diffuse PVNS include synovial chondromatosis and hemophiliac arthropathy. The former is usually recognized by the presence of cartilaginous nodules of the synovial membrane that can calcify and be seen on X-ray or MRI. The site of detachment...
of the nodule should be identified at the imaging study. A single isolated nodule without endochondral ossification may be wrongly taken for localized PVS. Hemophilic arthropathy may show similar findings, namely bone erosions and hemosiderin deposition, but clinical history is essential for differential diagnosis.

The risk of malignancy is very low and malignant transformation can occur de novo or be associated with recurrent disease. The prevalence of malignant transformation is 3%, being more frequent for DPVS\textsuperscript{1-3}. These lesions are sarcomatous tumors with synovial origin and poor prognosis.

In the pathologic evaluation of PVS\textsuperscript{4,11}, gross examination shows villous synovial proliferations, which are typically reddish because of hemosiderin deposition within the lesion, while on microscopic examination the lesion consists of finger-like projections of hyperplastic synovium. In the early phase of disease, multinucleated giant cells, lymphocytes and a small amount of hemosiderin are seen, while in the late phase, hemosiderin deposition is much more evident and fibrosis predominates. In our experience, histologic samples showed in all cases an evident inflammatory process with hemosiderin deposition, which suggests that this finding is present in all forms of PVS evaluated (fig. 8).

Treatment of the various forms of PVS is required to prevent progressive loss of function and destruction of the joints (DPVS or FPVS) or the tendon or bursa (GCTTS or DPVS). Surgical resection is the therapeutic management of choice for all forms of PVS and its success depends on complete resection of the disease. The efficacy of the surgical approach depends on the joint involved, extent of disease, and experience of the surgeon\textsuperscript{1-3}, but logically, the more localized the lesion, the more likely the cure. In this respect, DPVS is more difficult to eradicate, and adjuvant radiation therapy may be often required.

**Conclusion**

PVS represents a diverse group of proliferative lesions of the synovium with hemosiderin depositions. Even if these entities are histologically similar, they have specific imaging characteristics on MRI, which, together with location of disease enable us to distinguish 4 patterns of presentation. Thus, when the site of origin is intraarticular, the focal or diffuse forms may be seen, and extraarticular disease is subdivided into bursal or tendon sheath lesions (GCTTS). MRI is the tool of choice for accurately defining the extent of disease and its relationship to the surrounding tissues, in any form of presentation. These features are important to establish an adequate follow-up and management of patients.

**Conflicts of interest**

The authors declare no conflicts of interest.

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**References**