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Pigmented Villonodular Synovitis: Radiologic-Pathologic Correlation[1]

CME FEATURE

See accompanying test at http://www.rsna.org/education/rg_cme.html

LEARNING OBJECTIVES FOR TEST 6

After reading this article and taking the test, the reader will be able to:

- Describe the radiologic manifestations of pigmented villonodular synovitis.
- Identify the pathologic basis of radiologic features of pigmented villonodular synovitis.
- Discuss the spectrum of disease processes grouped with pigmented villonodular synovitis, as well as the treatment options and prognosis.

TEACHING POINTS

See last page

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Pigmented villonodular synovitis (PVNS) represents an uncommon benign neoplastic process that may involve the synovium of the joint diffusely or focally (PVNS) or that may occur extraarticularly in a bursa (pigmented villonodular bursitis [PVNB]) or tendon sheath (pigmented villonodular tenosynovitis [PVNTS]). Pathologic specimens of the hypertrophic synovium may appear villous, nodular, or villonodular, and hemosiderin deposition, often prominent, is seen in most cases. The knee, followed by the hip, is the most common location for PVNS or PVNB, whereas PVNTS occurs most often in the hand and foot. PVNTS is also referred to as giant cell tumor of the tendon sheath (GCTTS). PVNTS is the most common form of this disease by a ratio of approximately 3:1. Radiographs reveal nonspecific features of a joint effusion in PVNS, a focal soft-tissue mass in PVNB or PVNTS, or a normal appearance. Extrinsic erosion of bone (on both sides of the joint) may also be seen and is most frequent with intraarticular involvement of the hip (>90% of cases). Cross-sectional imaging reveals diffuse involvement of the synovium (PVNS), an intimate relationship to the tendon (PVTNS), or a typical bursal location (PVNB), findings that suggest the diagnosis. However, the magnetic resonance (MR) imaging findings of prominent low signal intensity (seen with T2-weighting) and "blooming" artifact from the hemosiderin (seen with gradient-echo sequences) are nearly pathognomonic of this diagnosis. In addition, MR imaging is optimal for evaluating lesion extent. This information is crucial to guide treatment and to achieve complete surgical resection. Recurrence is more common with diffuse intraarticular disease and is difficult to distinguish, both pathologically and radiologically, from the rare complication of malignant PVNS. Recognizing the appearances of the various types of PVNS, which reflect their pathologic characteristics, improves radiologic assessment and is important for optimal patient management.

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Abbreviations: GCTTS = giant cell tumor of the tendon sheath, H-E = hematoxylin-eosin, PVNB = pigmented villonodular bursitis, PVNS = pigmented villonodular synovitis, PVNTS = pigmented villonodular tenosynovitis, STIR = short inversion time inversion recovery

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Introduction

Pigmented villonodular synovitis was first described by Chassaignac (1) in 1852 as a nodular lesion of the synovial membrane that affects the flexor tendons of the fingers. However, the current description of this entity was not applied until 1941 by Jaffe and colleagues (2), who used the designations pigmented villonodular synovitis (PVNS), pigmented villonodular bursitis (PVNB), and pigmented villonodular tenosynovitis (PVNTS). Previous terminology for PVNS included synovial xanthoma, synovial fibroendothelioma or endothelioma, benign fibrous histiocytoma, xanthomatous giant cell tumor of the tendon sheath, myeloplaxoma, chronic hemorrhagic villous synovitis, giant cell fibrohemangioma, fibrohemosideric sarcoma, sarcoma fusigiganocellulare, benign or malignant polymorphocellular tumor of the synovial membrane, and fibrous xanthoma of the synovial membrane (3–8). The current World Health Organization nomenclature describes these lesions as giant cell tumor of the tendon sheath (GCTTS) for PVNTS or PVNB and diffuse-type giant cell tumor for the diffuse intraarticular form of PVNS (2,9). Although the etiology of this disease process remains uncertain, current cytogenetic abnormalities and its potential for autonomous growth suggest that PVNS is a neoplastic process.

PVNS represents a benign, hypertrophic synovial process characterized by villous, nodular, and villonodular proliferation and pigmentation from hemosiderin. These components vary in prominence from lesion to lesion. Jaffe et al (2) proposed the terminology of PVNS, as well as PVNB and PVNTS, to denote involvement of not only intraarticular synovium but also its extraarticular extensions that affect bursa and tendons. Granowitz and co-workers (10) further subclassified these lesions as localized (PVNB or PVNTS) or diffuse (PVNS). The localized extraarticular form of PVNS is frequently referred to as GCTTS. Localized disease may also affect an intraarticular location; such a case is referred to as the localized intraarticular form of PVNS, and these cases occur almost exclusively in the knee (3–5,7). Malignant PVNS has also been reported in rare cases (3–5,7).

Imaging manifestations of PVNS, PVNB, and PVNTS are frequently characteristic of the disease process. Radiographic features are nonspecific, and radiographs may appear normal. The diffuse intraarticular form of PVNS often demonstrates a joint effusion and extrinsic erosion of bone on both sides of the joint, but the joint space is unaffected. The localized forms of disease usually reveal only a soft-tissue mass. Computed tomography (CT) depicts these lesions as either diffuse thickening of the tissue about the joint (PVNS) or as a localized soft-tissue mass (PVNB or PVNTS). The attenuation of the lesion may be somewhat increased relative to that of muscle owing to the presence of hemosiderin. Extrinsic erosion on both sides of the joint is well demonstrated by CT and is more frequent in lesions in less capacious joints such as the hip. The magnetic resonance (MR) imaging appearances of PVNS, PVNB, or PVNTS are often characteristic of these lesions, which demonstrate low to intermediate signal intensity with all pulse sequences. Use of gradient-echo pulse sequences allows confirmation of the presence of hemosiderin, which appears as a prominent "blooming" of low signal intensity due to magnetic susceptibility artifact. MR imaging is optimal for identifying the extent of synovial disease with diffuse intraarticular involvement (ie, PVNS), for demonstrating its intimate relationship to the tendon sheath in PVNTS, or for revealing its typical bursal location in PVNB. Detection of disease location and extent are important both for diagnosis and to guide treatment. In this article, the clinical features, pathologic characteristics, spectrum of radiologic appearances, complications, treatment, and prognosis of all types of PNVS are discussed and illustrated.

Clinical Characteristics

PVNS, either diffuse intraarticular, localized extraarticular, or localized intraarticular, is a relatively uncommon disease. The average annual

incidence has been estimated to be 9.2 and 1.8 cases per 1 million population for the extraarticular and intraarticular forms of the disease, respectively, in the study by Myers and Masi (11). Localized disease, whether extraarticular or intraarticular, represents 77% of cases (with the tenosynovial form being the most common), compared with diffuse intraarticular involvement, which accounts for 23% (a 3.3:1 ratio) (11). Localized intraarticular involvement represents 6% of all cases (11). The localized extraarticular form of the disease accounts for 1.6%–3.9% of all benign soft-tissue masses, whereas the diffuse intraarticular form represents 0.9% (4–8,12). Patients are most commonly in the 3rd to 5th decades of life (mean age, late 30s) at diagnosis (4–8,12). The diffuse intraarticular form of the disease occurs with equal frequency in both sexes, whereas the localized extraarticular form has a mild female predilection (1.5–2.1:1) (4–8,12).

Clinical symptoms vary greatly and depend on lesion location as intraarticular or extraarticular. Extraarticular localized PVNB or PVNTS most frequently manifests clinically with a soft-tissue mass (83%–99% of cases) and pain (22%–71%), whereas joint dysfunction or swelling are unusual (0%–4%) (4–8,11,12). Common clinical symptoms associated with the intraarticular type of PVNS are pain (79%–90% of cases) and swelling (72%–79%) (3–8,13), with joint dysfunction being a less frequent finding (26%–28%) and a soft-tissue mass being unusual (6%–19%) (5,11). The duration of symptoms also ranges widely, from 1 to 120 months, with a mean duration of 19 months for localized extraarticular disease and 15 months for diffuse intraarticular disease (11). However, in most cases—88% of localized extraarticular disease and 93% of diffuse intraarticular involvement—the onset of disease is chronic (duration of months to years); in the study by Myers and Masi (11), no patients presented with acute symptoms (duration of days). Intraarticular lesions may demonstrate intermittent, fluctuating symptoms with slow overall progression (11,14). A history of trauma has been found in 44%–53% of patients. Lesions are almost invariably monoarticular or solitary and are unassociated with other disorders (5). Polyarticular or multifocal involvement is distinctly unusual, but it appears more often in children, an otherwise rarely affected age group (11,15–17). PVNS in children has also been associated with a variety of other abnormalities, including vascular lesions, cherubism, multiple lentigines syndrome, extremity lymphedema, Noonan syndrome, and jaw lesions (5,6).

The distribution of lesion location also depends on the lesion subtype, either localized or diffuse, and on whether the lesion is intraarticular or extraarticular. PVNTS most commonly involves the hand or wrist (65%–89% of cases), specifically the index and long fingers (5,18–21). PVNTS represents the second most common soft-tissue mass of the hand and wrist and is exceeded in frequency only by ganglia (5,6). The volar aspect of the hand is affected approximately twice as often as the dorsal aspect (5–7,12). The next most common site of localized extraarticular disease is the foot and ankle, which account for 5%–15% of lesions (5–7,12). Rare sites of PVNTS include the knee, hip, elbow, and shoulder (22,23). PVNB most commonly occurs about the hip or knee in our experience. Localized intraarticular involvement of PVNS almost exclusively involves the knee (11,24) and represented 6% of all cases in the series of Myers and Masi (11).

Teaching Point

The diffuse intraarticular form of PVNS most frequently affects the large joints, with the knee involved in 66%–80% of cases (25,26). The hip is the second most commonly affected joint, accounting for 4%–16% of cases (4–8,12). Other large joints affected include the ankle, shoulder, and elbow in decreasing order of frequency (5,6,8,12). However, virtually any articular site may be involved, and rarely affected locations include the apophyseal joints of the spine, small joints of the hands or feet, sacroiliac joint, subtalar joints, and the temporomandibular joint (27–31).

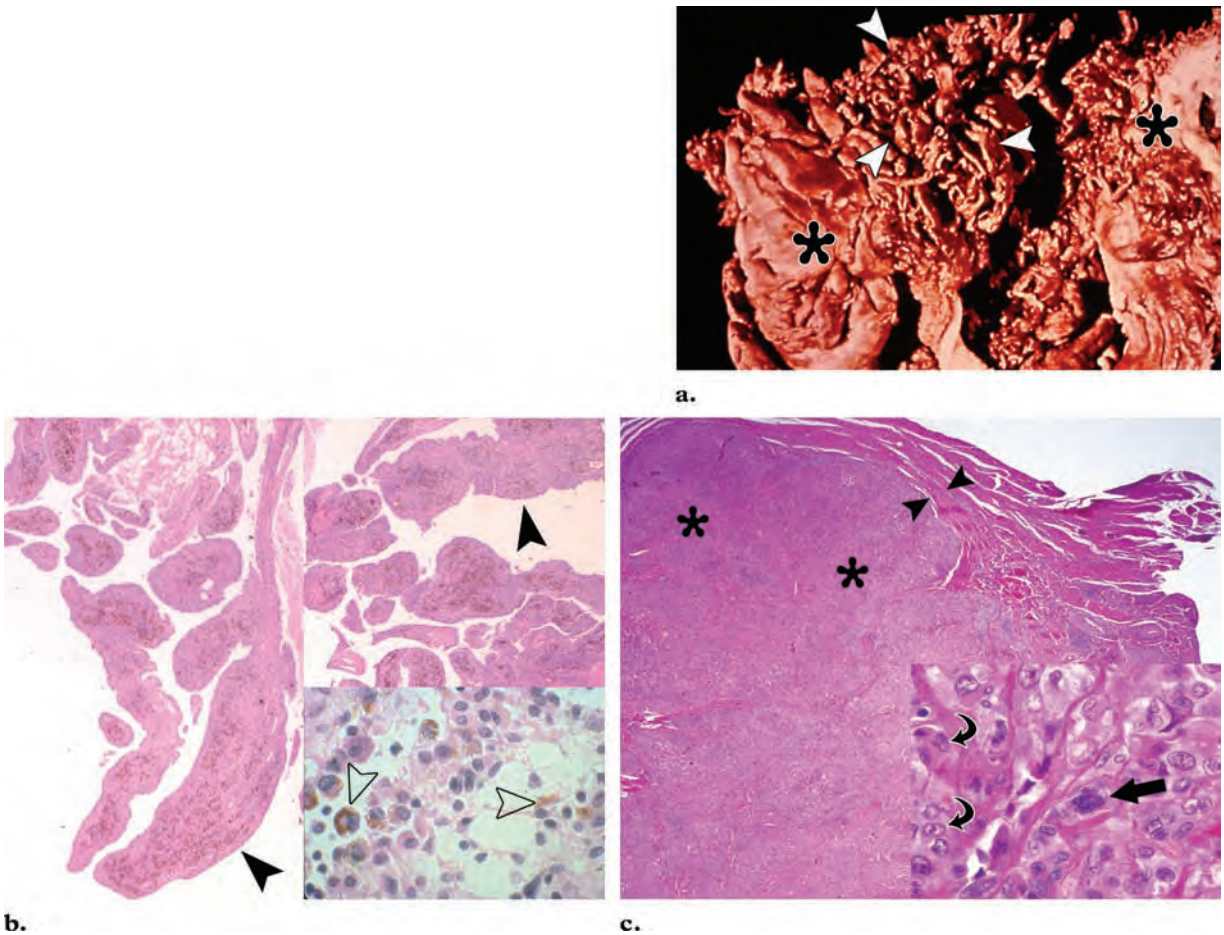


Figure 1. Pathologic characteristics of PVNS. **(a)** Photograph shows the typical appearance of diffuse intraarticular disease, which appears brown (*) owing to hemosiderin deposition and has villonodular protrusions (arrowheads). **(b)** Photomicrograph (original magnification, ×100; hematoxylin-eosin [H-E] stain) shows the villonodular fronds with overlying synovial tissue (solid arrowheads). Brownish areas represent extensive hemosiderin deposition. Inset photomicrograph (original magnification, ×400; H-E stain) demonstrates golden brown globular cytoplasmic intracellular hemosiderin (open arrowheads). **(c)** Photomicrograph of localized extraarticular PVNTS (original magnification, ×175; H-E stain) shows abrupt transition between the nodular tumor (*) and fibrous pseudocapsule (between arrowheads). Inset photomicrograph (original magnification, ×400; H-E stain) shows both multinucleated giant cells (straight arrow) and reniform mononuclear histiocytoid cells (curved arrows).

Pathologic Features

The gross pathologic appearance of PVNS varies, depending on the location (intraarticular or extraarticular) and type of disease (localized or diffuse) (Fig 1). The localized extraarticular forms of the disease PVNTS or PVNB generally manifest as a circumscribed, lobulated, cauliflower-like, nodular soft-tissue mass that is attached to the tendon sheath or that resides within a known bursal site (7,9,12) (Fig 1). Initially, the lesion may reveal a villous projection into the synovial space of the tendon sheath or bursa that blends with the synovial membrane; however, continued lobular growth of the tumor obscures this morphology and this appearance is not typically observed pathologically. Cut sections demonstrate pink-gray tumor with mottled brown or

yellow, depending on the amount of xanthoma cells (foamy histiocytes) or hemosiderin present. Lesion size typically ranges from 0.5 to 4.0 cm in greatest dimension (4,7,9,12).

The gross pathologic appearance of diffuse intraarticular PVNS is generally an infiltrative mass that extensively involves and thickens the synovium of the entire joint. The morphology of the synovial hypertrophy consists of irregular papillary or villous projections and larger nodular or villonodular protrusions (Fig 1). The villous architecture is best seen when the specimen is distended in water. The thickened synovium is typically mottled dark brown and yellow, depending on the amount of xanthoma cells, which appear yellow, and hemosiderin, which appears brown (7,9,12). Prominent xanthomatous areas have been mistaken for lipid components, which are not usually present in PVNS. Diffuse intraarticular lesions are larger than those seen in localized extraarticular disease, a feature that reflects the diffuse synovial involvement.

At microscopic analysis, localized extraarticular disease typically reveals a multinodular, well-delineated process embedded in a dense, partially collagenous pseudocapsule (4,7,9,12) (Fig 1). In contradistinction, diffuse intraarticular PVNS is not embedded in a collagenous pseudocapsule and has a diffuse infiltrative sheetlike growth pattern along synovium with cleftlike spaces and discohesive zones. All types of PVNS are composed of mononuclear histiocytoid cells with reniform nuclei and plump, eccentric eosinophilic cytoplasm, admixed with multinucleated giant cells and xanthoma cells (Fig 1). The multinucleated giant cells contain a variable number of nuclei, from three or four to 50 (4,7,9,12). Overall, the multinucleated giant cells are less conspicuous and not as uniformly dispersed in diffuse intraarticular PVNS. In fact, multinucleated giant cells may be absent in up to 20% of cases of diffuse intraarticular PVNS and are rarely seen in highly cellular regions (4,7,9,12). Hemosiderin deposition is usually apparent in all forms of PVNS (Fig 1). However, the extent of hemosiderin deposition varies, and hemosiderin is usually a much more prominent feature of diffuse intraarticular disease. In rare cases, chondroid or osteoid metaplasia with associated calcification is seen (18,32). Mitotic activity may be high, but generally, there are less than 10 mitoses per 10 high-power fields (4,7,9,12). Necrosis or sarcomatoid spindling should not be present. Both the mononuclear and multinucleated cells are positive for CD68 and CD163 (histiocytic markers), but the diagnosis is generally made from the H-E–stained tissue without use of immunohistochemical stains (7,12,33).

The overall histologic appearance of the diffuse intraarticular form of PVNS may be worrisome for a more aggressive malignant entity such as rhabdomyosarcoma, synovial sarcoma, or epithelioid sarcoma (4,7,9,12). These features include extremely large size of the cells, invagination of cytoplasm into the nucleus that resembles large nucleoli, and increased numbers of mitoses. Correlation of the histologic features with the imaging findings of a diffuse synovial process is essential to arrive at the correct diagnosis of a benign lesion (Fig 2).

Teaching Point

The etiology of PVNS has historically been attributed to either an inflammatory process; repeated hemorrhage into the joint, perhaps related to an occult synovial hemangioma or repetitive mild trauma; neoplasia; or a disorder of lipid metabolism. However, the capability of these lesions for autonomous growth, the identification of malignant transformation, and the more recent detection of cytogenetic aberrations all strongly support the hypothesis that PVNS has a neoplastic origin. The most consistent genetic rearrangement in both localized and diffuse types of PVNS is in chromosome 1p11-13, a site for *CSF-1* gene, which most commonly fuses to *COL6a3* on chromosome 2q35 (34,35). Trisomy of chromosomes 5 and 7 has also been reported in diffuse and malignant PVNS (8,36). These cytogenetic aberrations are seen in the majority but not all cases of PVNS.

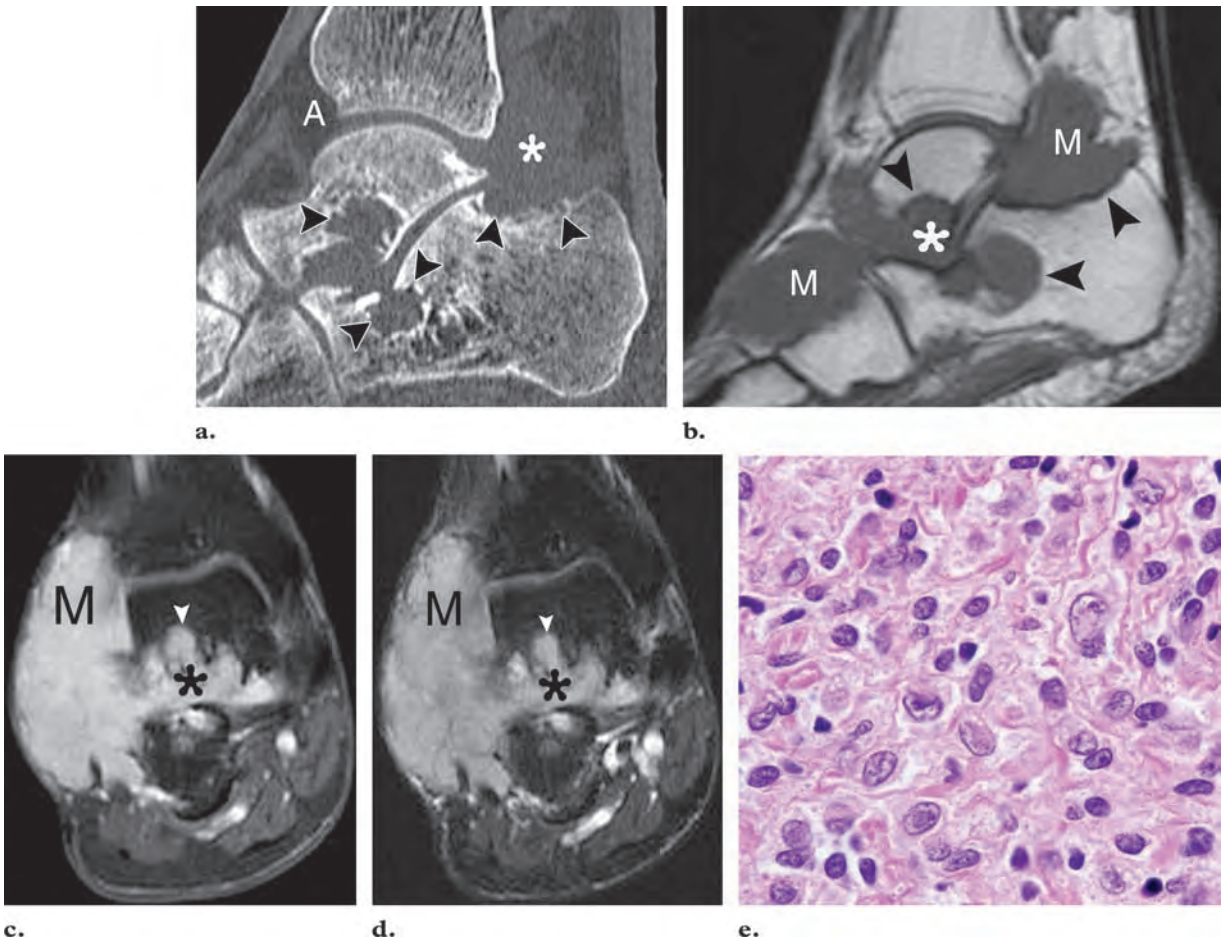
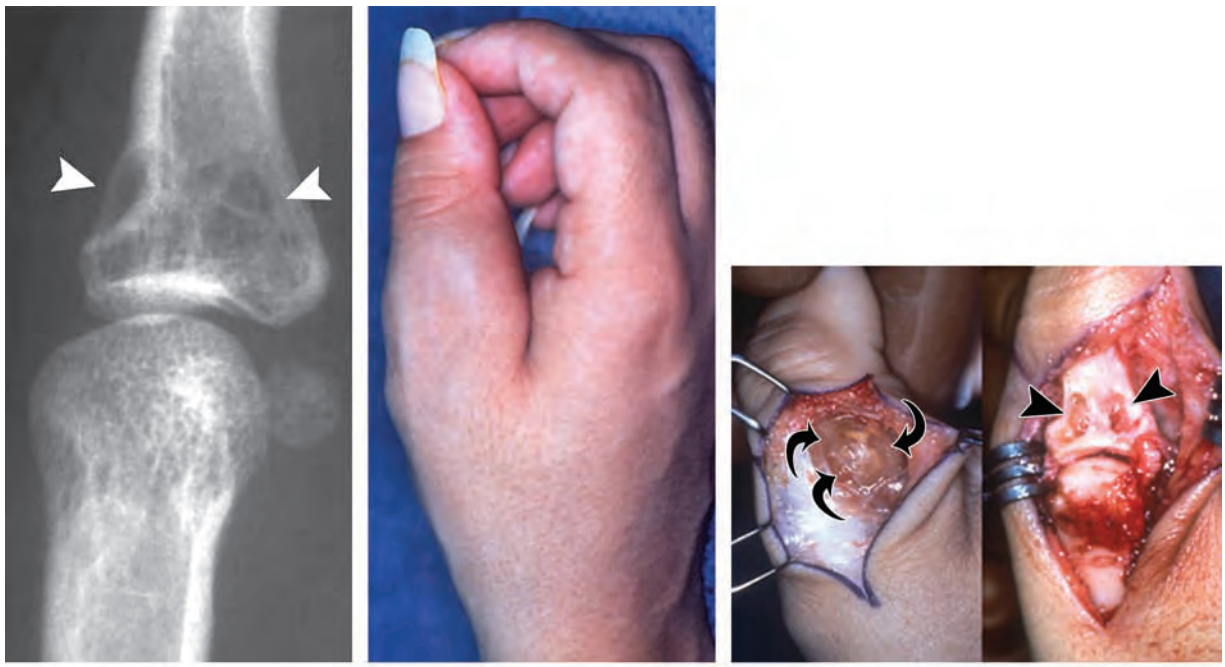


Figure 2. Diffuse intraarticular PVNS in an unusual site, affecting the subtalar joint in a 63-year-old man with hindfoot pain. **(a)** Sagittal CT scan shows extensive erosion about the subtalar joint (arrowheads) and large soft-tissue components posteriorly (*) with sparing of the ankle joint, as evidenced by normal fat in the region of the anterior recess *(A)*. **(b–d)** Sagittal T1-weighted (448/127 = repetition time msec/echo time msec) precontrast **(b)**, short-axis T1-weighted (565/10) postcontrast **(c)**, and short-axis short inversion time inversion recovery (STIR) (4000/48, 180° inversion pulse) **(d)** MR images reveal that the large mass emanates from the subtalar joint (*) with the largest component in the surrounding soft tissues *(M)* because of the small capacity of the site of origin. There is low to intermediate signal intensity on T1-weighted **(b)** and STIR **(d)** MR images, prominent diffuse enhancement, and bone erosion (arrowheads). **(e)** Photomicrograph (original magnification ×400; H-E stain) of a specimen from the lesion was initially viewed as worrisome for malignancy owing to the extremely large tumor cell size, apparent cytologic atypia, and paucity of multinucleated giant cells. However, correlation of histologic findings with imaging results reinforces the articular origin of the lesion and helps confirm the diagnosis of diffuse intraarticular PVNS with soft-tissue extension.

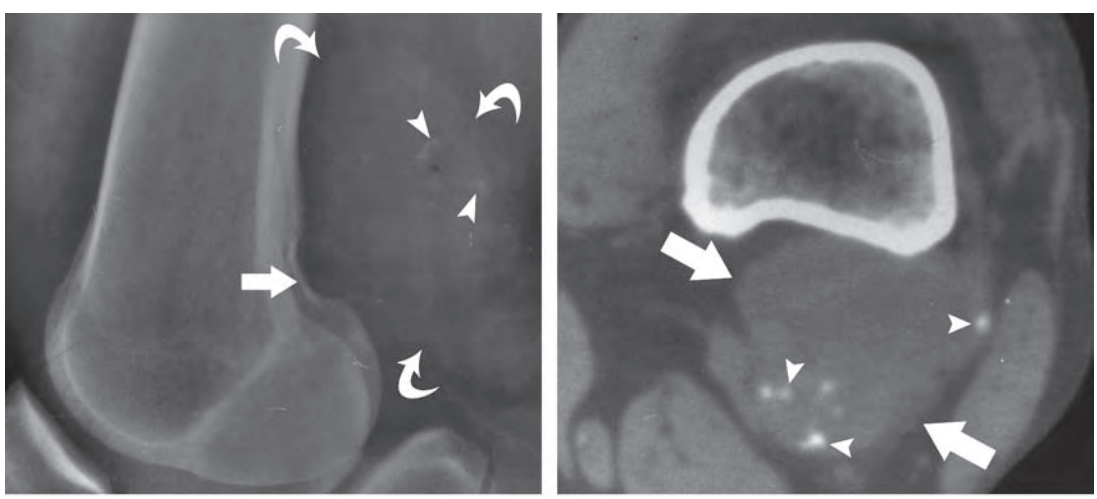
Imaging Features

Radiographic appearances of PVNS vary by the subtype, localized or diffuse, and the specific joint involved with intraarticular lesions. The localized extraarticular subtype of this disease, PVNTS or PVNB, typically manifests as a soft-tissue mass in 50%–70% of cases (37–40) (Fig 3). However, radiographic findings are normal in up to 20% of cases (5,37–40). Overall, osseous abnormalities are present in 15%–25% of cases (5,37,38,40,41). Extrinsic erosion, often with well-defined sclerotic margins, of the underlying bone is the most common osseous abnormality, seen in 9%–25% of cases (5,37–41) (Figs 3, 4). This feature appears more often in lesions of the foot or ankle, owing to the overlying dense



a. b. c.

Figure 3. Localized extraarticular PVNTS involving the thumb in a 26-year-old woman with a palpable, slowly enlarging, soft-tissue mass. **(a)** Oblique radiograph of the thumb shows soft-tissue swelling and well-defined areas of subchondral lysis with sclerotic margins that represent bone erosions (arrowheads). **(b)** Clinical photograph reveals soft-tissue swelling and a soft-tissue mass involving the thumb. **(c)** Intraoperative photographs demonstrate the nodular brownish soft-tissue mass (curved arrows) and, following lesion resection, extrinsic erosion of the proximal phalanx (arrowheads). The erosion corresponds to that seen on the radiograph **(a)**.



a. b.

Figure 4. Localized extraarticular PVNB in a bursa posterior to the knee in a 30-year-old man with a palpable soft-tissue mass. **(a)** Lateral radiograph of the knee shows a soft-tissue mass (curved arrows) with small foci of calcification (arrowheads) and subtle extrinsic erosion of the femur (straight arrow). **(b)** Axial CT scan also reveals the soft-tissue mass (arrows) and small areas of calcification (arrowheads).

ligaments that restrict outward growth of lesions (38,42). The extrinsic erosions can be quite deep, a finding that simulates marrow invasion and a more aggressive process (Fig 2), although this feature is rare. Additional radiographic features associated with GCTTS include periosteal reaction (8% of cases) and calcifications (6%) (14,41) (Fig 4).

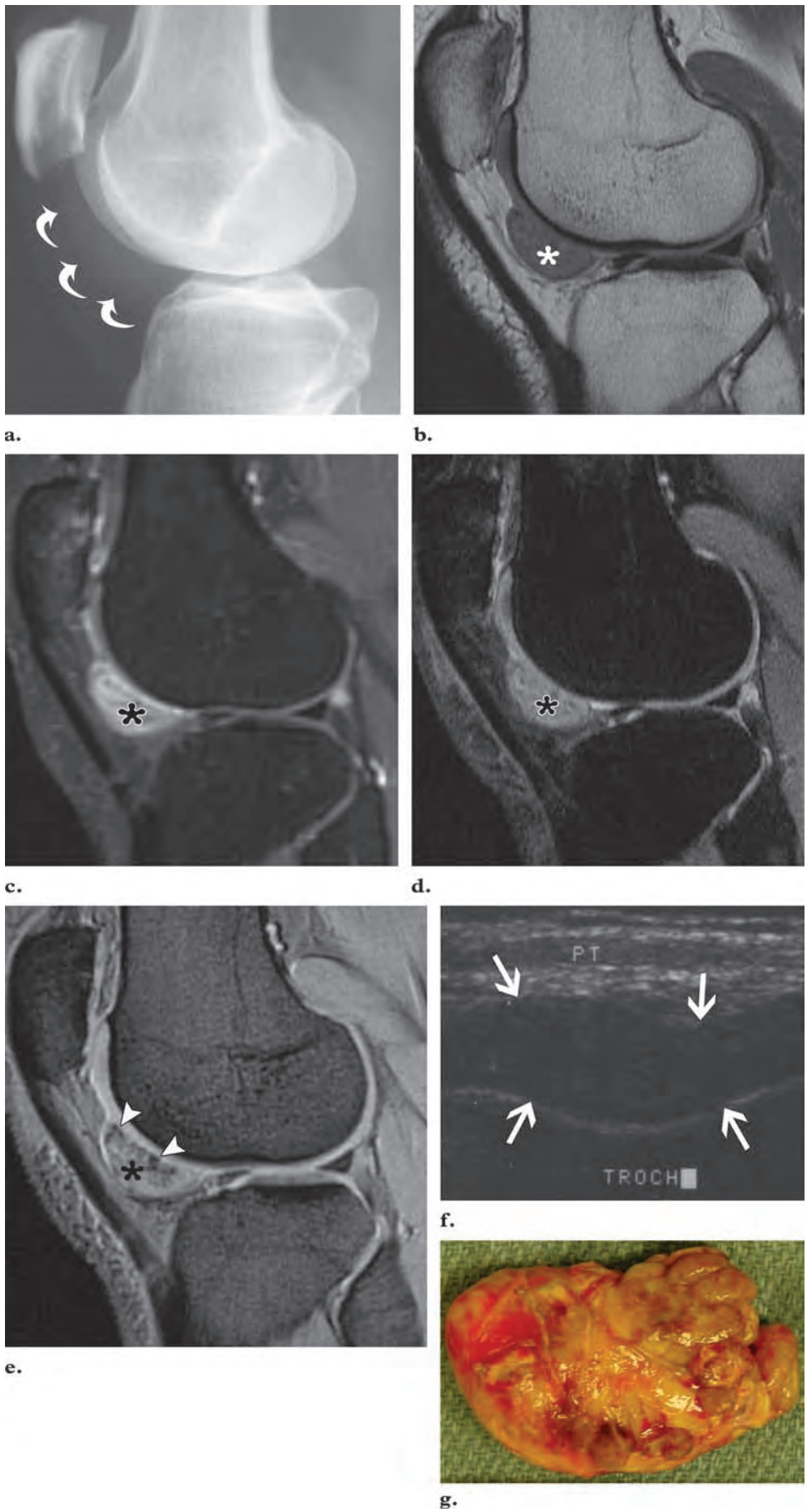


Figure 5. Localized intraarticular PVNS of the knee in a 40-year-old woman with knee pain. **(a)** Lateral radiograph shows an ill-defined area of soft-tissue opacity that replaces the normal Hoffa fat pad (arrows). **(b, c)** Sagittal proton-density–weighted (2150/32) **(b)** and T1-weighted (567/13) fat-suppressed postcontrast **(c)** images reveal the localized intermediate-signal-intensity soft-tissue mass (*), which has prominent diffuse enhancement. **(d, e)** Sagittal proton-density–weighted (2600/32) fat-suppressed **(d)** and gradient-echo (450/10, 30° flip angle) **(e)** images show overall increased signal intensity of the intraarticular soft-tissue mass (*). The gradient-echo image **(e)** also shows focal hypointense areas (arrowheads), findings that represent the blooming artifact from hemosiderin. **(f)** Transverse sonogram of the knee reveals the hypoechoic intraarticular soft-tissue mass (arrows). *PT* = patellar tendon, *TROCH* = trochlear notch. **(g)** Photograph of the resected specimen shows the brownish appearance caused by hemosiderin.

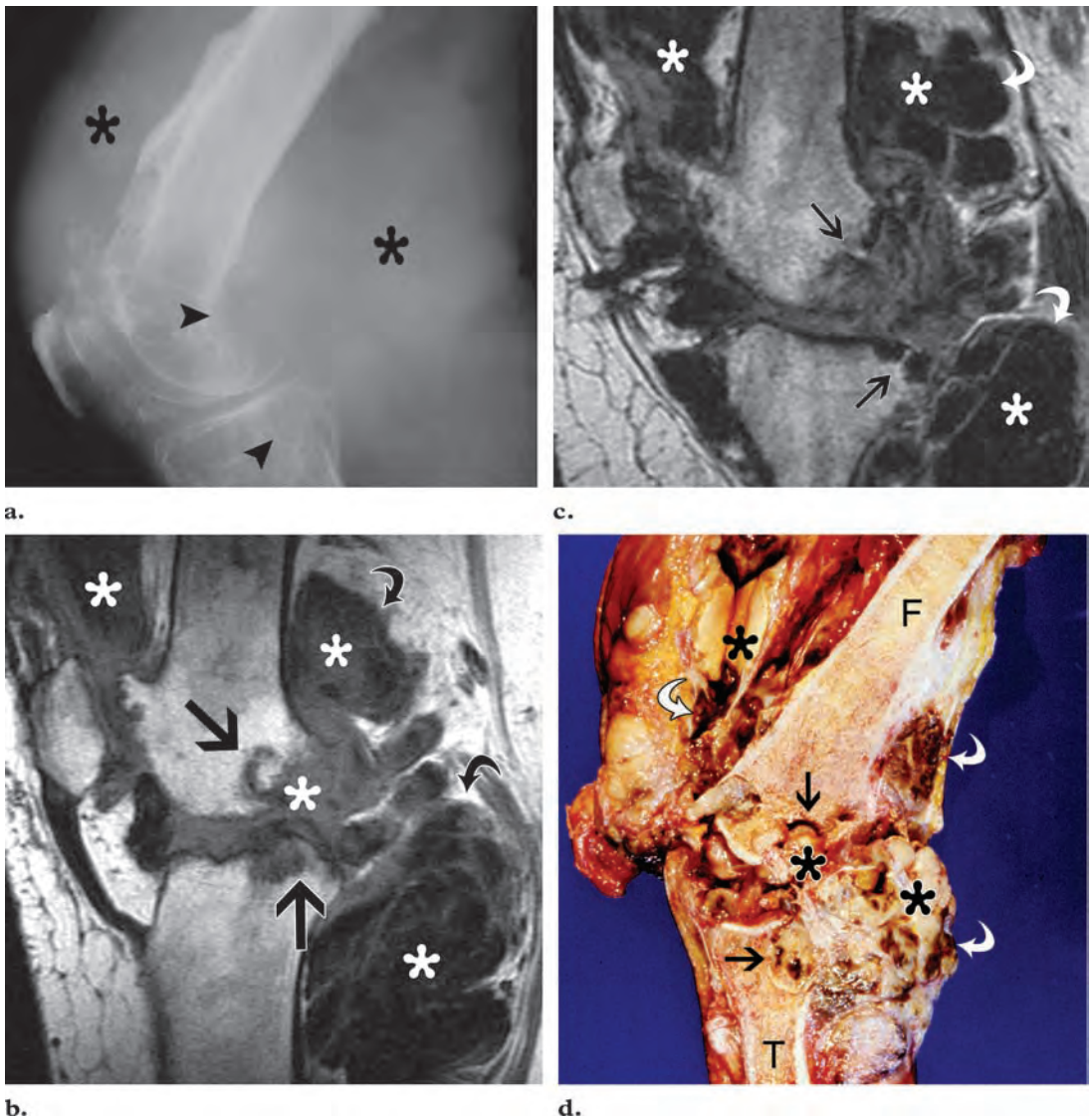


Figure 6. Diffuse PVNS of the knee in a 45-year-old man; multiple recurrences of PVNS ultimately led to amputation. **(a)** Lateral radiograph shows a large suprapatellar effusion and soft-tissue fullness about the knee (*). There are extrinsic erosions with marginal sclerosis (arrowheads) on both sides of the knee joint and patellofemoral degenerative changes. **(b, c)** Sagittal T1-weighted (684/14) **(b)** and gradient-echo (21/7, 40° flip angle) **(c)** MR images reveal a large amount of low-signal-intensity tissue (*) with posterior extension (curved arrows) that replaces the entire knee joint. Extrinsic erosions of the femur and tibia with low-signal-intensity margins are also seen (straight arrows). Mild blooming artifact is seen on the gradient-echo image **(c)**. **(d)** Photograph of the sagittally sectioned gross specimen demonstrates the large mass throughout the knee joint (*). The prominent brownish areas (curved arrows) result from more prominent hemosiderin deposition. Extrinsic erosions (straight arrows) of the femur *(F)* and *tibia* (T) are also seen.

The localized intraarticular form of PVNS appears normal on radiographs in most cases (11,24). An area of soft-tissue opacity that replaces the normal region of adipose tissue in the Hoffa fat pad may be seen (Fig 5). Myers and Masi (11) noted extrinsic erosion of bone in 20% of their cases involving the knee, although in our experience this finding is more unusual.

Radiographs of patients with diffuse intraarticular PVNS may appear normal in up to 21% of cases (38,39,43–45). However, more commonly, radiographs demonstrate joint effusion, soft-tissue swelling, absence of calcification, extrinsic erosion of bone, preservation of joint space, and normal bone mineralization (38,39,43–45) (Fig 6). Extrinsic erosion of bone and subchondral lucent areas are well-defined geographic lesions that frequently display a rim of sclerosis and that

Teaching Point

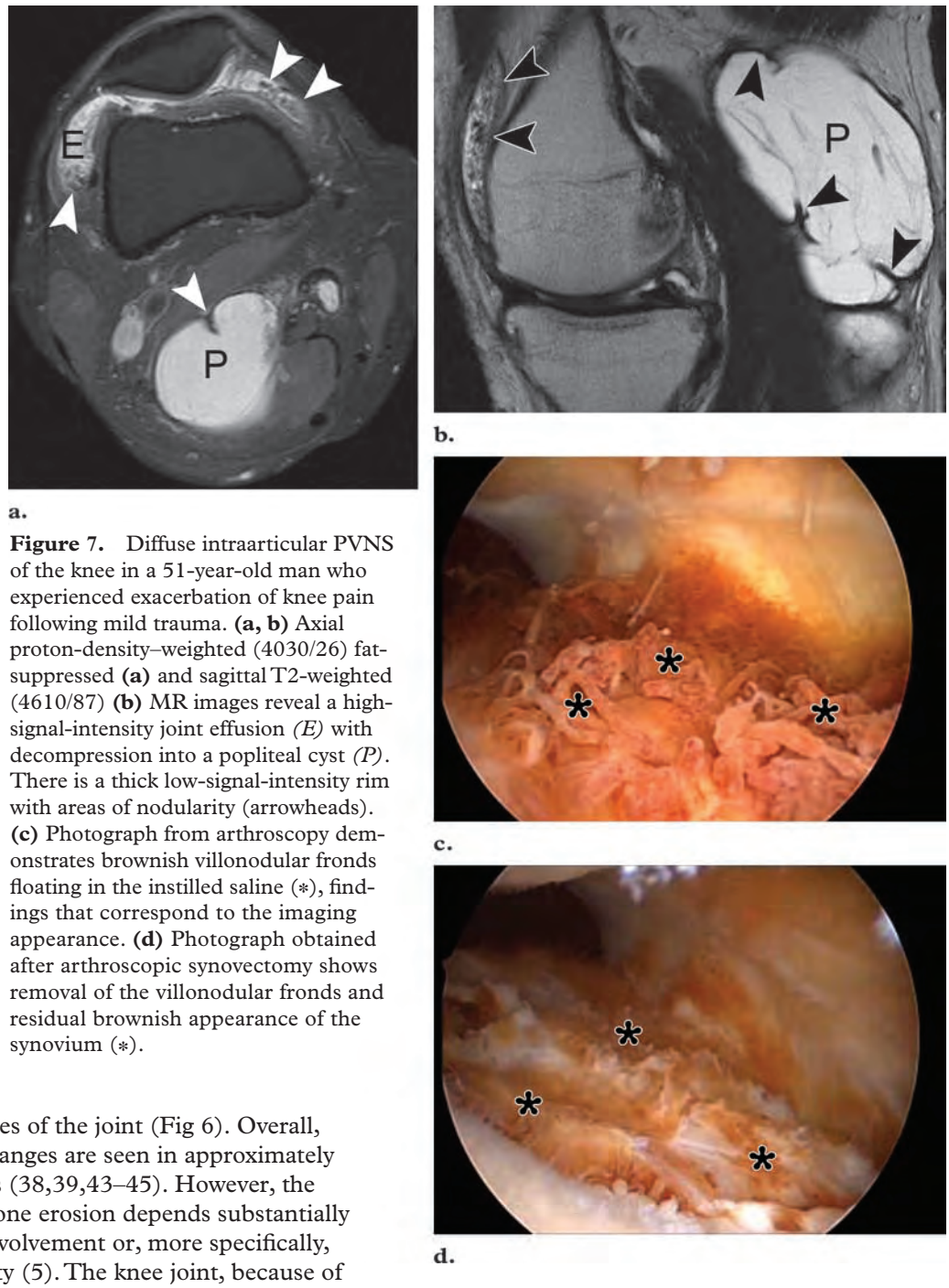
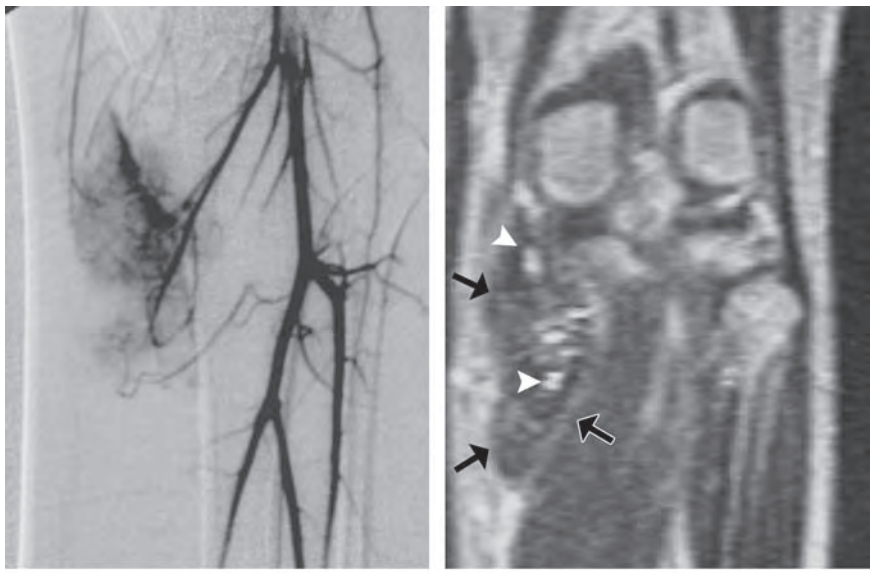


Figure 7. Diffuse intraarticular PVNS of the knee in a 51-year-old man who experienced exacerbation of knee pain following mild trauma. **(a, b)** Axial proton-density–weighted (4030/26) fat-suppressed **(a)** and sagittal T2-weighted (4610/87) **(b)** MR images reveal a high-signal-intensity joint effusion *(E)* with decompression into a popliteal cyst *(P)*. There is a thick low-signal-intensity rim with areas of nodularity (arrowheads). **(c)** Photograph from arthroscopy demonstrates brownish villonodular fronds floating in the instilled saline (*), findings that correspond to the imaging appearance. **(d)** Photograph obtained after arthroscopic synovectomy shows removal of the villonodular fronds and residual brownish appearance of the synovium (*).

involve both sides of the joint (Fig 6). Overall, these erosive changes are seen in approximately 50% of all cases (38,39,43–45). However, the prevalence of bone erosion depends substantially on the site of involvement or, more specifically, the joint capacity (5). The knee joint, because of its large capacity that allows extension and decompression of normal tissue into multiple adjacent bursal regions (such as the suprapatellar and gastrocnemius/semimembranosus bursae), is less frequently affected by extrinsic erosion of bone (Fig 7). This characteristic has been described in the radiology literature and in the review by Dorwart and Czerniak (7), who reported that osseous erosions were seen in only 26%–32% of PVNS cases involving the knee joint (Fig 6). In contradistinction, smaller capacity joints such as the hip, shoulder, elbow, and ankle are much more likely to demonstrate extrinsic erosion of bone on radiographs (seen in 93%, 75%, 63% and 56% of cases, respectively [7]). Less common radiographic features associated with diffuse intraarticular PVNS include osteopenia (7% of



a. b.

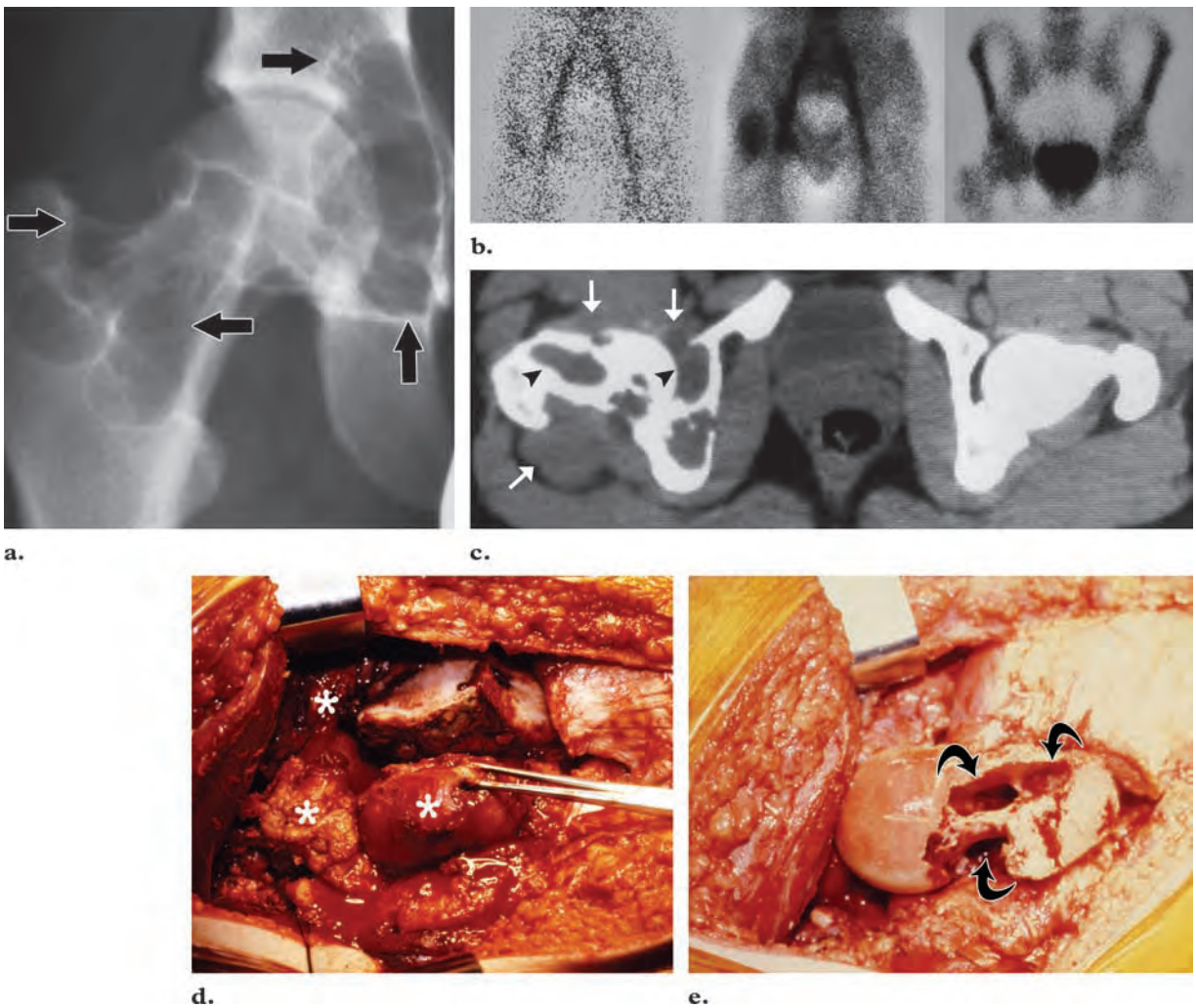
Figure 8. Localized extraarticular PVNB about the pes anserine bursa in a 26-year-old woman who presented with a slowly enlarging, soft-tissue mass in the medial knee. **(a)** Anteroposterior view from conventional late-arterial-phase arteriography shows prominent hypervascularity of the mass. **(b)** Coronal T2-weighted (2000/80) MR image reveals prominent low signal intensity of the mass (arrows), a finding that results from hemosiderin deposition. Small foci of high signal intensity (arrowheads) likely represent fluid surrounded by hemosiderin-laden tissue, an appearance similar to that of the cleft sign described in localized intraarticular PVNS.

cases), joint space narrowing (7%), intraarticular osteochondral bodies (7%), and degenerative disease (4%) (Fig 6) (7,11,39,43–45). An arthritis mutilans appearance may be seen in rare cases of PVNS, particularly subsequent to multiple recurrences.

Arthrography of diffuse intraarticular PVNS (typically followed by CT or MR imaging) reveals extensive synovial thickening with villous or nodular projections that extend into the joint (5,7,46). This appearance is relatively nonspecific, since an inflammatory arthritis such as rheumatoid arthritis would exhibit similar findings. However, multiple filling defects, such as those seen in primary synovial chondromatosis, are not an arthrographic finding of diffuse intraarticular PVNS. The localized intraarticular form of PVNS may show a singular nodular area of synovial thickening at arthrography. The fluid that is aspirated before injection of contrast material for arthrography typically demonstrates bloody effusion (69%–75% of cases), yellow fluid (22%–25%), or brownish fluid (9%) (5,7,11).

Significant vascularity develops in most cases of PVNS (all types, although the degree of vascularity varies among them) and is seen at angiography and bone scintigraphy (5,46) (Figs 8, 9). Angiography frequently shows prominent neovascularity, mild arteriovenous shunting, and tumor blush that may simulate a malignant neoplasm (5,7) (Fig 8). Blood flow and blood pool images from three-phase bone scintigraphy often reveal increased radionuclide uptake, which is substantially more prominent than the activity seen on delayed images (5,7) (Fig 9). Thallium-201 scintigraphy also demonstrates radionuclide accumulation in PVNS on both early and delayed images (5). In addition, PVNS has shown hypermetabolic activity at fluorine-18 fluorodeoxyglucose position emission tomography (PET) (47,48), with average and maximum standard uptake values of 5.9 and 11.3, respectively (48). Overall, these nuclear medicine studies reveal more focal abnormality in localized PVNS and more extensive radionuclide accumulation in diffuse intraarticular lesions.

Figure 9. Diffuse intraarticular PVNS of the hip in an 18-year-old man with hip pain of 2 years duration. **(a)** Anteroposterior radiograph shows extensive erosion of the femoral neck and acetabulum with sclerotic margins (arrows) and maintained hip joint space. **(b)** Images from three-phase bone scintigraphy reveal prominent increased radionuclide activity during the blood flow (left) and blood pool (middle) phases but only mild activity during the delayed phase (right), findings that reflect increased blood flow in PVNS. **(c)** Axial CT scan demonstrates features similar to those seen in **a**, with extensive erosions (arrowheads) about the hip and synovial thickening (arrows) that is most prominent posteriorly. **(d, e)** Intraoperative photographs obtained during open arthrotomy show nodular, brownish, hemosiderin-laden tissue (∗ in **d**) and, after complete synovectomy, extrinsic erosion of the femoral neck (arrows in **e**).



Sonographic appearances of intraarticular PVNS are nonspecific. Sonographic features of diffuse intraarticular disease include joint effusion, complex heterogeneous echogenic masses, and markedly thickened hypoechoic synovium that may have nodular and villous projections (5,39,46,49) (Fig 10). Extrinsic erosion of underlying bone may also be seen. Localized intraarticular PVNS has features similar to those seen in diffuse disease but only a solitary focal synovial mass is detected (Fig 5). PVNTS manifests as a hypoechoic solid mass with well-defined margins that is intimately related to the associated involved tendon (49). We believe a helpful hint for making the correct diagnosis of GCTTS is to observe the length (mean, 5.7 cm) and circumference (mean, 136°) of involvement about the tendon; these findings suggest that the lesion has a tenosynovial origin, as described by Middleton and co-workers (49). However, because the lesion arises from the tendon sheath, GCTTS does not move with the tendon during dynamic sonography (39,49). Doppler imaging commonly reveals increased blood flow in all types of PVNS (Fig 10).

Figure 10. Diffuse intraarticular PVNS of the knee in an 8-year-old girl with a 5-year history of intermittent knee pain and swelling. **(a)** Lateral radiograph shows a radiopaque suprapatellar effusion (*) and thickening about the patellar tendon. **(b)** Sagittal gradient-echo (27/9, 30° flip angle) image reveals prominent, diffuse hypointense synovial thickening (arrows). The blooming artifact is more prominent here, with the gradient-echo sequence, than with T1-weighting, owing to hemosiderin. **(c)** Coronal T2-weighted (7000/60) fat-suppressed image shows cartilage thinning caused by the chronic synovial process with a cleft extending through the articular surface (arrow) and an underlying hyperintense subchondral cyst (*) surrounded by hypointense hemosiderin-laden tissue. **(d)** Axial T1-weighted (418/12) fat-suppressed postcontrast image shows enhancing villonodular synovial fronds (arrowheads). **(e)** Long-axis Doppler sonogram shows the hypervascular synovial fronds (arrowheads) surrounded by the suprapatellar bursal fluid. **(f)** Photograph obtained during arthrotomy resection reveals villonodular protrusions (arrows) and brown discoloration, owing to extensive hemosiderin.

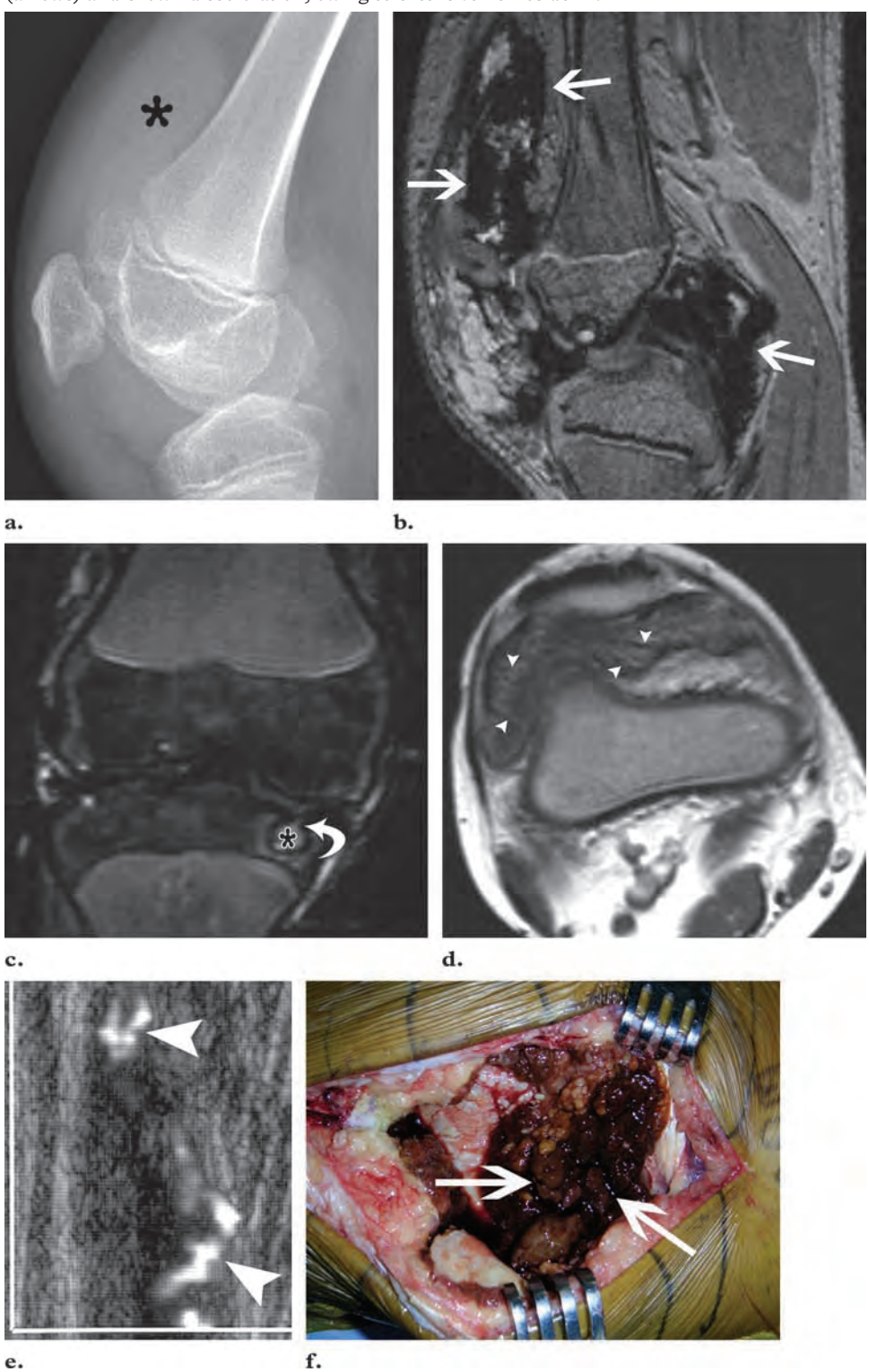
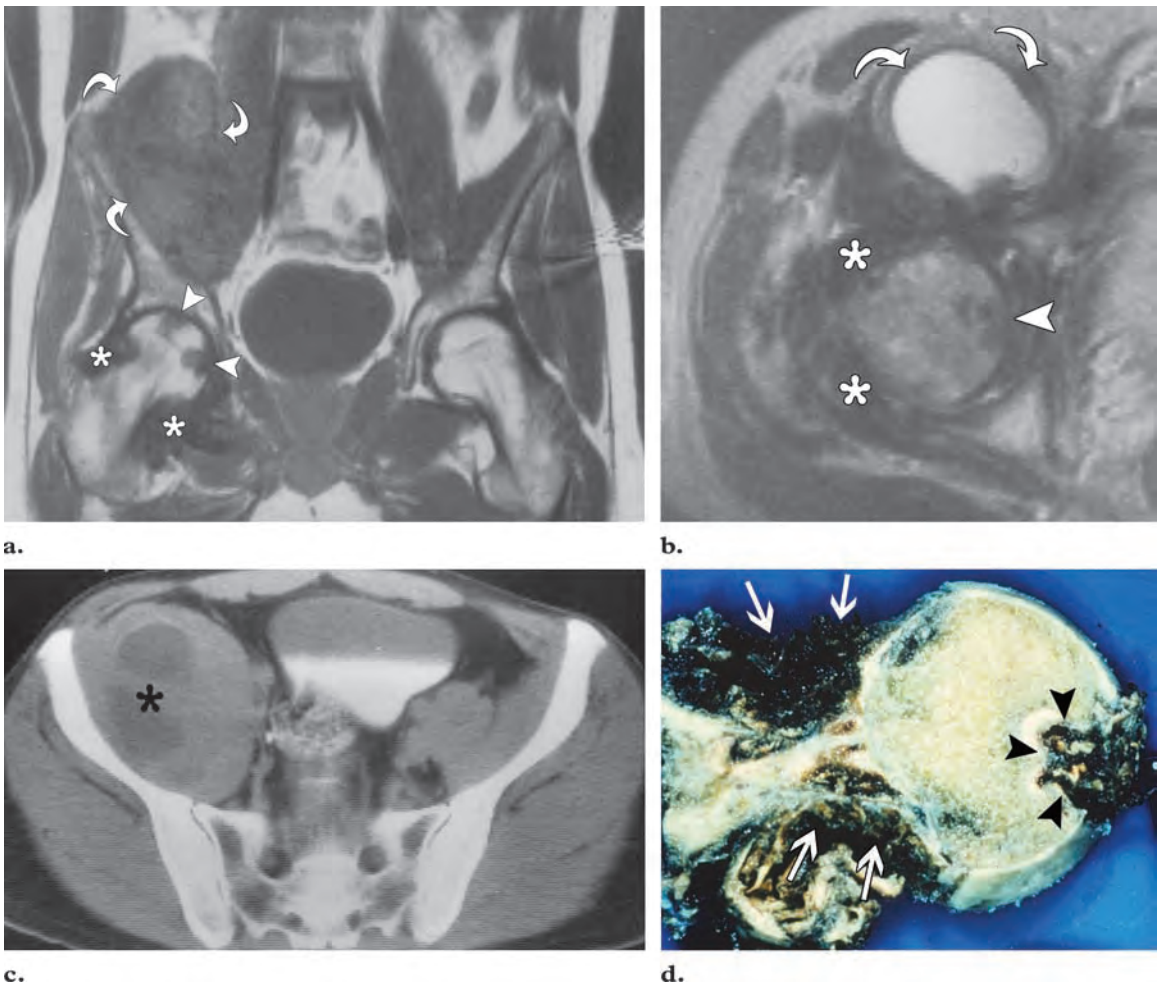


Figure 11. Diffuse intraarticular PVNS in the hip of a 29-year-old man with pain and limited range of motion. **(a, b)** Coronal T1-weighted (500/30) **(a)** and axial T2-weighted (2000/75) **(b)** MR images show femoral head erosions (arrowheads) and synovial thickening (*) about the hip, with low signal intensity on **b**. The involvement extends superiorly to the iliopsoas bursa (arrows), a substantial distance from the joint. **(c)** Axial CT scan also reveals involvement of the iliopsoas bursa (*), which normally communicates with the hip joint in 10%–15% of people. **(d)** Photograph of the sectioned gross specimen shows synovial thickening (arrows) and erosion in the region of the fovea (arrowheads).



CT reveals synovial thickening in diffuse intraarticular PVNS (5,46,50) (Figs 2, 9). Its attenuation may be slightly increased relative to that of muscle, a finding that reflects hemosiderin deposition as seen in 29% of seven cases described by Jelinek et al (50). It is also frequently associated with low-attenuation joint effusion. Lesion extent is not as well depicted with CT, compared with MR imaging, owing to the more limited contrast resolution of CT (Figs 2, 11). However, CT is optimal for demonstrating extrinsic erosion of bone on both sides of the joint, and it may also reveal subchondral cyst formation (Figs 2, 9). The CT appearance of the localized form of PVNS, either intraarticular or extraarticular, has not been extensively described. In our experience, localized PVNS manifests as a nonspecific well-defined soft-tissue mass with attenuation similar to that of adjacent muscle (Figs 4, 12). Mildly increased attenuation is uncommon (Fig 12), a characteristic that likely reflects the more variable amount of hemosiderin in these lesions, compared with diffuse intraarticular PVNS (5,50).

MR imaging is typically used to evaluate diffuse intraarticular PVNS after radiography because of the nonspecific clinical symptoms of a monoarticular arthropathy (6,39,45,46,50–54). MR imaging reveals characteristic features of a heterogeneous, diffuse, synovially based, plaquelike thickening that

Figure 12. PVNB of the greater trochanteric bursa in a 55-year-old man with a slowly enlarging, soft-tissue mass in the buttock. **(a, b)** Axial **(a)** and coronal **(b)** CT scans show a soft-tissue mass (*) adjacent to the greater trochanter and extending superiorly. The lesion has attenuation similar to that of adjacent muscle, with small areas of slightly higher attenuation. **(c, d)** Axial T1-weighted (680/10) fat-suppressed postcontrast **(c)** and sagittal T1-weighted (676/10) **(d)** MR images also reveal the intermuscular soft-tissue mass (*) with surrounding fat in the expected location of the greater trochanteric bursa. The mass shows intermediate signal intensity and prominent diffuse enhancement. **(e)** Coronal STIR (6210/35, 150° inversion pulse) MR image shows intermediate to high signal intensity in the mass (*), which surrounds the greater trochanter (arrow). There is no evidence of hip joint involvement. **(f)** Photograph of the sectioned gross specimen shows the massively distended greater trochanteric bursa (*) with a narrowed neck distally (arrow). Extensive brownish discoloration from hemosiderin is seen (arrowheads).

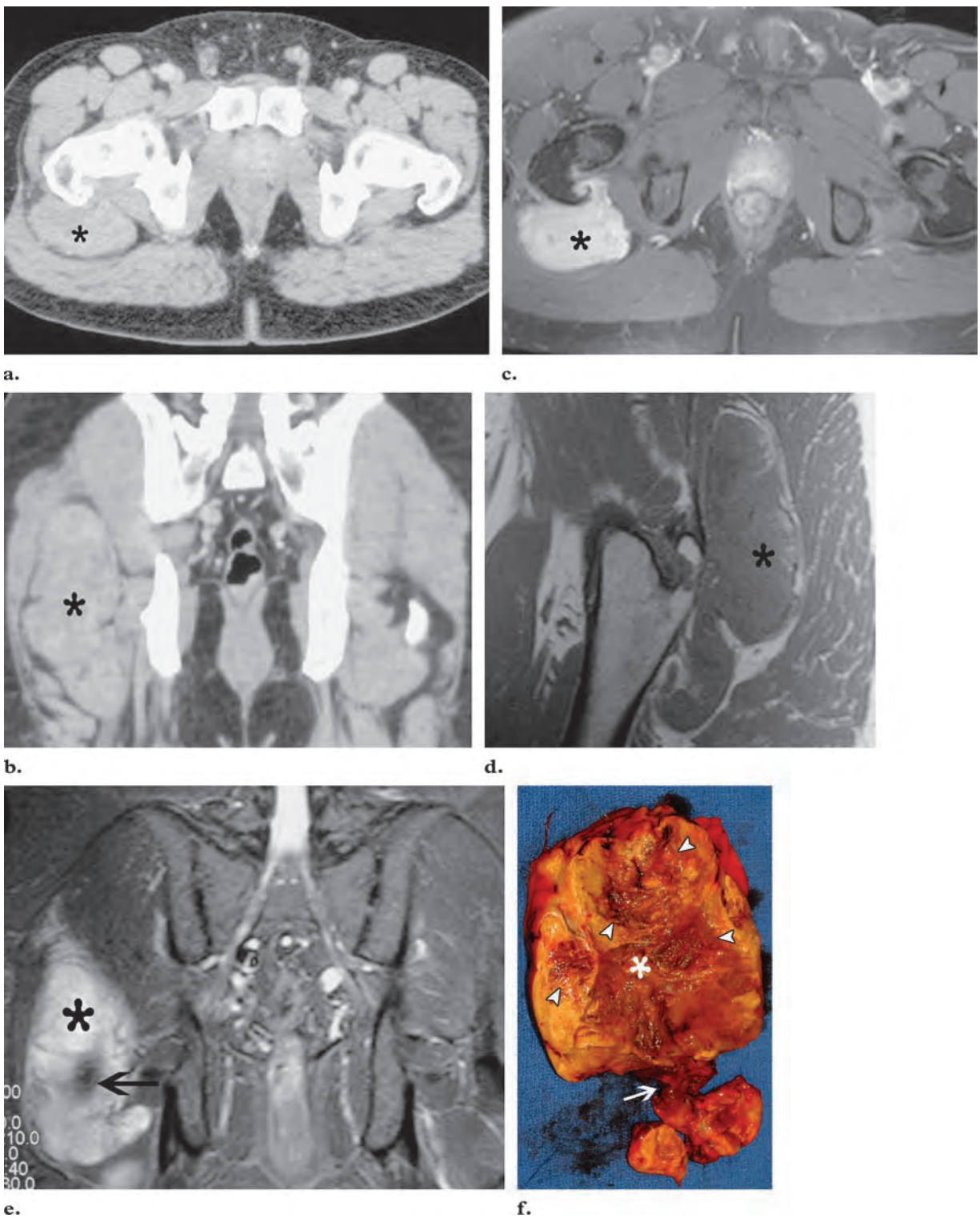
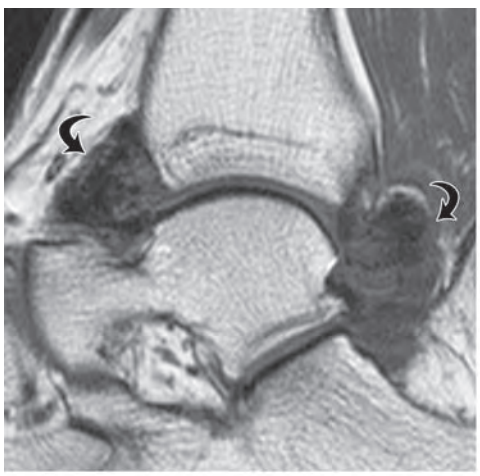
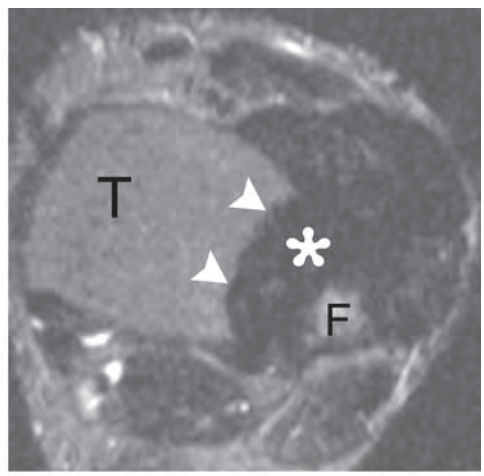


Figure 13. Diffuse intraarticular PVNS of the ankle in a 43-year-old man with pain and a soft-tissue mass. **(a, b)** Sagittal T1-weighted (600/20) **(a)** and axial T2-weighted (4830/90) **(b)** MR images show involvement of the anterior and posterior recesses of the ankle (arrows in **a**), syndesmotic extension (∗ in **b**), and osseous erosion (arrowheads in **b**). The mass has intermediate signal intensity with T1-weighting and predominantly low signal intensity with T2-weighting. *F* = fibula, *T* = tibia. **(c)** Intraoperative photograph shows the syndesmotic component (∗) with prominent brownish areas from hemosiderin deposition.



a.



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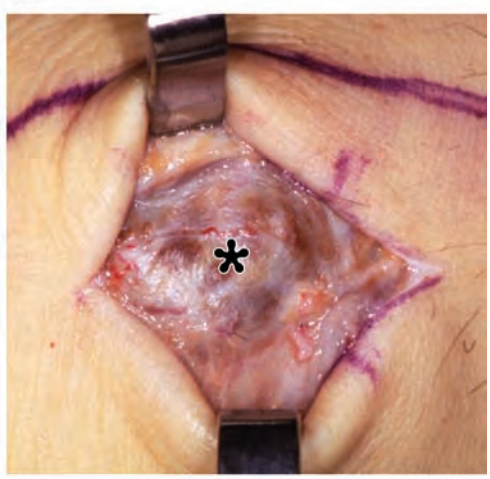


c.

often is associated with nodularity (Figs 2, 6, 7, 10, 11, 13). A villous or villonodular appearance is seen less frequently in our experience (Fig 10). Associated joint effusion is common, particularly in large joints such as the knee, but the effusion is generally surrounded by thickened synovial rinds of hemosiderin-laden tissue (Figs 7, 10). The signal intensity of the synovial thickening is intermediate to low on T1-weighted images (Figs 2, 6, 11). Low signal intensity also predominates on T2-weighted MR images, owing to the preferential shortening of T2 relaxation time caused by hemosiderin, an effect that is accentuated at higher field strength (Figs 7, 11, 13).

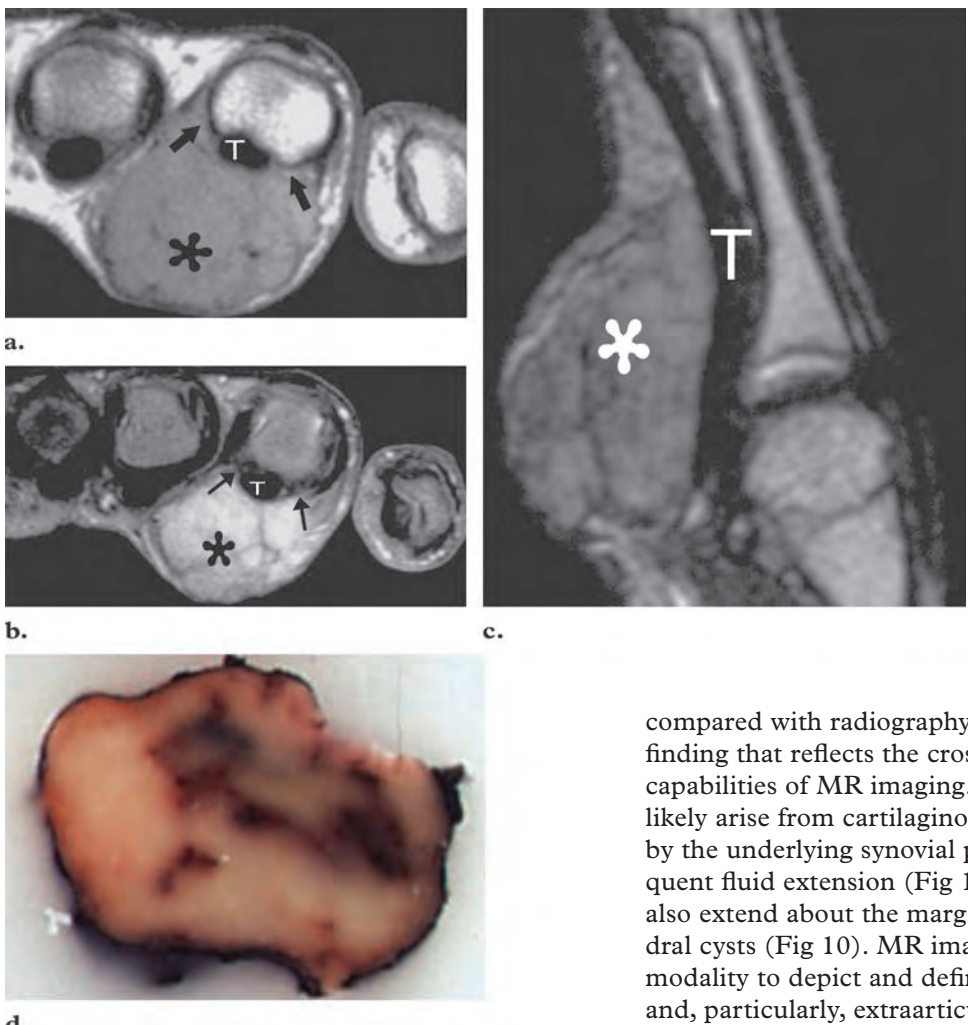
Teaching Point

This effect is particularly pronounced on gradient-echo images, which demonstrate an enlargement of the low-signal-intensity areas ("blooming") that is caused by magnetic susceptibility artifact (Figs 6, 10). The blooming effect, which specifically signifies the presence of hemosiderin as the cause of low signal intensity, is nearly pathognomonic of PVNS at MR imaging. Although synovial hemangioma and hemophiliac arthropathy may show similar MR imaging findings (caused by repetitive intraarticular hemorrhage and synovial hemosiderin deposition), diffuse intraarticular PVNS can be distinguished from these conditions because it is not associated with either serpentine vascular channels (hemangioma) or a clinical history of hemophilia (5,7).

Teaching Point

STIR MR images have been reported to reveal predominantly high signal intensity in PVNS (Fig 2). However, we have not observed this finding, as most cases show intermediate signal intensity on STIR images, and we believe this suggested appearance may be related to windowing and leveling of the images (Figs 2, 12). Areas of intermixed high signal intensity that represent lipid-laden macrophages on T1-weighted images have also been reported in the literature in PVNS (5,45,53). We agree with Bravo et al (46) that this intermixed high signal intensity is an uncommon finding. In addition, we believe that this finding may also represent entrapped fronds of perisynovial fat, a view supported by histopathologic analysis, which reveals xanthomatous tissue but does not demonstrate

Figure 14. Localized extraarticular PVNTS of a flexor tendon sheath of the index finger in a 25-year-old man with a slowly enlarging soft-tissue mass for 2 years. **(a, b)** Axial T1-weighted (650/19 in **a**, 800/17 in **b**) precontrast **(a)** and fat-suppressed postcontrast **(b)** MR images show an intermediate-signal-intensity soft-tissue mass (*) adjacent to and partially encasing (arrows) the flexor tendon *(T)*. The mass is diffusely enhanced in **b**. **(c)** On a sagittal T2-weighted (5000/92) MR image, the mass (*) has heterogeneous, predominantly intermediate signal intensity without prominent low-signal-intensity areas. *T* = tendon. **(d)** Photograph of the sectioned gross specimen shows the predominantly yellowish to tan, soft-tissue mass with a smaller brownish component resulting from a more prominent xanthomatous component (not shown).



fat. Enhancement of PVNS is common after intravenous administration of gadolinium; although the extent of enhancement varies, it is often a prominent feature (5,39,45,46,52,53,55) (Figs 2, 10). Additional MR imaging findings of diffuse intraarticular PVNS reported by Hughes et al (6) include bone erosion or subchondral cyst (62% of cases), septations (67%), edema in the adjacent bone or soft tissue (23%), and articular cartilaginous defects (31%) (Figs 2, 6, 10, 11). We agree with Hughes et al (6) that erosions about the knee are seen with increased frequency at MR imaging (60% of cases) as compared with radiography (26%–32%), a finding that reflects the cross-sectional imaging capabilities of MR imaging. Subchondral cysts likely arise from cartilaginous defects (created by the underlying synovial process), with subsequent fluid extension (Fig 10). PVNS tissue may also extend about the margins of these subchondral cysts (Fig 10). MR imaging is the optimal modality to depict and define the intraarticular and, particularly, extraarticular disease extent in order to guide complete surgical excision (56) (Figs 2, 6, 11).

Localized extraarticular PVNS, particularly when it involves the fingers (eg, PVNTS or GCTTS), is less commonly evaluated with cross-sectional imaging because the diagnosis is often suggested clinically. When it is performed, MR imaging typically demonstrates a well-circumscribed, soft-tissue mass adjacent to the involved tendon (50,57–59). In our experience, small extensions about the tendon margin are seen in most cases and suggest a tenosynovial origin (Figs 14, 15). This finding is similar to that seen

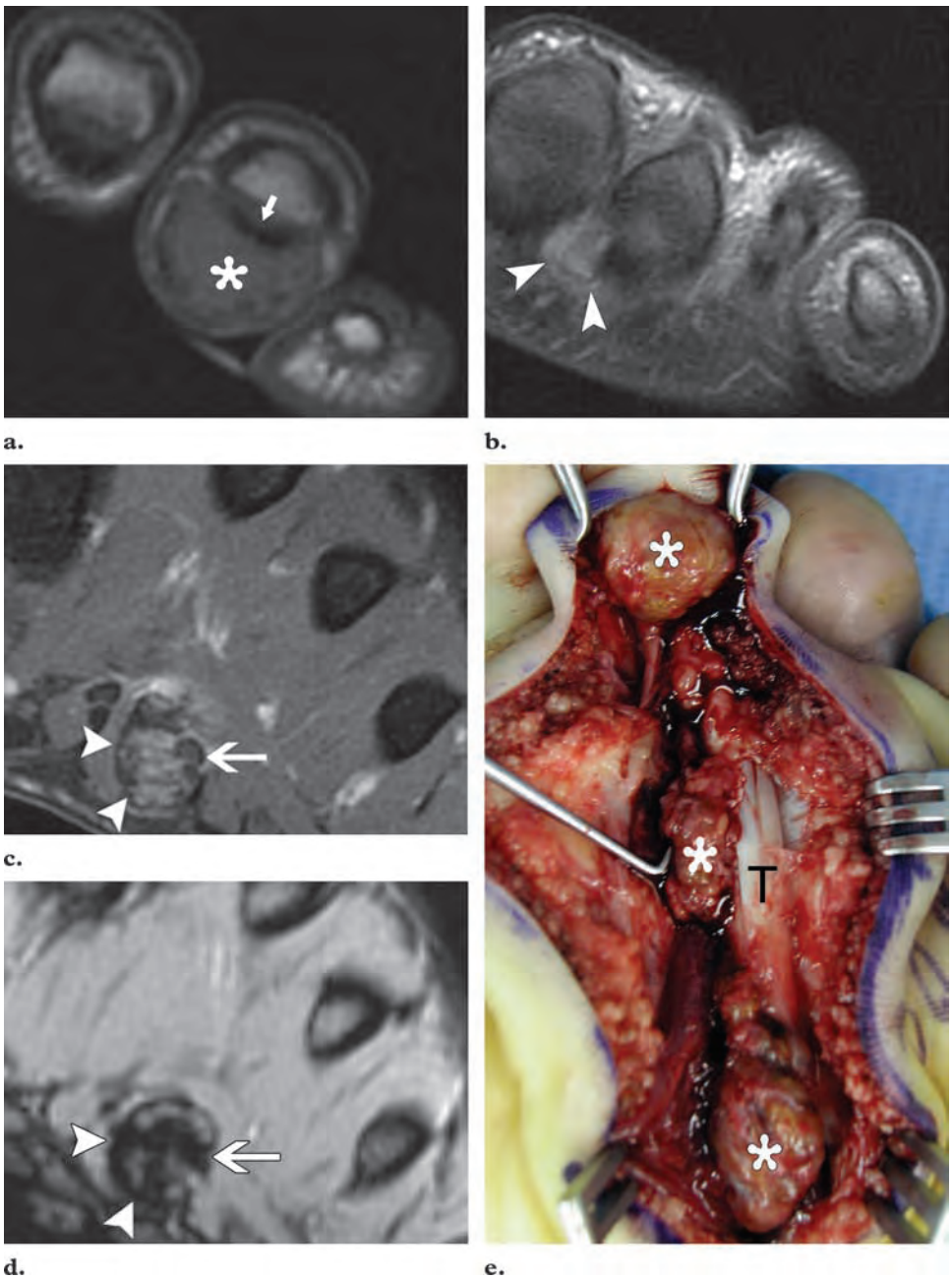


Figure 15. Recurrent localized extraarticular PVNTS of the third toe in a 19-year-old man with multiple satellite nodules. **(a)** Short-axis T1-weighted (400/14) MR image, obtained at the level of the proximal phalanx, shows a recurrent intermediate-signal-intensity (*) soft-tissue mass that surrounds the flexor tendon (arrow). The lesion appears similar to the original PVNTS resected 4 years previously (not shown). **(b)** Short-axis T1-weighted (767/11) fat-suppressed postcontrast MR image, obtained at the level of the metatarsophalangeal joint of the third toe, shows a second satellite nodule (arrowheads) with similar signal-intensity characteristics and diffuse enhancement but without continuity to the distal lesion. **(c, d)** Short-axis T1-weighted (767/11) fat-suppressed postcontrast **(c)** and gradient-echo (600/15, 20° flip angle) **(d)** MR images, obtained at the level of the distal metatarsal of the third toe, reveal a third lesion (arrowheads) with similar intrinsic characteristics that encases the flexor tendon (arrow). On the gradient-echo image **(d),** the lesion has lower signal intensity, resulting from hemosiderin deposition, and mild diffuse enhancement. **(e)** Intraoperative photograph demonstrates the multiple discontinuous foci (*) of PVNTS with brownish discoloration from hemosiderin deposition and the involved tendon *(T)* (the fourth toe is at the top of the image).

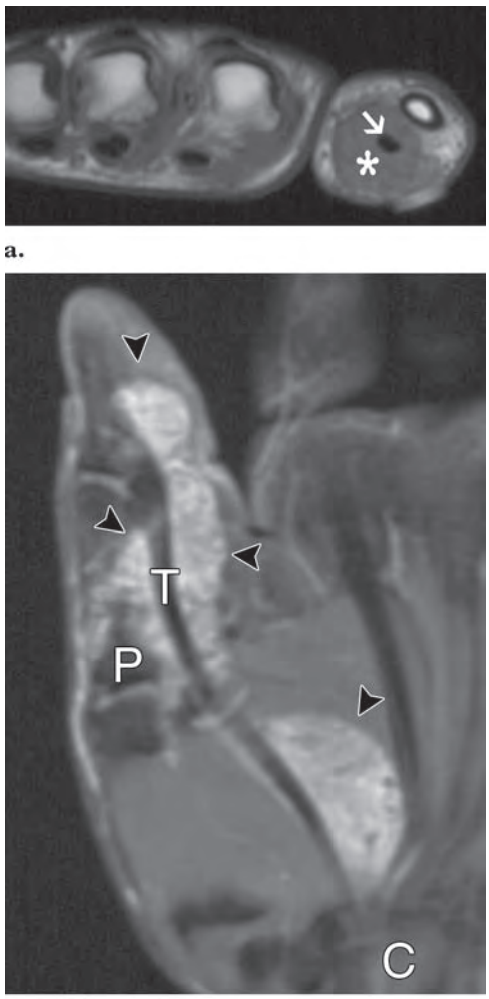


Figure 16. Rare example of extraarticular PVNTS diffusely involving the entire tendon sheath in a 24-year-old woman with a slowly enlarging mass in her thumb. **(a)** Axial T1-weighted (450/13) MR image shows an intermediate-signal-intensity soft-tissue mass (*) that encircles the flexor pollicis tendon (arrow). **(b)** Coronal T1-weighted (400/13) fat-suppressed postcontrast MR image reveals diffuse enhancement of the extensive tenosynovial involvement (arrowheads), which extends circumferentially about the tendon *(T)* from its attachment at the base of the distal phalanx *(P)* of the thumb to the carpal tunnel *(C)*.

at sonography (49). These extensions may entirely encase the tendon (Fig 16). Localized satellite nodules adjacent to the primary lesion may be seen but are uncommon (Fig 15). In rare cases, the tendon sheath may be involved more diffusely in a longitudinal extent (Fig 16).

Localized extraarticular disease frequently demonstrates low to intermediate signal intensity on both T1- and T2-weighted MR images. In our experience, the MR imaging effects of hemosiderin vary more in the localized forms of PVNS (both intraarticular and extraarticular) (Figs 5, 14), a pattern that reflects the pathologic characteristics of these lesions. However, the bursal form of extraarticular disease, PVNB, is more similar to diffuse intraarticular PVNS, because it demonstrates prominent MR imaging effects of hemosiderin (Fig 8). In cases with more limited hemosiderin deposition, signal intensity may be higher than expected (compared with the appearance of diffuse intraarticular PVNS) on long repetition time or water-sensitive MR images (Figs 12, 14). Similarly, the appearances of such lesions on gradient-echo images are more variable (Fig 15), and they may not reveal a prominent blooming effect. Diffuse contrast enhancement of varying degree is seen in most cases of localized disease, either PVNB or PVNTS (50,57–59) (Figs 12, 14, 15).

Localized intraarticular PVNS almost exclusively involves the knee (Fig 5). In a series of 21 cases, Huang and colleagues (24) reported that 67% of the lesions were infrapatellar in location, 24% suprapatellar, and 10% posterior intercondylar. The mean lesion diameter was 2.7 cm (24). The lesions appeared similar to other forms of PVNS, manifesting as low- to intermediate-signal-intensity soft-tissue masses with well-defined nodular or lobular margins on T1-weighted MR images (Fig 5). Heterogeneous and variable signal intensity was seen on T2-weighted images. In our experience and in all the illustrated cases of Huang and co-workers (24), T2-weighted signal intensity was low to intermediate and much lower than that of adjacent joint effusion (present in 38% of cases). Focal circular areas of low signal intensity, corresponding to hemosiderin deposition, within a soft-tissue mass were seen in 76% of cases on T2-weighted images or gradient-echo images (Fig 5), although these areas were much less extensive than those seen in diffuse intraarticular disease (24). Linear or cleftlike areas of high signal intensity were seen within the synovial soft-tissue mass in 33% of cases (24) (Fig 8). Although the authors suggested that this finding likely represents necrosis, we believe that necrosis is unlikely because it is not a histologic feature of PVNS (4,5,12,24). In our opinion, the cleftlike areas of high signal intensity may be related to small focal areas of joint fluid entrapped and surrounded by the hemosiderin-laden soft-tissue mass. Moderate contrast enhancement of the localized intraarticular form of PVNS was seen in 48% of cases reported by Huang et al (24) (Fig 5). As is true for other types of PVNS, MR imaging is vital to detect the anatomic location and exclude diffuse synovial involvement to guide optimal treatment.

Radioisotopes Used in Radiosynoviorthesis for PVNS

Isotope	Maximal Synovial Penetration (mm)	Half-life	Dose (mCi)	Injectate
Yttrium 90	11.0	64 hr	15–25	Colloid
Dysprosium 165	5.7	2.3 hr	300	Macroaggregate
Phosphorus 32	8	14.3 d	6	Colloid

Treatment and Prognosis

Treatment of PVNS is required to prevent progressive loss of function and destruction of the involved joint (in diffuse intraarticular disease) or the tendon or bursa (in localized extraarticular disease) (60). Treatment options include surgical resection, radiation therapy, pharmaceutical modulation of the disease, or a combination of these approaches (25,61,62). Surgical excision is the preferred method of treatment (41,60,61) for all forms of PVNS. However, the long-term success of surgery for PVNS, as with other tumorous lesions, depends on the ability to resect the disease completely. Logically, cure is more likely in the setting of localized disease (including PVNB or PVNTS and both intraarticular and extraarticular disease), and surgical resection alone is typically the only treatment employed for this form of PVNS (63). Diffuse intraarticular involvement is more difficult to eradicate with surgical resection alone, and adjunct therapies may be used. The surgical approach for diffuse intraarticular PVNS depends on the joint involved, extent of disease, and experience and preference of the surgeon.

The goals of surgery for patients with diffuse intraarticular PVNS are to restore function of the joint and to prevent the destruction of articular cartilage by completely resecting the diseased synovium (64). The majority of literature that discusses the surgical management of diffuse intraarticular PVNS describes the approach to the knee (63). Synovectomy may be performed with either an arthroscopic or open arthrotomy technique, but, regardless of the approach, complete resection of disease is required to reduce the likelihood of recurrence.

Arthroscopic resection of diseased tissue is preferred by some investigators (65,66). Complete resection of PVNS is more likely with this approach when the disease is relatively localized and can be accessed easily with an arthroscope through an anterior portal and when the arthroscopic surgeon is experienced (65,66) (Fig 7). Although arthroscopic surgery offers the ability to resect disease with minimal loss of function and faster rehabilitation times, these advantages must be balanced against the possibility that diseased tissue may not be resected completely with this approach (65,66).

Open arthrotomy with synovectomy increases the likelihood of complete resection of disease (60) but usually requires immobilization and a longer recovery. In a large-capacity joint such as the knee, where both anterior and posterior portals are required to excise the entirety of disease, open surgical techniques and multiple approaches increase the risk of neurovascular injury and prolong the recovery period because of the incisions that are required to expose the joint completely. Immobilization is particularly necessary after surgery to prevent breakdown of incisions. Open synovectomy may be augmented by applying a cryosurgical surface spray to all nonarticular surfaces at the time of surgery; however, the efficacy of this intervention has yet to be proved in a randomized study (67).

Radiation therapy may be used as the primary treatment for diffuse intraarticular PVNS, but it is best used to augment surgery following the incomplete resection of disease. Radiation can be administered either by external beam or through intraarticular injection of radioactive isotopes, also known as radiosynoviorthesis (68–70). Some authors prefer the clinical results achieved with arthroscopic partial synovectomy followed by irradiation, compared with those of open radical resection of disease, because of reduced morbidity (68–70).

External beam irradiation has been used to treat diffuse intraarticular PVNS for decades, with excellent clinical results (68,70). Clinical control of disease has been reported in as many as 98% of patients following external radiation therapy. External beam irradiation is usually employed after the incomplete resection of disease. Radiation doses ranging from 20 Gy to 50 Gy are administered in 15–25 fractions, beginning about 6–8 weeks following surgery (68,70). Minimal side effects of erythema have been reported, but the patients usually tolerate the therapy well without skin breakdown. Theoretical concerns include development of malignancy, either in the synovium or bone, following external radiation therapy. Layfield and co-workers (71) reported two cases of malignant PVNS in patients who

underwent radiation therapy for previous benign synovial involvement. However, to the best of our knowledge, no other cases of patients treated for PVNS have been reported in the literature.

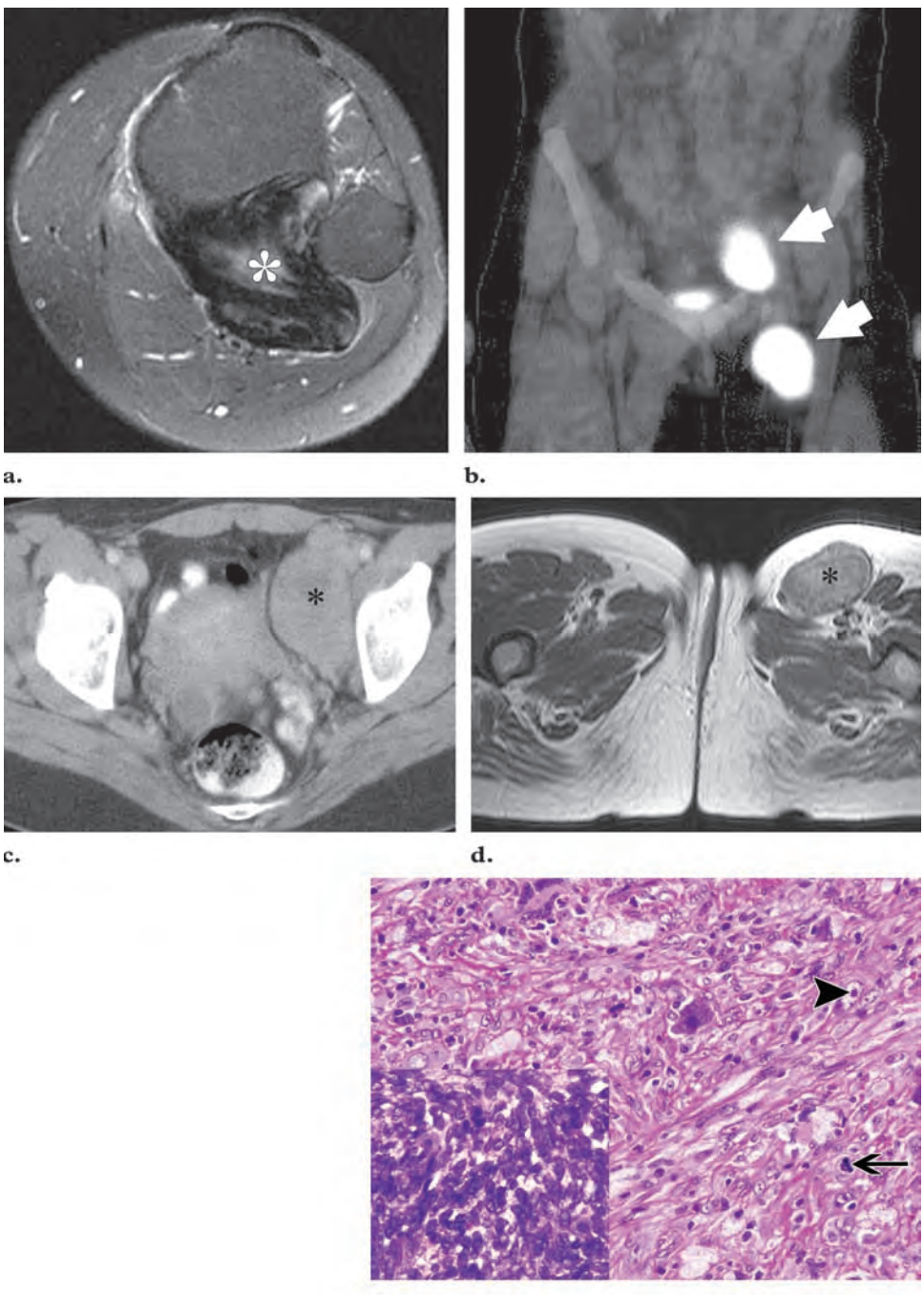
Radiation may also be administered to the joint through an intraarticular injection of a radioactive isotope. Several isotopes have been used, but all share similar properties of being a β-emitting colloid that is bound to a bulky molecule to minimize resorption and leakage (60,61,69,72) (Table). The injection is performed as an outpatient procedure. Arthrography is used to confirm the intraarticular placement of the needle before the injection, and steroid can be added to the injectate to reduce inflammation that may be induced by the therapy (60). Despite the theoretic concern that radiosynoviorthesis may not be able to control disease such as extraarticular extension or bulky postoperative residual disease, the clinical results are frequently excellent. Intraarticular radiosynovectomy has an excellent safety profile, particularly when it is used in large joints. However, an occurrence of skin ulceration that developed after the administration of yttrium 90 in ankle joints and that required muscle flap surgical reconstruction prompted one group of investigators to advise against use of this technique in small joints (73).

The treatment options for diffuse intraarticular PVNS have been limited to surgery and irradiation until recently. Kroot and co-workers (74) reported the use of α-TNF (tumor necrosis factor) blockade in a patient with PVNS that was refractory to conventional therapy. After the administration of infliximab, the patient experienced gradual improvement in clinical symptoms and reduction of pain. In this patient, the total volume of synovitis as assessed by MR imaging was unchanged, but the population of macrophages in the synovium was markedly decreased at pathologic examination. The authors concluded that the reduction of inflammation induced by α-TNF blockade reduced pain, and they postulated that osseous and articular destruction may be prevented or delayed in such patients as a result (74). A future controlled randomized study will be required to understand the true impact of this new class of drugs on the longitudinal behavior of PVNS.

The recurrence rate for localized disease is generally lower than that for diffuse intraarticular PVNS. Most authors have reported 100% cure rate with total resection of localized intraarticular disease, regardless of whether the operative technique is open or arthroscopic (4–6,52,63). The recurrence rate following surgical resection of localized extraarticular disease (ie, PVNTS or PVNB) ranges from 0% to 44% and was 8% in the largest series reported by Ushijima and colleagues (19). The interval between initial treatment and recurrence ranged from 2 to 63 months, with a mean of 24 months in the study of Ushijima et al (19). Recurrence of localized disease may be related to incomplete resection of satellite nodules near the main tumor that were not initially recognized (Fig 15). MR imaging has an important role to play in the detection of such satellite nodules and in the direction of complete surgical excision (Fig 15).

The recurrence rate of diffuse intraarticular PVNS following initial treatment ranges from 8% to 56%, although the actual rate may be higher if MR imaging were used as the detection modality (5,8,25,46). Reported factors that increase the likelihood of recurrence include disease location (PVNS recurs more frequently in the knee, owing to the large capacity of the joint and the difficulty in performing a true total synovectomy), history of previous operations, and positive surgical margins (5,8,25,46). Recurrent disease usually occurs intraarticularly, but subcutaneous tissues may also be involved in arthroscopic portal tracts or in open synovectomy incisions (75). Schwartz and co-workers (25) reported that 7% of recurrences developed at 1 year, 15% at 5 years, and 35% at 25 years after initial treatment. The mean time to recurrence was 5 years (25). Variations in recurrence for diffuse intraarticular PVNS have been reported with different treatment options (62). Chin and colleagues (60) reported no difference in recurrence rate among patients who underwent arthroscopic versus open complete synovectomy. However, not unexpectedly, Ogilvie-Harris (76) and De Ponti et al (65) both reported higher recurrence rates for incomplete synovectomy (56% and 50%, respectively) compared with those for complete resection (7% and 20%, respectively). Blanco and colleagues (77) reported a recurrence rate of 14% for patients who underwent external beam irradiation after partial synovectomy, a rate that was comparable to the recurrence rate for patients who underwent total synovectomy for diffuse intraarticular PVNS. Shabat and co-workers (61) reported similar excellent outcomes with adjunct radiosynovectomy. However, results of surgery and adjuvant therapy are not uniformly positive. de Visser et al (78) reported that use of radiosynovectomy alone or in combination with surgery, compared with use of surgery alone, did not improve patient outcome. Recurrent diffuse intraarticular PVNS is often treated with combined therapy, joint replacement, or amputation (the latter is used in rare cases, typically only those with multiple recurrences) (25,46) (Fig 6).

Figure 17. Malignant transformation of longstanding diffuse intraarticular PVNS of the knee in a 35-year-old woman with multiple recurrences following synovectomy for the initial lesion 20 years before. **(a)** Axial T2-weighted (3800/77) fat-suppressed MR image shows predominantly low signal intensity in a recurrent primary lesion of the knee (*). **(b)** Coronal fused PET/CT image reveals two hypermetabolic foci in the iliac and inguinal regions (arrows). **(c)** Axial CT scan of the pelvis demonstrates iliac adenopathy (*) that corresponds to one of the foci in **b** and represents metastatic disease. **(d)** Axial T2-weighted (3530/69) MR image shows the intermediate-signal-intensity inguinal adenopathy (*). **(e)** Photomicrograph (original magnification, ×200; H-E stain) shows marked cytologic atypia, large prominent nucleoli (arrowhead), paucity of multinucleated giant cells, and mitotic activity (arrow). Inset photomicrograph (original magnification, ×400; H-E stain) from a different area of the lesion reveals marked increased cellularity with spindling and high nuclear-to-cytoplasmic ratio. Necrosis was seen in other regions (not shown).



Malignant PVNS

Malignant transformation of PVNS is rare, and it can occur de novo or be associated with recurrent disease (usually multiple episodes) (79,80). The prevalence of malignant transformation of PVNS was 3% in a study by Bertoni and colleagues (79). These lesions are high-grade aggressive sarcomatous tumors with evidence of synovial origin and typically poor outcome (33,36,71,81). Carstens and Howell (82) first reported malignant transformation of PVNS in 1979, when they described a patient with a lesion in the dorsum of the foot who experienced repeated local recurrences over many years and who ultimately developed multiple metastases. They referred to the lesion in this case as a malignant GCTTS (82). There is controversy in the literature about the diagnosis of malignant transformation of PVNS. Malignant PVNS is composed of a rare collection of tumors with some disagreement over which lesions should actually be included in the diagnosis. The lesions presently thought of as malignant PVNS frequently demonstrate multiple local recurrences as well as lymph node or pulmonary metastases (Fig 17). The designation of malignant transformation is further evidence of the neoplastic origin of this group of lesions.

A minority of proponents suggest that diffuse forms of PVNS with locally aggressive behavior should be considered malignant on clinical grounds, regardless of their histologic characteristics. However, the majority of researchers reserve the moniker malignancy for lesions with sarcomatous histologic features and do not use it for lesions with only aggressive clinical behavior (33,36,71,79,81). Several malignant synovial lesions with varied histopathologic features were previously lumped in with malignant PVNS. Some prior authors considered any sarcoma that contained giant cells and that originated adjacent to a tendon sheath to be malignant PVNS. With this very broad definition, lesions of other histopathologic characteristics such as fibrosarcoma, malignant fibrous histiocytoma, clear cell sarcoma (also known as malignant melanoma of soft parts), and epithelioid sarcoma could be included.

Enzinger and Weiss (12) have defined malignant GCTTS or PVNS as being *(a)* a lesion in which a phenotypically benign, localized or diffuse PVNS coexists with overtly malignant areas or *(b)* a malignant tumor that develops in a joint, bursa, or tendon sheath previously afflicted by PVNS. This definition was modified by Bertoni and co-workers (79) to include malignant-appearing synovial lesions with the following histologic characteristics: *(a)* a nodular, solid infiltrative growth pattern; *(b)* large, plump, round or oval cells; *(c)* cells with large nuclei with deep eosinophilic cytoplasm and prominent nucleoli; *(d)* fewer numbers of benign giant cells, xanthomatous cells, and inflammatory cells (compared the numbers present in PVNS); *(e)* lack of the normal zonal pattern of maturation of PVNS; and *(f)* areas of necrosis. Criteria for histologically malignant PVNS have also been developed at the Armed Forces Institute of Pathology (AFIP); according to these guidelines, malignant PVNS must have at least five of the following eight features: diffuse pleomorphism, prominent nucleoli, high nuclear to cytoplasmic ratio, mitotic activity greater than 10 per 10 high-power fields, necrosis, discohesion of tumor cells, paucity of giant cells, and a diffuse growth pattern (83) (Fig 17).

In a series of eight cases of malignant PVNS, Bertoni and colleagues (79) reported a slight female predominance, an age range of 12–79 years, and a peak incidence in the 6th decade of life. The most common location of malignant PVNS, as determined in a literature review of 15 cases by Bhadra and co-workers (84), was the knee (47% of cases). The foot was the second most frequently affected site (20% of cases), followed by the ankle (13%), hip (7%), and thigh (7%) (84). The largest review of malignant PVNS was performed by Fanburg-Smith and Miettinen (83) at the AFIP. In this series of 27 cases, the mean patient age was 48 years and lesions most commonly affected the knee, followed by the hand or toes (83). Imaging of malignant PVNS has only rarely been reported, although in our experience extensive bone marrow invasion should be viewed with suspicion for malignant disease (85). At radiologic evaluation, lymph node or pulmonary metastases may also be seen (85) (Fig 17).

Despite aggressive therapy, including surgery, adjuvant chemotherapy, and external beam irradiation, malignant GCTTS or PVNS is associated with a guarded prognosis. Local recurrence is reported in 54%–70% of cases, and metastases (to lymph nodes or lung) have been observed in 38%–70%, with death occurring in approximately 50% of cases (79).

Summary

PVNS represents an uncommon benign neoplastic process. When the disease diffusely or focally involves the synovium of a joint, it is referred to as PVNS; when it occurs in the extraarticular synovium of a bursa or a tendon sheath, it is designated as PVNB or PVNTS, respectively. The hypertrophic synovium is typically villous, nodular, or villonodular and 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.

Intraarticular PVNS most frequently affects the knee, with the hip being the second most common site. The radiologic appearances of intraarticular PVNS, particularly its MR imaging findings, are frequently pathognomonic. Radiography shows nonspecific features, such as joint effusion, maintained joint space, and extrinsic erosion on both sides of the articulation (particularly in small-capacity joints such as the hip); calcification is not present. CT reveals the nonspecific synovial thickening and optimally depicts the bone erosion. MR imaging demonstrates the disease extent (particularly bursal involvement) to best advantage, and the predominant low signal intensity of the lesions on T2-weighted images is characteristic of the disease. However, the blooming of low signal intensity, caused by the magnetic susceptibility artifact from hemosiderin deposition in these lesions as seen on gradient-echo images, is nearly pathognomonic of this disease. The knees, followed by the hips, are the most common locations for PVNS or PVNB, whereas PVNTS (frequently referred to as GCTTS) occurs most often in the hands and feet. PVNTS is the most common form of this disease by an approximate 3:1 ratio. Radiographs of the extraarticular localized forms of the disease may appear normal or may reveal a nonspecific soft-tissue mass. Sonography and MR imaging are optimal for demonstrating the intimate relationship of extraarticular lesions to the tendon sheath, a finding that suggests the diagnosis. MR imaging findings of predominant low signal intensity (seen with T2-weighting) and particularly significant hemosiderin content—although the amount is more variable than that seen in intraarticular disease—(seen with gradient-echo sequences) are characteristic of this diagnosis.

The treatment of choice for PVNS of all types is surgical resection, with complete synovectomy being particularly important in cases of diffuse intraarticular disease. Recurrence is more frequent with diffuse intraarticular PVNS, and adjuvant radiation therapy may also be employed for treatment in these cases. Malignant PVNS is rare and difficult to distinguish, both pathologically and radiologically, from multiple local recurrences of benign disease unless there is metastatic involvement of the lungs or lymph nodes. Understanding and recognizing the spectrum of radiologic appearances and their pathologic bases allow improved patient assessment and are important to optimize clinical management.

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RadioGraphics

Pigmented Villonodular Synovitis with Radiologic-Pathologic Correlation

Mark D. Murphey et al

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Page 1495
The diffuse intraarticular form of PVNS most frequently affects the large joints, with the knee involved in 66%--80% of cases (25,26).

Page 1497
Correlation of the histologic features with the imaging findings of a diffuse synovial process is essential to arrive at the correct diagnosis of a benign lesion (Fig 2).

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However, more commonly, radiographs demonstrate joint effusion, soft-tissue swelling, absence of calcification, extrinsic erosion of bone, preservation of joint space, and normal bone mineralization (38,39,43--45) (Fig 6).

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Low signal intensity also predominates on T2-weighted MR images, owing to the preferential shortening of T2 relaxation time caused by hemosiderin, an effect that is accentuated at higher field strength (Figs 7, 11, 13).

Page 1508
This effect is particularly pronounced on gradient-echo images, which demonstrate an enlargement of the low-signal-intensity areas (“blooming”) that is caused by magnetic susceptibility artifact (Figs 6, 10). The blooming effect, which specifically signifies the presence of hemosiderin as the cause of low signal intensity, is nearly pathognomonic of PVNS at MR imaging.