

Pigmented villonodular synovitis of proximal tibiofibular joint: Rare site of involvement treated with medical management

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Dear Editor,

We present a case of pigmented villonodular synovitis (PVNS) demonstrating an extensive disease of leg originating from isolated involvement of proximal tibiofibular joint. Earlier considered to be reactive in origin, recent studies have demonstrated possibility of neoplastic etiology instead.^[1-3] As per our knowledge, only four cases of PVNS showing isolated involvement of the proximal tibiofibular joint have been reported in English literature till date.^[4-7]

A 59-year-old gentleman presented with a mildly painful, slow growing swelling of the right upper calf for 6 years. Clinical examination revealed a fixed, nontender, firm palpable mass in the upper outer calf, extending up to the popliteal fossa. The examination of knee joint was normal with no restriction of range of motion. A provisional clinical diagnosis of soft tissue sarcoma was made. Patient underwent radiographical and magnetic resonance imaging (MRI), followed by biopsy of the lesion for histopathological confirmation.

Plain radiograph of the leg revealed a diffuse soft tissue swelling of the calf with suspicious involvement of the fibular head and adjacent part of tibia [Figure 1]. The knee joint appeared normal. MRI revealed a lobulated mass interposed between proximal tibia and fibula, extending across the interosseous membrane into both the anterior and posterior compartments of leg with encasement of otherwise patent neurovascular bundle. Cranially, it extended into the proximal tibiofibular joint and was associated with erosions of articular surfaces on both aspects suggestive of intraarticular origin of the mass [Figure 2a]. The mass showed hypointense signal to muscle on both T1- and T2-weighted images [Figure 2b]. Gradient echo images [Figure 2c and d] revealed magnetic susceptibility artifacts with marked signal loss (“blooming”) indicating extensive hemosiderin deposition, a feature highly suggestive of PVNS.



Figure 1: Plain radiograph of the right leg and knee, anteroposterior and lateral views. Proximal tibiofibular joint shows erosion in juxta articular area in both tibia and fibula

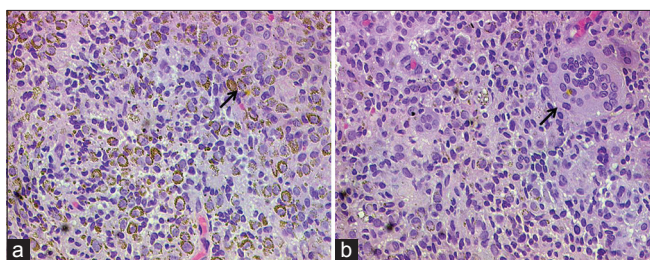


Figure 3: Photomicrograph of the lesion (H and E, x40) shows sheets of dyscohesive round to oval cells with extensive intracytoplasmic hemosiderin pigment (arrow) (a) and osteoclastic type of multinucleate giant cells (arrow) within the tumor (b)

Histopathology confirmed the radiological diagnosis of PVNS showing conspicuous intracytoplasmic hemosiderin pigment [Figure 3a], obscuring the cellular morphology in some of the cells. Sparse mitotic activity and a few osteoclastic types of multinucleate giant cells along with the absence of pleomorphism were observed [Figure 3b]. On immunohistochemistry, tumor cells were positive for antibody to CD68 while negative for antibody to S-100 and HMB-45.

The patient was advised both surgical (extended open synovectomy of the proximal tibiofibular joint with excision of the mass) and medical options. In view of extensive disease and associated risk of neurovascular injury, he deferred the surgery. The patient was treated conservatively with tyrosine kinase inhibitor, imatinib, 400 mg/day for 24 months. There are no adverse effects observed during the treatment course. Imatinib was stopped at 24 months due to observation of a stable disease on repeated MRI scans and relief of symptoms. The patient is under observation and is on regular follow-up since past 36 months. The patient is better symptomatically, and disease appears to be stable [Figure 4].

Usually, a monoarticular disease, it most frequently affects knee joint followed by hip, ankle, shoulder, and elbow.^[1-4,8] Radiographs may be normal or demonstrate a nonspecific joint effusion or soft tissue masses, which may appear dense due to high hemosiderin deposition. Preservation of bone mineralization and joint spaces until late in the disease is characteristic.^[1,4]

MRI is the modality of choice for making a specific diagnosis of PVNS.^[1-3] Diffuse PVNS demonstrates a typical lobulated mass-like synovial proliferation, appearing hypointense to isointense on T1- and T2-weighted images^[1,2] and showing characteristic magnetic susceptibility artifact (“blooming”) on gradient echo images due to local magnetic field inhomogeneities created by hemosiderin.^[1-3]

The condition is optimally managed with open or arthroscopic synovectomy, which may be partial or extended depending on the growth pattern.^[1,9,10] Localized form has excellent prognosis

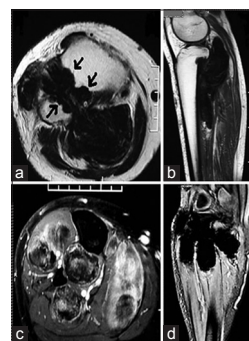


Figure 2: (a) Axial T2-weighted image of proximal leg reveals a lobulated heterogeneous soft tissue mass of predominantly hypo intense signal intensity centered at proximal tibiofibular joint associated with articular erosions (arrows) and extension into muscles of both anterior and posterior compartments. (b) Sagittal T1-weighted image shows hypo intense mass in proximal leg with preserved knee joint. (c) Axial postgadolinium fat-suppressed T1-weighted image shows intense, heterogeneous predominantly peripheral enhancement of the mass. The mass encases anterior and posterior interosseous neurovascular bundles. (d) Coronal gradient echo image shows characteristic “blooming” phenomenon

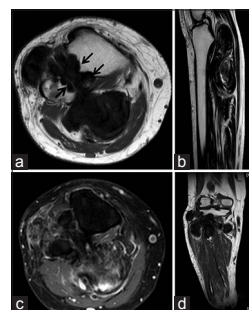


Figure 4: Post 36 months follow-up magnetic resonance images showing mild dimensional disease reduction. (a) Axial T2-weighted image of proximal leg. (b) Sagittal T1-weighted image shows hypo intense mass in proximal leg with preserved knee joint. (c) Axial postgadolinium fat suppressed T1-weighted image. (d) Coronal gradient echo image

while diffuse form tends to recur in 30–50% cases, especially in joints such as knee and shoulder with multiple recesses and communicating bursae.^[1,10,11] Extended synovectomy is recommended in all cases of diffuse PVNS.^[1,9-11]

Recently, few studies have shown that imatinib induces tumor regression in patients with unresectable or advanced PVNS.^[12] Imatinib is a tyrosine kinase inhibitor which blocks macrophage colony stimulating factor receptors, which is expressed in high levels in most of mononuclear and multinucleated stromal cells and are thought to be responsible for tumor formation.^[13] The use of imatinib in this indication is based on the observation of the presence of a t(1;2) translocation in bulk of the tumors, leading to the over expression of active colony-stimulating factor 1, which attracts inflammatory cells that dominate in PVNS.^[12,14] No prospective studies on use of imatinib in PVNS are available as of now.^[15]

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Conflicts of interest

There are no conflicts of interest.

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