Pigmented Villonodular Synovitis as a Differential of Subchondral Cysts

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Pigmented villonodular synovitis (PVNS) is a rare disease which commonly involves the knee joint. It can be seen in association with bursa, tendon sheath or intra-articular; it could be localized or diffused form. Subchondral cysts, giant cell tumor, and geode degenerative cysts as a differential of chronic erosive form could be diagnosed by plain radiography.

A twenty-year-old female presented with one year history of right knee pain and mild swelling. Plain radiography and CT revealed an initial diagnosis of benign-looking lytic subchondral lesions and subchondral cysts were suspected. MRI raised the possibility of PVNS. Arthroscopic resection was performed. The diagnosis was confirmed as PVNS by histopathological examination. The patient is currently in good condition. A multi-disciplinary team decided to proceed with radio-synovectomy to prevent a recurrence.

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Pigmented villonodular synovitis (PVNS) is a rare disease[1]. It results in a villous, nodular, or villonodular proliferation and hyperplasia of the synovium or tendon sheath, with intra and extra-cellular hemosiderin pigmentation deposition and multinucleated giant cells[2,3,4,5].

The term PVNS should be used for intra-articular pathology with pigmented villonodular bursitis for lesions localized to bursas, and the term pigmented villonodular tenosynovitis for those arising from the tendon sheath (known as giant cell tumors of the tendon sheath)[2,3]. It is seen in patients 20-50 years of age[3]. It commonly occurs as a monoarticular joint disease; however, rare cases have been reported with polyarticular pathology with bilateral synchronous or metachronous involvement[4,5]. The intra-articular entity can be of localized or diffused forms[1].

The etiology remains is not clear. Repeated trauma, reactions to unknown stimuli, chronic inflammatory process and benign neoplasm were suggested[6,7,8].

Symptoms are usually non-specific, most patients presenting with knee swelling, pain and locked knee[1,5,6,8,10,11]. MRI reveals the characteristic appearance of blooming artifact related to hemosiderin particle deposition[12,13].

The treatment is usually arthroscopic resection and/or open surgery. The local recurrence rate is 18 to 46%[8,18,19].

The aim of this report is to present a case of PVNS which was treated by arthroscopic resection and radio-synovectomy to prevent a recurrence.

THE CASE

A twenty-year-old female presented with one year history of right knee pain; the pain is worsened with exertion and prolonged standing. No other joint involvement. On examination, there was swelling of the right knee. She had no history of trauma, diabetes mellitus, hypertension or a family history of arthritis.

Vitamin D level was low and was prescribed a regular dose of D 50,000 units once weekly for 3 months. The immunology profile was negative. The patient continued to complain of knee pain with no improvement.

Right knee plain X-ray revealed a well-defined proximal meta-epiphyseal lytic tibial lesion with a sclerotic thin border with no evident septation or trabeculation. A narrow zone of transition was noted. No periosteal reaction or soft tissue mass. A lateral radiograph of the right knee showed posterior joint soft tissue possibly due to synovial thickening/effusion. Suspicion was raised for bone cyst, see figure 1 (A and B).

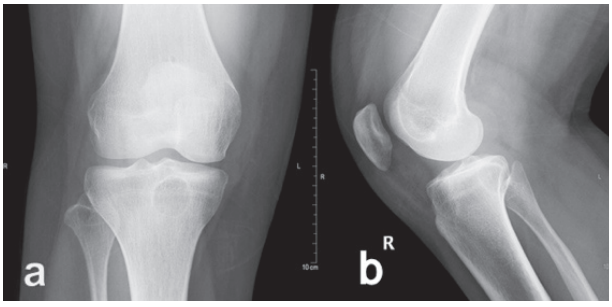

Figure 1 (A and B): Antero-Posterior (A) and Lateral Radiographs (B) of the Right Knee Demonstrate A Lytic Lesion within the Meta-Epiphyseal Junction of the Proximal Tibial End with Well Sclerotic Outline and Narrow Zone of Transition Suggesting Benign Lesion. Figure 1 B Shows Posterior Joint Soft Tissue like Synovial Thickening

CT scan of the right knee joint revealed a large well-defined subchondral cyst seen in the posterior aspect of the medial tibial plateau, measuring about 2.1x1.9x1.7 cm, minimal bone

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expansion, thinning and scalloping of posterior cortical bone with focal defects reaching the articular surface, see figure 2 (A and B). Two smaller subchondral cysts were seen in the anterior aspect of the medial tibial plateau and inner aspect of the lateral femoral condyle, measuring about 1x0.9x0.7 cm and 0.5x0.5cm, respectively. Both had similar CT features with a focal bony defect of related articular surface, see figure 3 (A-D). Moderate knee joint effusion was reported with clear soft tissues around the knee joint.

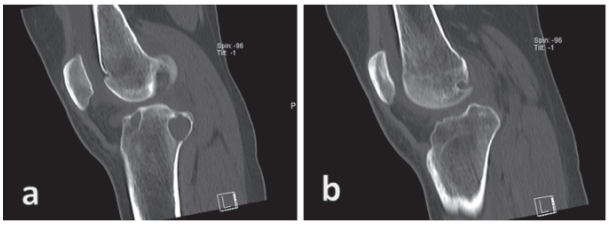

Figure 2 (A and B): Coronal Non-Contrast CT Scan of Right Knee: (A) A Large Well-Defined Subchondral Cyst Seen in the Posterior Aspect of Medial Tibial Plateau Showing Minimal Bone Expansion, Thinning and Scalloping of Posterior Cortical Bone. It Has Narrow Zone of Transition. No Associated Periosseus Soft Tissue Component Could Be Detected. Another Smaller one seen close to Tibial Spine Anterior Aspect (B) Shows Much Smaller Similar Cyst close to Femoral Condyle

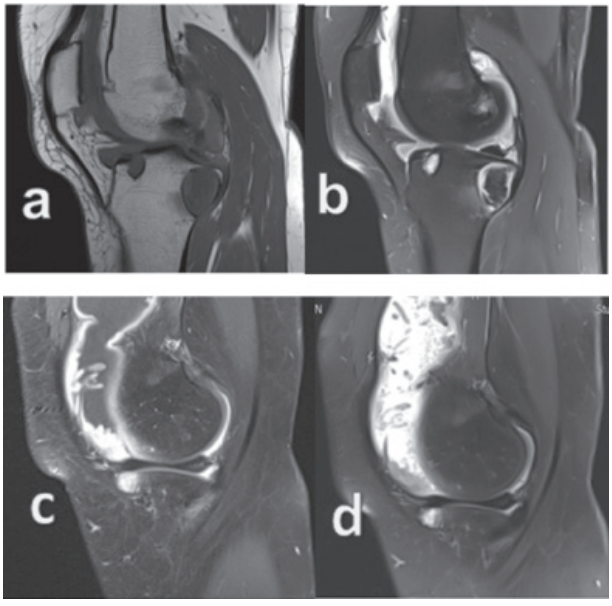

Figure 3 (A-D): Sagittal T1-Weighted Spin-Echo (500/12) (A), Sagittal T2-Weighted Spin-Echo (4030/59) (B), Sagittal Post Contrast T1 Weighted (480/12) (C) and, Sagittal GRE Sequence (D). (A) Low T1WIs Subchondral Lesions with Signal Void Sclerotic Rim (B) Joint Effusion associated with Subchondral High T2WIs SI Cystic-Like Lesions (C) Multiple Enhancing Frond-Like Synovial Proliferation Noted within the Supra-Patellar Bursa with Enhancing Synovial Outline (D) Signal Loss along The Surface of the Synovium (Blooming) Represent Hemosiderin-Laden Synovial Tissue

MRI revealed large effusion and synovial proliferation. The synovium demonstrated low signal intensity on both T1-weighted (500/12 - repetition time msec/echo time msec) and T2-weighted (4030/59) images, see figures 3A and 3B. The synovium also demonstrated magnetic susceptibility artifact with low signal intensity ("blooming") on gradient-echo (GRE), see figure 3D. Contrast images revealed polypoidal-like enhancement of the synovial outline, see figure 3C. The signs were suggestive of primary synovial lesion, the primary differential diagnosis was of PVNS and was advised diagnostic biopsy.

The patient underwent endoscopic subtotal synovectomy. A multi-disciplinary team decided to proceed with radiosynovectomy to prevent recurrence.

DISCUSSION

The etiology remains unclear, however, repeated hemarthrosis, repeated trauma, reactions to unknown stimuli, chronic inflammatory processes and benign neoplasms have been implicated. Other possibilities include benign neoplastic processes and reactions[6,7,8].

The mechanism of bone erosions seen in patients with PVNS is not clear. The pressure of synovial overgrowth within the involved joints versus a release of a substance that causes bone erosion by the synovium has been implicated[9].

Patients with the diffuse form of the joint disease usually present with progressive pain, joint swelling, repeated non-traumatic joint effusion and decreased range of motion with joint locking[1,5,6,8,10,11]. Patients with localized forms are difficult to diagnose as symptoms may mimic meniscal lesions[7].

The differential diagnoses, on plain films, with multiple subchondral cysts include gout, synovial chondromatosis, tuberculosis (TB) and hemophilia[7]. MRI is essential for establishing the diagnosis and planning the treatment[7]. MRI typically shows lobulated margin, a mass-like synovial proliferation, diffuse form or well-defined single nodule. Joint effusion is frequently seen[12,13].

PVNS is a benign lesion, but malignant degeneration has been reported rarely[13]. Mononuclear cells in association with multinucleated giant cells are usually reported on microscopy[1,14,15,16,17].

Synovectomy is the recommended treatment of choice for PVNS[8]. Radiotherapy is used in cases of multiple recurrences and as a prophylactic measure to prevent a recurrence, as in our case[8,18,19]. MRI is used to monitor the recurrence[8,19]. Local recurrence is 12% - 48%[8].

CONCLUSION

Pigmented villonodular synovitis is a rare disease commonly involving the knee joint. Synovectomy is the recommended treatment of choice for such cases; radiotherapy is reserved for multiple recurrence cases.

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