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Case History
A 10 year old male presented after a direct fall onto his left knee while playing basketball one week ago. He was unable to continue playing. Two days later he developed intermittent pain “all over the knee” that worsened with weight-bearing. His mother noted swelling and limping. He treated his pain with Acetaminophen and ice.

Past medical history was remarkable for strabismus surgery. Family history revealed mother had insulin dependent diabetes mellitus; maternal grandmother had history of ITP. He denied prior joint pain or swelling, rash, fever, change of weight, easy bruising/bleeding, and gastrointestinal symptoms.

Physical exam:
Vital signs: heart rate 66 bpm, blood pressure 90/52 mmHg, weight 108 pounds. He was a well appearing Caucasian male, in no acute distress.

Knee Exam:
Inspection: A 2+ effusion was present in the left knee.

Palpation: A tender area of ecchymoses was noted over the tibial tubercle and lateral femoral condyle.

Range of Motion: He had full active knee extension bilaterally. Left knee lacked 5 degrees of flexion compared to right. Full flexion was painful.

Strength: 4/5 strength with left leg knee extension and straight leg raise.

Stability testing: poor endpoint on Lachman testing of the left knee, however the patient was guarding. Anterior and posterior drawer, as well as valgus and varus stress tests were negative bilaterally.

Special test: McMurray’s was negative for click bilaterally.

Hips / Pelvis exam:
Range of Motion: full pain free ROM of bilateral hips.

Inspection / Palpation: Non-tender pelvis, surrounding hip musculature.

Extremity exam: No other joint swelling present

He had normal gait.
Neurovascular and skin exam were unremarkable.

Laboratory evaluation: Blood work obtained by his primary care physician included a negative ANA, Rheumatoid factor of 3, and a normal CBC.

Imaging: AP, Lateral, and Merchant views of the left knee revealed an effusion, and normal appearing physes.

He was instructed on range of motion exercises and strengthening. Reassessment was scheduled for one week. The patient did not return until 3 months later. Mom states she did not follow-up after the initial episode of pain and swelling because it resolved spontaneously within 48 hours of the visit.

Three days prior to the second evaluation, patient had played a dancing videogame on a dance mat for 1.5 hours in socks. He squatted down to put his shoes on when his left knee locked in flexion. He was unable to extend his knee, though he reported no significant pain. He was taken to an Emergency Department where he was sedated and the left knee was passively extended. Radiographs were performed; he was placed in a knee immobilizer, and given crutches. He subsequently developed diffuse left knee pain and swelling.

Two days later his repeat physical exam included heart rate 114 bpm, blood pressure 130/80 mmHg, and a temperature of 98.3°F. He was anxious in appearance, and lay supine on the exam table, hesitant to move the left lower extremity. Active range of motion was very limited, with flexion extension between 5° and 15° comfortably. A 3+ boggy effusion was present, extending from distal metaphysis of the femur to the proximal tibial plateau. He was diffusely tender throughout the left knee. Left sided straight leg raise strength was 2/5. Ligamentous testing and McMurray’s could not be performed secondary to patient pain and restricted motion. He was unable to bear weight on the extremity. Neurovascular exam was unremarkable.

AP and Lateral radiographs of the left knee from the Emergency Department were performed status post extension maneuver, and were only notable for a large suprapatellar joint effusion.

An MRI of the left knee showed a very large joint effusion with intra-articular deposits of low signal intensity on all sequences, suggestive of hemosiderin deposition, likely due to repeated intra-articular hemorrhage. Widened intracondylar notch and slight irregularity of femoral condylar articular surfaces was present. A partial thickness tear of the vastus lateralis at the level of the distal femoral metaphysis was present. No fracture was present. No meniscal or ligamentous injury was visualized. The hemosiderin deposition was concerning for a diagnosis of hemophilia. The patient was brought back in for evaluation by a Hematologist who found normal coagulation studies. The patient was offered a therapeutic aspiration of the effusion. Sero-sanguinous synovial fluid, 45cc, was removed with a glucose level of 127 mg/dl and total protein 5 g/dl. Gram stain revealed 3+ granulocytes and no organisms, culture was negative.
Patient had improvement in left knee motion, strength, and pain with physical therapy. Patient subsequently underwent diagnostic arthroscopy which revealed gross hemosiderin-laden soft tissue throughout patellofemoral joint, medial compartment, and lateral compartment. The synovium was covered with abnormal stained finger-like projections. Frozen section was sent intra-operatively which showed villonodular processes with hemosiderin, foam cells, and densely packed synovioocytes with focal giant cells, consistent with a final diagnosis of pigmented villonodular synovitis. A total compartment synovectomy was performed. Patient subsequently completed physical therapy and obtained full pain free motion and strength of the left knee. He has remained active with no episodes of recurrence in the last 1.5 years.

**Discussion**

Pigmented villonodular synovitis (PVNS) is a rare disease involving over proliferation of the joint synovium. PVNS was initially described in 1852 by Chassaignac. The incidence is estimated at 1 in 1.8 million. It is a benign fibrohistiocytic disorder that may occur in a joint, a tendon sheath, or a bursa. The etiology is unknown, although attempts have been made to determine if it is inflammatory or neoplastic in origin. PVNS can occur in any age group, although the third and fourth decades are the most common.

Two types exist: a diffuse form that affects the entire synovium, and a localized form that appears as a discrete mass. They are histologically identical, but are considered separate entities because of their divergent clinical course. Our patient suffered from the diffuse form of PVNS, which will be the focus of this discussion. The diffuse form of PVNS can affect any synovial joint, but most commonly affects the large joints, especially the knee (80% of cases), hip, and shoulder. Monoarticular disease is the typical presentation, although rare cases of polyarticular have been reported. Although PVNS is benign it is a locally aggressive lesion that can cause significant joint and soft tissue destruction.

Clinical presentation is characterized by insidious onset of pain which can be progressive in nature. Recurrent episodes of swelling and localized warmth are also common symptoms. Synovial hypertrophy may cause limited range of motion and significant mechanical symptoms such as locking or catching. The non-specific symptoms, insidious onset of PVNS, and rarity of the disease may lead to a delay in diagnosis.

Plain radiographs may be normal or reveal findings of soft tissue densities consistent with synovial hypertrophy or joint effusion. Bony erosion and cystic changes may be evident in advanced disease. Currently, MRI is the preferred imaging modality. Hyperplastic synovium may be evident, and a heterogeneous signal intensity is present in all imaging sequences. T2-weighted images produce the characteristic low signal density due to hemosiderin deposition. Although MRI is highly suggestive of the diagnosis, definitive diagnosis of PVNS is made through tissue biopsy. Arthroscopic evaluation frequently reveals a hemosiderin stained synovium with multiple hypertrophic villi and nodular projections.
Treatment of diffuse PVNS may include: surgical synovectomy, radiation therapy, arthrodesis, bone grafting, or arthroplasty. Total synovectomy is the primary treatment of choice, however there is no standardized treatment currently. Patient age, joint involved, the chronicity of the disease and the recurrence rate may all affect treatment choices. Due to the difficulty in removing the entire synovium, recurrence rates are high, between 10 and 56%. Unfortunately the most commonly involved joint, the knee, also has the highest recurrence rate. Bone grafting and arthroplasty are typically reserved for cases with extensive joint destruction with arthritis. Adjuvant radiation can also be used. Typically it is reserved for diffuse forms of recurrent PVNS. Unfortunately, radiation therapy is not without its own set of risks.

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i Chassaignac M. Cancer de la gaine des tendons. Gaz Hop Civ Milit. 1852;57:185-6


