Intra-articular Nodular Fasciitis of the Knee in a Pediatric Patient

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abstract

The differential diagnosis for an intra-articular lesion in the knee of a pediatric patient is broad. Diagnostic considerations include pigmented villonodular synovitis (PVNS)—the most common intra-articular tumor—and a variety of both benign and malignant tumors, including lipomas, hemangiopericytomas, nodular fasciitis, parosteal osteosarcomas, and fibromyxoid sarcomas. If there is concern over possible malignant lesions, a tumor surgeon should be consulted. Precise pathologic diagnosis is ideal for identifying these enigmatic lesions and for determining the appropriate treatment plan. This article presents the case of a 13-year-old boy who presented with 1-month duration of knee pain and no history of trauma to the extremity. Physical examination revealed pain along the medial and lateral joint lines, pain with range of motion, and limited range of motion of the affected knee. Magnetic resonance imaging revealed a 3×1×3-cm lesion in the posterolateral corner that was believed to be localized PVNS. Arthroscopically, there was no evidence of PVNS, but a posterolateral soft tissue mass was found and removed, which was pathologically diagnosed as a rare, benign, intra-articular nodular fasciitis. When working with intra-articular masses, it is important to assess the likelihood of malignancy and to both consult a tumor surgeon and use the appropriate surgical tumor principles when malignancy is a concern. Additionally, the pathology team should be consulted prior to surgery and be on standby during arthroscopic evaluation of the knee to help with precise diagnosis of the intra-articular mass. Discussing the case with the pathologist with imaging studies present is helpful and often aids in the diagnosis of the lesion.

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There are a variety of intra-articular knee lesions in the pediatric population. Mechanical symptoms of the knee (snapping, locking, popping, giving way, localized tenderness, and pain) are most commonly associated with meniscal tears, chondral injuries, and loose bodies; however, in rare cases, intra-articular tumors, benign or malignant, can occur, with localized pigmented villonodular synovitis (PVNS) being the most common of these lesions.\textsuperscript{1} Localized PVNS has been known to mimic other intra-articular lesions, such as meniscal pathology or loose bodies.

The use of arthroscopy in the pediatric population is increasing, with rates rising by 62\% between 1996 and 2006, with most of this increase being attributed to knee and shoulder arthroscopy.\textsuperscript{2} A recent retrospective review found that approximately 0.8\% of individuals seeking arthroscopic surgery for mechanical knee symptoms had benign intra-articular masses.

This article presents the case of a young boy who presented with mechanical knee symptoms and was believed to have PVNS based on magnetic resonance imaging (MRI). Intraoperatively, there were no signs of PVNS, and pathologic analysis revealed a rare intra-articular mass.

**Case Report**

A 13-year-old boy presented with left knee pain of 1 month’s duration, which had prevented him from playing baseball, and no history of trauma to the extremity. He reported anterior knee pain that worsened with knee extension to a straight leg position, swelling, and decreased range of motion. He reported no rashes, tick bites, mechanical symptoms, fever, paresthesias, warmth, open wounds, or febrile illness. No joints other than the left knee were symptomatic, and there was no family history of juvenile rheumatoid arthritis. Prior to presentation, laboratory tests ruled out Lyme disease. Ibuprofen, rest, and ice had been ineffective in treating the pain.

On physical examination, the patient was afebrile, with an antalgic gait. He had a moderate effusion in the patellar fat pad, which was debrided. There was no gross clinical evidence of PVNS. On arthroscopy, an initial view of the knee through an anterolateral portal revealed a discrete soft tissue mass directly posterior to the lateral femoral condyle extending distally into the popliteal hiatus (Figure 2), inhibiting range of motion of the knee. Magnetic resonance imaging (MRI) was performed to further evaluate possible intra-articular derangements.

Magnetic resonance imaging revealed a 2.8×1.0×3.4-cm lesion posterior to the lateral femoral condyle. The lesion was mildly hyperintense on T1-weighted MRI and mildly hypointense on T2-weighted images (Figure 1), with signal characteristics most consistent with localized PVNS. Once informed consent was obtained, the patient was scheduled for surgical arthroscopy and debridement of the left knee with biopsy.

On arthroscopy, an initial view of the knee through an anterolateral portal revealed moderate synovitis in the patellar fat pad, which was debrided. There was a discrete soft tissue mass directly posterior to the lateral femoral condyle extending distally into the popliteal hiatus (Figure 2), inhibiting range of motion of the knee. No gross clinical evidence of PVNS was noted. Frozen sections from multiple intraoperative biopsies revealed spindle cell proliferation set in myxoid or chondroid matrix. An accessory posterolateral portal was established under direct visualization to perform an extensive synovial debulking of all visibly involved tissue.

The final pathology report revealed a benign spindle cell proliferation with haphazard arrangement and foci of short in-
terlacing fascicles. Foci of myxoid degeneration and extravasation of erythrocytes were noted. Neither hemosiderin pigment nor clusters of foamy macrophages were identified. The lesional cells were positive for vimentin, smooth muscle actin, muscle specific actin, and CD10. No reactivity was seen with CD34, bcl-2, desmin, S-100, and cytokeratin AE1/AE3 (Figure 3). These histological findings and immunohistochemical staining properties were consistent with nodular fasciitis. Fluorescent in situ hybridization studies for fused in sarcoma gene break apart yielded negative results and MUC-4 immunostaining revealed only scattered positive cells, making a diagnosis of fibromyxoid sarcoma unlikely.

**DISCUSSION**

Intra-articular nodular fasciitis is a rare diagnosis, especially in pediatric patients. To the best of the authors’ knowledge, this is only the second report of intra-articular nodular fasciitis occurring in a pediatric patient. The other case occurred in the ankle of a 9-year-old boy.\(^3\)

Nodular fasciitis is a benign soft tissue lesion composed of proliferating fibroblasts. It is the most common neoplasm of fibrous tissue and may grow rapidly with high mitotic activity, mimicking a more aggressive lesion.\(^4\) Nodular fasciitis typically affects patients 20 to 40 years old and may cause mild pain or tenderness in approximately 50% of cases.\(^5\) It most commonly affects the upper extremity (46% of cases), but also affects the head and neck (20%), trunk (18%), and, at times, the lower extremities (16%).\(^5\) Lesions have been reported with a diameter of 0.5 to 10 cm, but most are typically 2 cm or smaller and recurrences are rare, even after partial resection.\(^6\)

After ruling out common intra-articular lesions such as PVNS, the authors relied on the expertise of pathologists to determine the type of intra-articular lesion encountered. Based on the initial frozen section review, the pathologic differential diagnosis included PVNS, parosteal osteosarcoma, fibromyxoid sarcoma, nodular fasciitis, and malignant hemangiopericytoma.

Pigmented villonodular synovitis typically has a characteristic hemosiderin-laden appearance, which was absent in this case, and parosteal sarcoma does not typically have an intra-articular location. Often, reviewing imaging findings with the pathologist can aid in the correct diagnosis of the intra-articular lesion. In PVNS, MRI will show articular effusion and hyperplasia of the synovial membrane. Nodular growths with weak T1 and T2 signal can be seen because of hemosiderin deposits, which are also responsible for the unique microscopic appearance of PVNS. There may be significant inflammatory changes with destructive potential in affected joints with marked synovial hypertrophy.\(^7\)

Parosteal osteosarcoma usually presents on MRI as a dense, lobulated, bony...
mass with a radiolucent line that appears to separate the dense bony mass of the tumor from the bone cortex. Contrast enhanced T1-weighted images are valuable in delineating the extent of the tumor and can aid in guiding the biopsy. Fibromyxoid sarcoma is frequently hypointense or isointense to muscle in fluid-sensitive MRI sequences, often with enhancing gyriform patterns on MRI. Malignant hemangiopericytoma has nonspecific MRI with intermediate T1 and high T2 signal intensity. The most characteristic feature is the presence of serpentine vessels in an otherwise solid nonspecific soft tissue mass. These vessels are usually high flow (low signal on T1 and T2) vascular structures.

On MRI, nodular fasciitis has a signal intensity similar to, or slightly higher than, skeletal muscle. It usually is homogeneous on T1-weighted images with high signal intensity on T2-weighted sequences, greater than subcutaneous fat. At times, however, it may demonstrate intermediate signal intensity. Thus, it is difficult to make a definitive diagnosis on MRI and pathologic review is necessary.

Although the authors acknowledge that classic tumor principles stress the use of longitudinal incision without violation of multiple compartments, in this situation the lesion was intra-articular with exposure to both the proximal tibia and the distal femur. When PVNS was not encountered on diagnostic arthroscopy, an intraoperative decision was made by the senior author (L.W.), given the lower likelihood of malignancy based on advanced imaging, to access and excise the lesion arthroscopically with the use of an additional posterolateral portal. Although the possibility of seeding another portal tract was introduced, in the event of malignancy, the definitive procedure would involve either wide excision with a limb-sparing procedure or an above the knee amputation. Both would address whatever seeding occurred during the less invasive arthroscopic procedure.

**Conclusion**

The differential diagnosis for an intra-articular lesion in the knee of a pediatric patient is broad. If concern exists about possible malignant lesions, a tumor surgeon should be consulted. The pathology team should be consulted prior to surgery and be on standby during arthroscopic evaluation of the knee to help with the diagnosis. Discussing the case with the pathologist with the imaging studies present is helpful, often aiding in diagnosis of the lesion.

**References**