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Synovial Hemangioma of the Knee: A Case Report

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We present a case report of a rare synovial hemangioma of the knee. Magnetic resonance imaging (MRI) demonstrated typical signal characteristics and enhancement. Histologic findings were confirmatory. Imaging characteristics, differential diagnoses and classification are discussed. A review of the relevant literature is provided.

Case report

A 7-year-old female presented with inability to walk and a decreased range of motion of her left knee after a fall from a human-powered scooter. On physical examination, there is note of an approximately 4 x 3 cm suprapatellar soft tissue mass, thought to be related to a suprapatellar effusion. Review of the patient's past medical history reveals a report of a "mass" after blunt trauma that occurred three years prior to this injury. Radiographic imaging was initially thought to be negative but close inspection revealed a vague soft tissue density in the suprapatellar region. The lesion was arthroscopically removed without recurrence to date.

Magnetic resonance imaging (MRI) shows a suprapatellar mass demonstrating heterogenous signal with enhance-

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Abbreviations: MRI, Magnetic resonance imaging, CT, Computed tomography, PVNS, pigmented villonodular synovitis

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ment following intravenously administered gadolinium (Figure 1). No phleboliths were identified and there were no adjacent marrow or cortical abnormalities. Targeted sonography performed with color Doppler augmentation shows a solid and lobulated vascular mass in the suprapatellar knee. The histopathologic evaluation in our case revealed a cavernous hemangioma pattern, with large, thinwalled vessels (Figure 2).

Discussion

Synovial hemangiomas, first described by Bouchut in 1856, are rare benign vascular tumors that occur most frequently around the knee but have also been reported in the elbow, wrist, ankle, temporo-mandibular joint and tendon sheaths (1-3). They can be focal or diffuse in their involvement of the joint. The average age of onset is early adolescence. With minimal trauma, or spontaneously, they can hemorrhage, which often results in clinical presentation before the age of 16 (1-4). Misdiagnosis often contributes to a delay in diagnosis of many years.

The initial clinical presentation of synovial hemangiomas often includes pain, joint swelling and recurrent joint effusions, with or without limitation in range of motion (1-7). They can also present with mechanical symptoms mimicking internal derangement (3). On clinical examination, the mass is often palpable, compressible, and spongy.

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Figure 1A. Sagittal T1-weighted MR image shows a lobulated mass within the suprapatellar recess that is predominantly isointense to muscle with scattered tiny areas of fat.

Figure 1B. Sagittal fat-suppressed T2-weighted MR image shows a heterogenous mass demonstrating areas of both low and high signal intensities. Note the presence of a small amount of joint fluid (arrows).

Figure 1C. Sagittal fat-suppressed T1-weighted MR image following intravenous gadolinium-based contrast administration shows heterogenous enhancement of the lesion and a small amount of non-enhancing joint fluid.

Classification

Soft-tissue hemangiomas can be categorized based on specific site of origin as cutaneous, subcutaneous, intramuscular, synovial or subsynovial (1). Further classification is based on size or type of predominating vessels within the lesion: cavernous (large vessel), capillary, venous and arteriovenous (1-3, 7). The vast majority are cavernous (50%), followed by capillary (25%), arteriovenous (20%) and venous (5%) (1, 3). Another classification system, used primarily by interventional radiologists and orthopedic surgeons, classifies them by anatomical relationship to the joint: juxtaarticular, intra-articular or intermediate type. Juxtaarticular hemangiomas are situated on the outside of the actual joint capsule, with no intra-articular involvement. However, intra-articular lesions are actually situated within the joint capsule itself, and the last type, intermediate, show features of both juxta-articular and intra-articular lesions (2-4). Most reported cases have been of the juxta-articular and intermediate types (3).

Imaging

Radiographic findings of a synovial hemangioma are sparse or nonspecific; often the findings suggest or are similar to a joint effusion (1-3, 5, 6, 8). Although highly suggestive of the diagnosis in the presence of a clinical mass, phleboliths are occasionally seen. When there is prolonged diagnostic delay, degenerative changes resembling hemophilic arthropathy can develop (2, 4, 9).

Computed tomography (CT), if obtained, can confirm the presence of a soft tissue mass, identify phleboliths if present, and delineate any adjacent osseous change related to local mass effect. CT however, is limited in the actual characterization of the soft tissue tumor itself (1, 3, 5, 8).

MRI allows superior contrast resolution and multiplanar capability and is the modality of choice in the imaging evaluation of synovial hemangiomas (or any soft tissue tumor in general). This is true because MRI has the distinct ability to accurately identify the extent of the lesion and its relationship to surrounding tissue and structures (2-4). On T1-weighted images, synovial hemangiomas display low to intermediate signal intensity as compared to surrounding muscle and fat tissue, whereas T2-weighted images appear as high signal intensity (1, 4, 7, 8). Thin, fibrofatty septa are characteristically seen separating the serpentine vascular components. The identification of tiny, rounded signal voids is compatible with the presence of phleboliths, which are not common (1, 5, 7, 8, 10). Fluid-fluid levels are nonspecific but have been reported (1). Gadolinium-enhanced MRI provides clear demarcation of the frequently lobulated borders of the lesion, to include demonstration of any extra-articular involvement (1, 5, 8, 10). The vascular mass can be differentiated from joint fluid or adjacent muscle with the use of intravenously administered gadolinium (Figure 1C).

Differential Diagnosis

The differential diagnosis of synovial lesions of the knee includes; pigmented villonodular synovitis (PVNS), and

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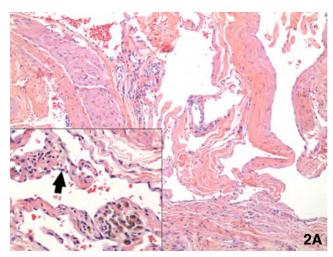


Figure 2A. Photomicrograph of histologic specimen reveals large, thin-walled, erythrocyte-filled vascular spaces lined by bland endothelial cells (arrow) within a dense connective tissue matrix with occasional hemosiderin-laden macrophages. (H&E, X100, inset X200)

synovial chondromatosis or osteochondromatosis (1). PVNS, also hemosiderin containing, characteristically shows low to intermediate (usually not high) T2 signal intensity. Synovial chondromatosis shows cartilage signal intensity, while osteochondromatosis is often diagnosed radiographically with identification of numerous loose bodies and pressure erosions, typically involving both sides of the joint. Rheumatoid arthritis, tuberculous arthritis, lipoma arborescens, and hemophilic arthropathy are additional differential considerations and can often be differentiated from synovial hemangioma clinically. Synovial sarcomas contain dystrophic calcification in about a third of cases. Vast majorities occur next to a joint but are not actually intra-articular in origin, unlike synovial hemangiomas (10).

Pathology

The histopathologic evaluation in our case revealed a cavernous hemangioma pattern, with large, thin-walled vessels, consistent with the most common (50% of cases) type of synovial hemangioma (Figure 2). The most important histologic differential is diffuse-type giant cell tumor (pigmented villonodular synovitis), which is much more likely to recur than synovial hemangioma and may also appear relatively vascular in its early stages. It is distinguished from synovial hemangioma by the presence of sheet-like proliferations of histiocytes and multinucleated giant cells. Synovial hemangiomas contain a matrix that is myxoid, edematous or focally hyalinized in between the vessels.

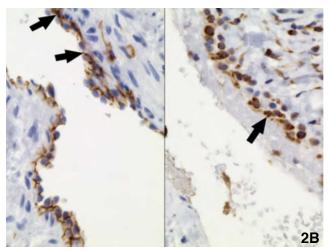


Figure 2B. Photomicrographs show endothelial cells stained positive for CD31 (and CD34) by immunohistochemistry (left, arrows). Synovial lining cells stained with CD68 (KP1) (right, arrow).

Treatment

Once identified, surgery should be initiated rapidly to reduce the long term risk of cartilage deterioration and chronic hemarthropathy (2-4). Treatment generally involves open or arthroscopic surgical excision of the entire hemangioma, with partial or total synovectomy (1, 4, 5). Currently, arthroscopic excision is the modality of choice if the hemangioma is pedunculated and well circumscribed (3, 6). Due to the vascular nature of such lesions, significant bleeding is a risk the surgeon must face, often leading to the decision for arthroscopic removal (3). However, if the lesion is diffuse in nature, open excision is likely the better option (3, 5, 6). Recurrence rates are generally much higher following open surgical excision due to the diffuse nature of the lesion (3, 6).

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