

# Indolent synovial sarcoma in adolescent male: MR, PET, microscopy, and arthroscopy correlation

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## CASE SUMMARY

A 16-year-old Native American male presented with 2 years of gradually worsening left knee pain initially treated with anti-inflammatory medication and physical therapy. Magnetic resonance imaging revealed the presence of an anterior lesion within Hoffa's fat pad. After referral to orthopedic surgery, he underwent arthroscopy with subtotal synovectomy/excision.

## IMAGING FINDINGS

Sagittal T1-, T2- and proton density (PD)-weighted MR images of the left knee (Figure 1, A-C) demonstrates the 2.3cm soft tissue mass (arrowheads) within the infrapatellar space displacing Hoffa's fat pad which shows predominantly T1 hypointensity, and T2/PD heterogeneity with areas of iso, hypo, and hyperintense signal (to muscle). Coronal PD image (Figure 1D) confirms location predominantly within the medial compartment of the left knee with questionable early involvement of the medial femoral condyle (arrow).

PET/CT images (Figure 2 A-B) performed two months following surgical excision reveal residual focal hypermetabolism in the intraarticular space at

the anterior aspect of the left knee joint along the intercondylar notch of the distal left femur. These findings could be seen with either residual tumor or post-operatively. Posterior view PET maximum intensity image (B) shows focal uptake of the left knee with maximum SUV of 2.1. No other foci of hypermetabolism were seen.

Histopathology images (Figure 3) with low (A), medium (B), and high (C) power H&E staining with characteristic biphasic squamous cell and spindle cell patterns. Epithelial membrane antigen immunoperoxidase stain (D) highlights the epithelial component. By RT-PCR this tumor is positive for an SYT-SSX1 fusion transcript.

Arthroscopy images (Figure 4) of the left knee reveal the lobulated mass within the anterior fat pad, having the gross appearance of pigmented villonodular synovitis, according to the operative report.

## DIAGNOSIS

Biphasic grade 2 synovial sarcoma. Differential diagnosis includes: neoplasm (undifferentiated pleomorphic sarcoma, peripheral nerve sheath tumor), pigmented villonodular synovitis, Hoffa's syndrome, or meniscal cyst.

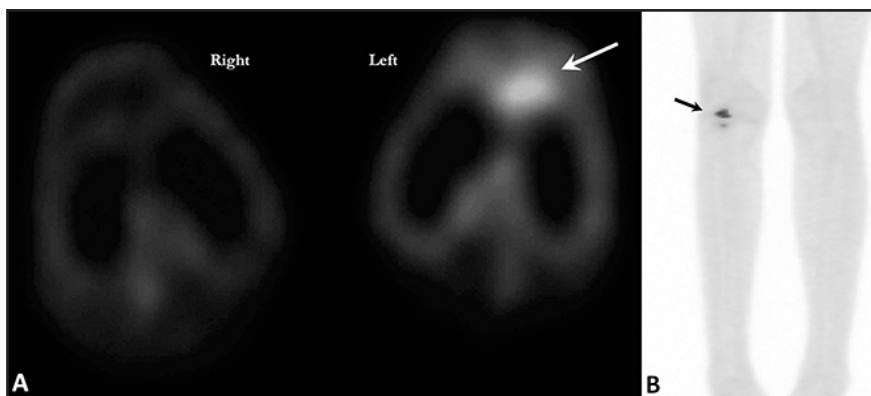
## DISCUSSION

Sarcomas are rare mesenchymal tumors that account for 1% of all malignancies.<sup>1,2</sup> Synovial sarcoma (SS) is a sarcoma subtype consisting of spindle-cell tumors with variable epithelial differentiation that is often associated with SYT-SSX gene fusion.<sup>1-5</sup> SS comprises 2.5%–10.5% of all primary soft-tissue malignancies worldwide<sup>2</sup> and occurs in both the pediatric and adult populations, remaining the commonest non-rhabdomyosarcoma soft tissue sarcoma (NRSTS) in childhood.<sup>3-5</sup> Approximately 30% of patients are younger than 20 years of age.<sup>4</sup>

SS most frequently affects the musculoskeletal system with the extremities involved in over 85% of cases. The lower extremities (~70%) especially the knee and peri-popliteal fossa, tendon sheaths, and bursae are effected in younger adults ( $\leq 40$  years of age).<sup>1-3,6</sup> Contrary to the implied nomenclature, these neoplasms do not arise intra-articularly or within the synovium, rather these mesenchymal cells differentiate just enough so as to resemble synovial spindle cells. An increasing number of reports are describing extra-extremity primary SS.<sup>2,6</sup> The lung is the most common site for metastasis, and



**FIGURE 1.** MR images of the left knee demonstrate the 2.3 cm soft tissue mass (arrowheads) within the infrapatellar space displacing Hoffa's fat pad. (A) Sagittal T1 image with hypointense signal, (B & C) T2 & PD sagittal images show marked heterogeneity with areas of iso, hypo, and hyperintense signal (with respect to muscle). (D) Coronal PD image confirms location predominantly within the medial compartment of the left knee with questionable early involvement of the medial femoral condyle (arrow).



**FIGURE 2.** Axial PET images (A) performed two months following surgical excision reveal residual focal hypermetabolism in the intraarticular space at the anterior aspect of the left knee joint along the intercondylar notch of the distal left femur (white arrow). This could be seen with residual tumor versus post-operative changes. Posterior view PET maximum intensity image (B) shows focal radiotracer uptake in the left knee with maximum SUV of 2.1 (black arrow). No other foci of hypermetabolism were identified.

post-metastatic survival is rare.<sup>4</sup> As an intermediate to high-grade tumor, the risk of local recurrence and distant metastasis is consequential, despite initial aggressive surgical resection.<sup>2</sup> Optimal treatment remains elusive in the absence of randomized clinical trials, owing to its rarity.<sup>3</sup> Recurrent SS is considered chemosensitive and typically treated with rhabdomyosarcoma protocols by pediatric oncologists.<sup>3,4</sup>

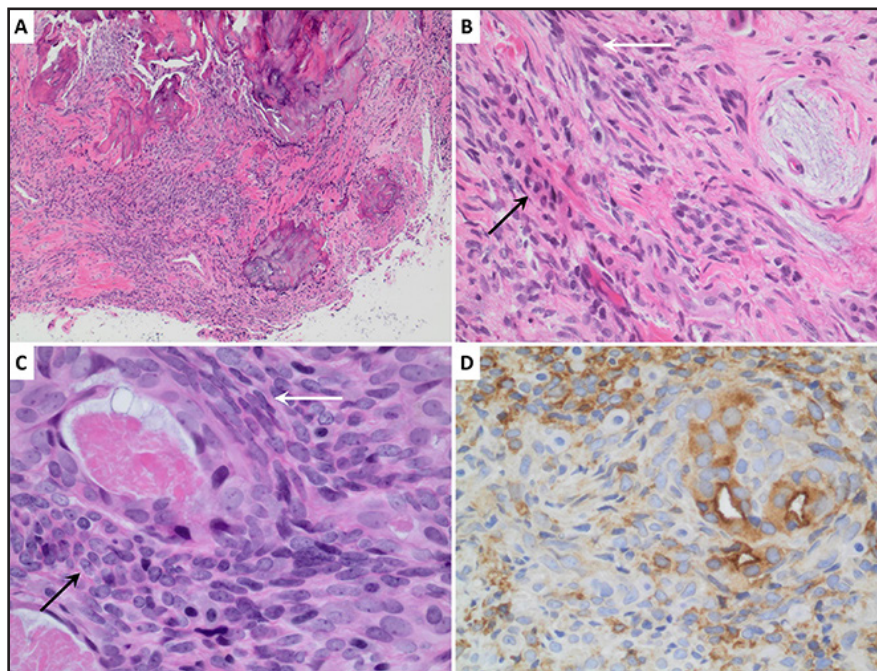
Patients typically present with slow-growing palpable masses (frequently  $\geq 5$ cm). The paradoxically insidious onset often delays the diagnosis up to several years, prolonging the time from initial symptoms to treatment and worsening the stage.

While radiographic features are infrequently diagnostic, common findings include a juxtaarticular soft tissue mass with peripheral calcifications.<sup>7-9</sup> CT findings show a well-circumscribed muscle-density mass with heterogeneous enhancement and peripheral punctate calcification. MRI is most useful demonstrating multi-lobulation and marked heterogeneity with the "triple sign" (areas of hypo, iso, and hyperintensity on T2) seen in upwards of 35% of patients.<sup>7-9</sup> Additional studies report a "bowl of grapes" sign with multiple internal septations and cystic necrosis.<sup>8</sup> PET imaging is particularly helpful for initial staging, post-treatment management with the literature reporting at least 80% F18-FDG sensitivity, and for prognostication utilizing SUVmax.<sup>10</sup>

## CONCLUSION

Synovial sarcomas are rare mesenchymal tumors representing the most common non-rhabdomyosarcoma soft tissue sarcoma of childhood, especially of the lower extremity and knee. These can masquerade as pigmented villonodular synovitis having a similar appearance on arthroscopy and MR imaging. Common MR findings include a hypointense homogenous appearance on T1 with a heterogenous multilobulated





**FIGURE 3.** Histopathology photomicrographs confirm synovial sarcoma. Low-power (A), medium-power (B), and high-power (C) H&E staining demonstrating characteristic biphasic squamous cell (black arrows) and spindle cell (white arrows) patterns. High-power staining with epithelial membrane antigen immunoperoxidase staining confirms dimorphic cell pattern. Additional confirmatory FISH studies for synovial sarcoma (18q11.2 with SS18/SYT) were performed (not shown).

appearance on T2 and varying degrees of internal septations (triple sign, bowl of grapes sign). The insidious onset and propensity for recurrence/metastasis warrants heightened diagnostic suspicion. Histopathology remains a crucial diagnostic component.

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**FIGURE 4.** Left knee arthroscopy photos reveal a lobulated mass within the anterior fat pad (black arrows).

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