UNUSUAL PRESENTATION OF LOW-GRADE SYNOVIAL SARCOMA AS FLEXION CONTRACTURE OF THE KNEE

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Abstract

Synovial sarcoma (SS) is an uncommon type of mesenchymal tumor that commonly involves the juxta-articular region, particularly around large joints like the knee. Although synovial sarcomas are usually high-grade tumors, this case highlights an instance of a low-grade variant with benign clinical features. A boy in late adolescence presented with persistent pain and progressive flexion contracture around the knee for four years. Imaging suggested a benign lesion; however, histopathology confirmed SS. Post-surgery, the patient received radiotherapy due to positive surgical margins. A year later, he exhibited a painless knee movement with a mild flexion deformity, showing no signs of recurrence. This report emphasizes the importance of molecular analysis and careful excision for low-grade SS, given the risk of recurrence despite benign appearances.

INTRODUCTION

Synovial sarcoma (SS) can show varying degrees of epithelial characteristics, either as a single-phase (monophasic) or two-phase (biphasic) structure. It accounts for less than 10% of all soft tissue sarcomas (1). When it comes to localized SS, patients who are younger and have the tumor confined to a specific area tend to have a more favorable prognosis.

Diagnosing SS can be challenging because it lacks distinctive clinical or radiological features. While larger SS tumors often appear more diverse in signal intensity on MRI scans, smaller synovial sarcomas (less than 5 cm) can exhibit well-defined borders and uniform signal intensity across all MRI images, regardless of the imaging technique used. This can make them resemble benign lesions like localized nodular synovitis within joints (2).

Molecular analysis has shown that over 95% of SS cases are characterized by a specific chromosomal translocation known as t(X;18)(p11;q11), which results in a fusion of the SYT-SSX genes (3). This genetic event is considered the primary driver of SS development. Therefore, conducting molecular analysis on biopsy samples should be the preferred method for diagnosing synovial sarcoma. In this report, we present our experience with a SS that clinically mimicked a slow growing benign lesion.

CASE PRESENTATION

A boy in late adolescence presented to our tertiary care center with chief complaints of pain and progressive flexion contracture in his right knee region for the past four years. The pain was initially moderate but became severe over the past few months. The flexion contracture had a gradual onset and increased progressively over the past year. He reported a history of multiple surgeries for the swelling, with temporary relief followed by recurrence. He denied any history of decreased appetite or weight loss, and there were no similar swellings noted elsewhere in his body.

On examination, he appeared to be of average build. There was mild fullness around the lateral aspect of the right distal thigh, which was tender to palpation. Additionally, he exhibited a fixed flexion deformity of 30 degrees in the right knee (Figure 1).

There were no enlarged lymph nodes and no signs of distal neurovascular deficit. A plain radiograph of the right knee revealed an elongated, small calcified mass on the lateral aspect of the right distal femur, without any involvement of the bone or joint (Figure 2).

MRI of the right knee demonstrated a hyperintense lesion on T2/STIR and isointense on T1, located along the anterolateral aspect of the right distal thigh, between the iliotibial band and lateral femoral condyle. The lesion appeared as a well-defined lobulated soft tissue intensity mass with focal calcification (Figure 3). There was no evidence of infiltration into surrounding soft tissues or adjacent bone involvement.

Based on the X-ray and MRI findings, a provisional diagnosis of benign or low-grade malignant swelling around the right knee joint, possibly pigmented villonodular synovitis (PVNS), was made. The prognosis was discussed with the patient. Other differential diagnoses considered included complex meniscal cyst, parameniscal cyst, or ganglion cyst.

Following discussions with family members and obtaining informed consent, the patient underwent planned marginal excision and biopsy of the swelling. Intraoperatively, a myxomatous mass was observed around the lateral aspect of the right lateral femoral condyle, containing calcification and bony spicules in the distal part (Figure 4). The entire abnormal mass, along with surrounding synovium, was excised and sent for histopathological examination.

Histopathological evaluation of the tumor tissue revealed a fragmented mass composed of short spindle cells arranged in interlacing fascicles, stag horn pattern, and sheets. The cells were elongated with scant to moderate cytoplasm, high nucleo-cytoplasmic ratio, ovoid hyperchromatic nuclei, and indistinct nucleoli (Figure 5), suggestive of a synovial sarcoma.

Immunohistochemistry performed on the sample confirmed the diagnosis of synovial sarcoma. Given positive margins for cancer, postoperative radiotherapy was recommended for the tumor bed using intensity-modulated radiotherapy with a radical intent. At the one-year follow-up, the patient showed no signs of local recurrence, with painless knee range of motion, and mild fixed flexion deformity of the right knee (Figure 6).

DISCUSSION

To the best of our knowledge, this is a very rare documented case of a patient who initially presented with what appeared to be a non-malignant primary lesion but was later found to have a positive indication for sarcoma. Such sarcomas in their early stages can be detected using additional molecular testing methods.

Orthopaedic surgeons should be familiar with a wide range of tumor presentations. When there is suspicion of a tumor, it's crucial to refer the patient to a specialized orthopedic oncology center. Additionally, radiologists and orthopaedic surgeons should be aware that most intraarticular synovial sarcomas measuring less than 5 cm exhibit non-specific MRI characteristics, which can lead to misinterpretation(1).

In the treatment of soft tissue sarcomas, surgical wide resection with clear, tumor-free margins is imperative (4). In the case of Synovial sarcoma of knee, achieving these clear margins is only possible through extraarticular resection. Keeping any part of the joint capsule intact carries the risk of sarcoma recurrence (5).

In most cases, synovial sarcomas (SSs) tend to develop around the outer parts of the body, particularly in the extremities (6). They are less frequently found in areas like the neck, heart, and lungs (7). When they do occur in the extremities, they are typically near large joints like the knee joint. These tumors are closely associated with structures such as joint capsules, tendon sheaths, bursa, and facial tissues. It's widely believed that SS doesn't originate from synovial cells but rather from mesenchymal cells (8). In cases where SS becomes calcified, it can be seen in soft tissue on X-ray images (7). Prognostic factors for SS include

factors like incomplete removal of the tumor, a tumor size larger than 5 cm, male gender, age over 20, a high degree of malignancy, the presence of tissue necrosis, invasion of nerves and blood vessels, a high rate of cell division, and a specific genetic variant called SYT-SSX1(9). When SS metastasizes, it typically spreads to nearby lymph nodes and the lungs (10).

On magnetic resonance imaging (MRI), SS appears as a diverse, multi-lobular soft tissue mass with a signal intensity similar to or greater than that of muscle on T1-weighted images. On T2-weighted images, the mass appears heterogeneous with a predominantly high signal intensity (11).

Although the radiological characteristics of SS are not unique, up to 30% of cases may show calcification in the soft tissues (11). It's important to consider this because calcified masses can also be indicative of other conditions, such as myositis ossificans (MO), which involves the abnormal formation of bone within soft tissues and often occurs after injuries (12).

From a histological perspective, SS has three subtypes: biphasic, monophasic, and poorly differentiated (13). In our case, the examination of tissue samples revealed spindle-shaped cells, arranged in interwoven bundles, forming a stag horn-like pattern as well as appearing in groups. The cells have a small to moderate amount of cytoplasm and a high ratio of nucleus to cytoplasm. The nuclei are ovoid, deeply stained, and have unclear nucleoli.

CONCLUSION

In conclusion, synovial sarcoma should be considered a differential diagnosis in cases of benign-appearing periarticular masses or progressive flexion contracture of joints, as early recognition significantly impacts management outcomes. Thorough molecular testing is essential for accurately diagnosing ambiguous soft tissue tumors, ensuring precise treatment planning. Complete surgical excision remains the cornerstone of management, with radiotherapy playing a vital role in cases of positive margins to reduce the risk of recurrence. Given the high recurrence potential associated with synovial sarcoma, regular and vigilant follow-up is crucial to monitor for any signs of disease progression or relapse, ultimately improving patient prognosis.

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FIGURES

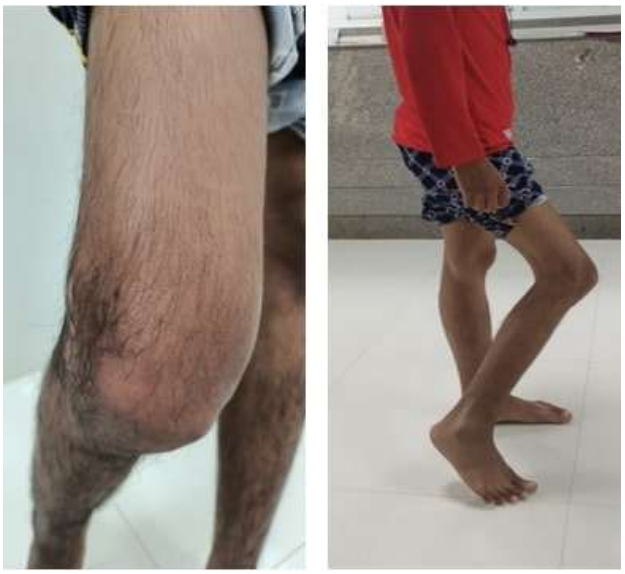

Figure 1: Clinical image showing mild fullness around the lateral aspect of the right distal thigh, along with a fixed flexion deformity of 30 degrees in the right knee.

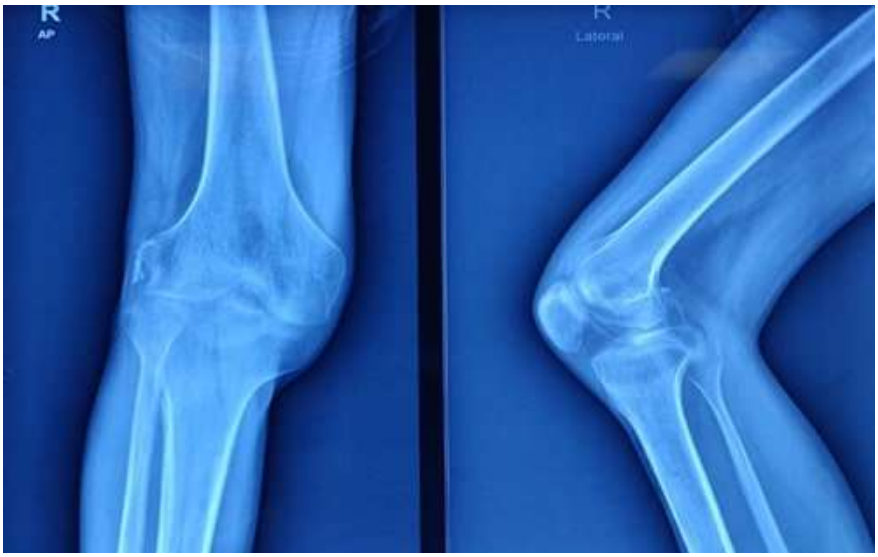

Figure 2: Plain radiograph of the right knee, showing an elongated, small calcified mass located on the lateral aspect of the distal femur without any bone or joint involvement.

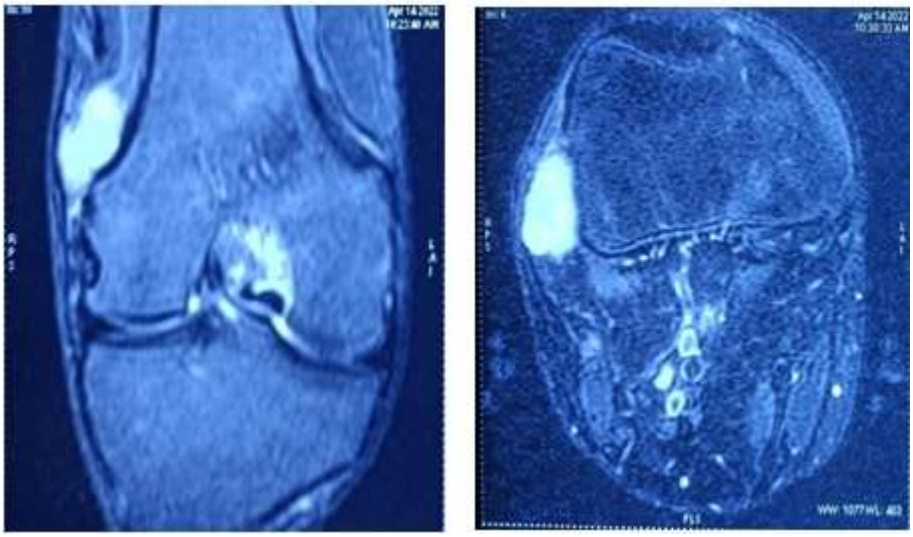

Figure 3: MRI of the right knee, T2/STIR sequence, revealing a well-defined hyperintense lobulated soft tissue mass with focal calcification along the anterolateral aspect of the right distal thigh, positioned between the iliotibial band and lateral femoral condyle.

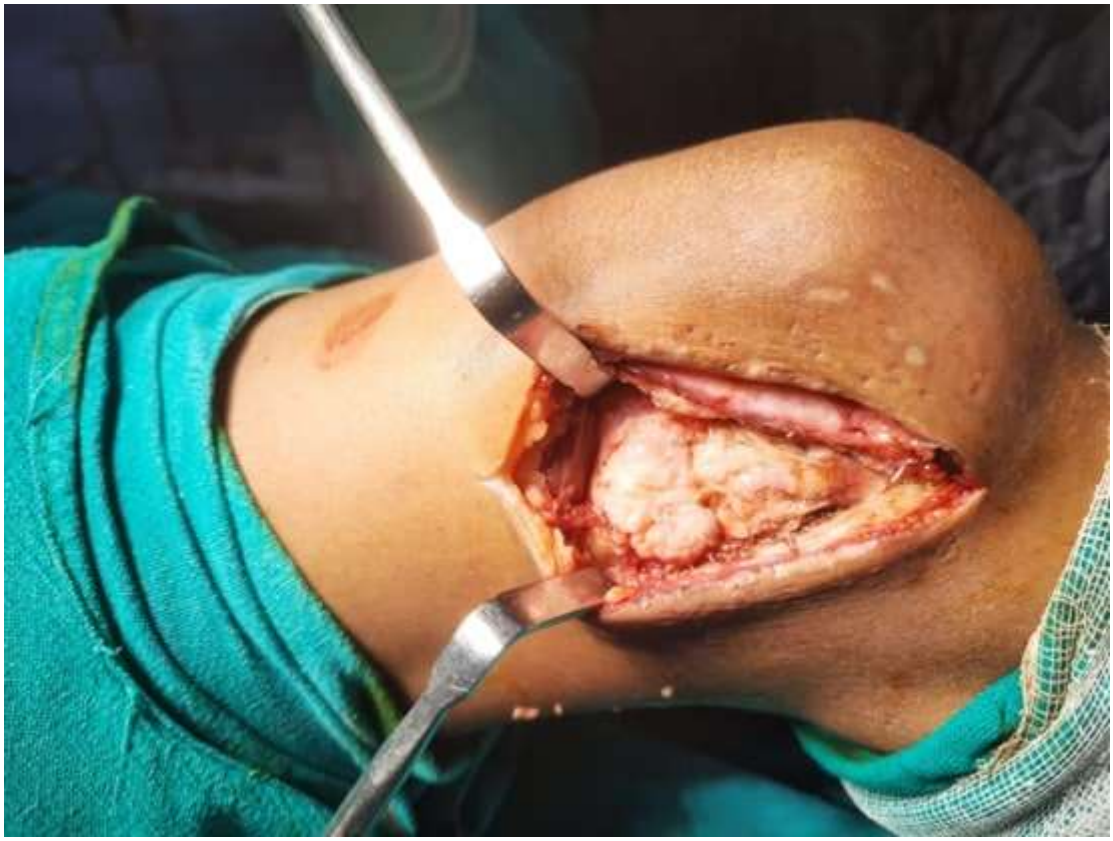

Figure 4: Intraoperative image showing the exposed mass around the lateral aspect of the distal femur after incision, with visible calcification and bony spicules in the distal part of the tumor.

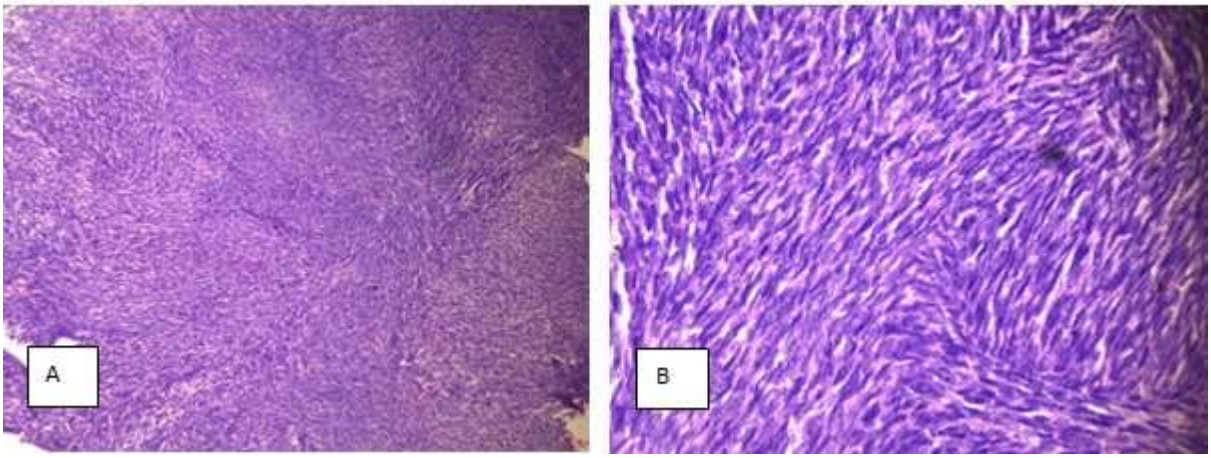

Figure 5: Histopathological slide ((A) Low magnification and (B) High Magnification) displaying short spindle cells arranged in interlacing fascicles, with a high nucleo-cytoplasmic ratio, ovoid hyperchromatic nuclei, and indistinct nucleoli, characteristic of synovial sarcoma.

Figure 6: Postoperative follow-up image at one year showing the right knee with no local recurrence and painless knee range of motion, though with a mild fixed flexion deformity.