Acta Scientific Orthopaedics (ISSN: 2581-8635)

Volume 6 Issue 6 June 2023

Case Report

Intraarticular Synovial Sarcoma of Knee Treated with wide Local Excision and Arthrodesis- A Case Report and Follow Up of 4 Years

Ashish Rustagi[1], Akshay MK[2]* and Bhupender Khatri[2]

[1]*Associate Professor, Department of Orthopaedic, Central Institute of Orthopaedic, VMMC and Safdarjang Hospital, New Delhi, India*

[2]*Senior Resident, Department of Orthopaedic, Central Institute of Orthopaedic, VMMC and Safdarjung Hospital, New Delhi, India*

***Corresponding Author:** Akshay MK, Senior Resident, Department of Orthopaedic, Central Institute of Orthopaedic, VMMC and Safdarjung Hospital, New Delhi, India.

DOI: 10.31080/ASOR.2023.06.0759

Received: April 14, 2023

Published: May 24, 2023

Abstract

Intra-articular Synovial sarcoma is rare and is morphologically distinct as compared to extra articular involvement. The periarticular synovial sarcomas can secondarily spread to the joint capsules. The occurrence of synovial sarcoma originating from an intraarticular location is considered to be rare.Discussion: This case report describes a patient of 34 year female diagnosed with Monophasic Intranodular synovial sarcoma of the left knee. She presented with complains of Pain, swelling, slight discomfort and inability to fully flex left knee which gradually increased over around 2 years. After Staging and biopsy the tumor was managed by wide location excision and arthrodesis using intramedullary nail with augmentation plate and followed up for 4 years for any local or systemic recurrence and also discusses the treatment options based on a review of the prior literature. Conclusion: We conclude that Intra-articular synovial sarcoma are rare and require multidisciplinary approach and interdepartmental correlation for effective management and dedicated orthopedic specialist. Orthopedic surgeons must keep a high suspicion for dealing with lesions of unknown etiology and should have a low threshold for seeking an opinion of musculoskeletal oncologist to assist in concluding appropriate differential diagnosis before intervention. These precautions can prevent complications and improve patient survival rates.

Keywords: Periarticular Synovial Sarcoma; Intra-Articular Synovial Sarcoma; Limb Reconstruction After Resection; Monophasic and Biphasic Synovial Sarcoma; Malignant Neoplasm Management; Synovial Sarcoma Four Year Follow-Up

Introduction

Synovial sarcoma is a morphologically distinct neoplasm which accounts for 5-10% of the soft tissue sarcomas [1-3]. Synovial sarcoma is characteristically found near joints with more than 50% cases found in lower extremities compared to upper extremities involving hip region more than the knee. Intraarticular involvement is rare compared to extra articular involvement[3].The periarticular synovial sarcomas can secondarily spread to the joint capsules [4]. The occurrence of synovial sarcoma originating from an intraarticular location is considered to be rare based on the paucity of documented case reports [5]. This case report describes a case of Monophasic Intranodular synovial sarcoma of the left knee treated by wide location excision and arthrodesis using intramedullary nail with augmentation plate and a follow up of 3 years and discusses the treatment options based on a review of the prior literature.

Case Report

A 34 year old married female presented to our outpatient department with complains of Pain, swelling, slight discomfort and inability to fully flex left knee which gradually increased over around 2 years. She was being treated by local general practitioner on the lines of early osteoarthritis with on and off analgesics and physiotherapy. There is no history of trauma, constitutional symptoms, any other joint involvement, morning stiffness and any other swelling or lump anywhere in the body. Her Knee examination showed nontender, diffuse swelling predominantly extending suprapatellar and infrapatellar region ,moderate effusion with palpable synovial thickening, there is no local rise of temperature, restricted terminal range of movement with fixed flexion deformity of 20 degree with no signs of instability or abnormal mobility without neurovascular deficits.

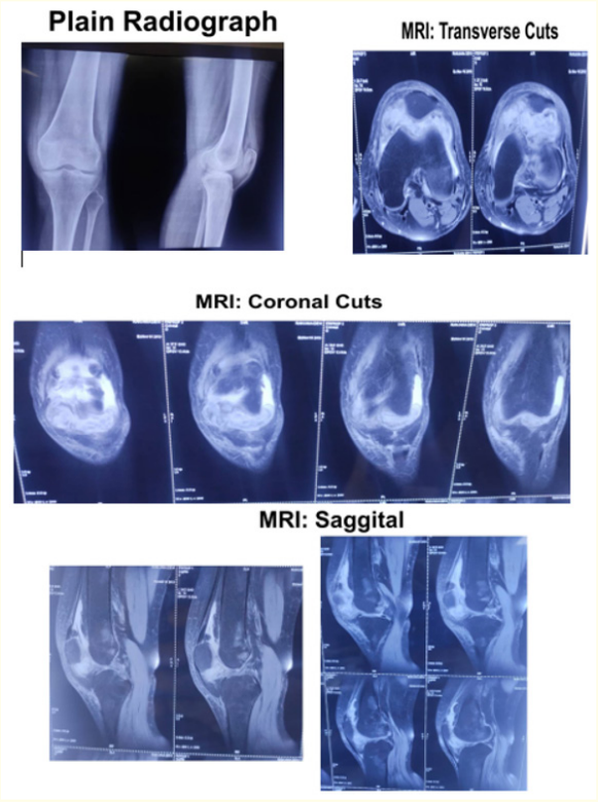

Figure 1

Plain radiographs showed linear and punctuate calcification within the medial and lateral compartment, but otherwise normal. A magnetic resonance imaging of the left knee showed grossly thickened synovium in the Hoffa's fat pad region with presence of T2W Hypo intense foci, it measures 5.7*1.9 cm in size, moderate joint effusion is seen appearing hypo intense on T1W and hyper intense on T2W images. It appeared to have continuity with joint capsule. Anterior and posterior cruciate ligaments appear normal in alignment and signal intensity. On positron emission tomography- computed tomography (PET-CT), the lesion had increased metabolic activity with no metastatic lesion were seen in any other parts of the body. Contrast enhanced computed tomography appeared to be normal. This suggested clinical diagnosis as Non-specific Synovitis left knee with differentials as PVNS and Synovial Sarcoma. The patient underwent ultrasound guided Core needle biopsy of the lesion followed by Histopathological examination which showed solidly packed plump oval or short-spindle cells with a pronounced hemangiopericytomatous pattern and a small portion of pseudoglandular or epithelial arrangement and IHC staining showed TLE1, BCL2 and Cytokeratin positive.

As the lesion size was less than 5 cm with deep involvement and no lymph node and distant metastasis (on PET-CT) the diagnosis was AJCC 8th Edition: Stage II (T1; N0; M0; G2/3) Synovial Sarcoma Left knee. The management involved multidisciplinary approach and included neoadjuvant chemotherapy followed by local resection and adjuvant chemotherapy.

The patient received 6 cycles of Ifosfamide/Adriamycin regime, the swelling decreased following neoadjuvant chemotherapy and it was followed by Extra-articular wide local resection of left knee and Reconstruction with Knee arthrodesis using nail-plate- cement spacer. The surgically resected specimen was sent to pathology for examination grossly mass 17 x 7 x 7 cms size with presence of an infiltrating tumor mass 3.5 x 2.5 cms, with around 30% necrosis. On Histopathological examination, sections showed features consistent with synovial sarcoma and Proximal, distal and lateral margins were free of tumor. After this, patient received 6 cycles of Ifosfamide/Adriamycin regime as adjuvant chemotherapy. Recent four year follow-up shows no evidence of any recurrence or distant metastasis.

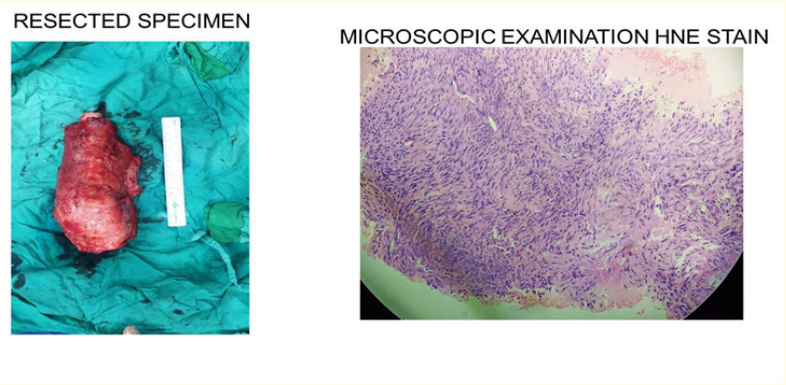

Figure 2

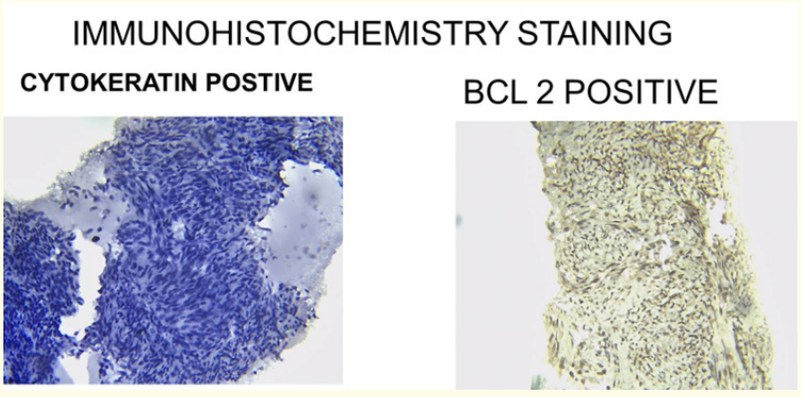

Figure 3

Discussion

Synovial sarcoma of intraarticular origin is rare. The lower extremity is the most common reported site of intra-articular synovial sarcoma in which knee joint is most common followed by hip joint [1]. It shows equal sex distribution[1]. Imaging shows prognostic significance in outcome [6].

Plain radiographs are normal in half of the synovial sarcomas, especially in smaller lesions; calcifications can be present in 30% cases [4]. MRI remains the main imaging modality for intra-articular synovial sarcomas and it also aids in surgical planning [5,6]. MRI scan shows nonspecific heterogeneous mass with T1 iso or low signal and T2 high signal [5]. Tumour size, regional invasion, as well as involvement of adjacent structures may be seen on MRI [5].

The management of intra-articular synovial sarcoma involves a multi-disciplinary and step wise approach beginning with a thorough history and clinical examination. Imaging must include Plain Radiographs, MRI sequences (T1, T2, fat suppression, and post Gadolinium contrast series) [7]. Additionally CT scan, PET scan and ultrasound may be used to rule out any distant metastasis. Imaging modalities showing potentially aggressive or malignant neoplasm require biopsy which can be core needle or open biopsy. Arthroscopic biopsy is not indicated as it risks the intraarticular spread of the tumour cells and potential extravasation [8].

Microscopically, Intra-articular synovial sarcoma is of two main subtypes: Biphasic and Monophasic spindle cell. The Biphasic has 2 components: spindle cells- gland-like epithelial structures and Glandular lumina containing mucin. The Cytological features of biphasic subtype include epithelial component that has moderate, amphiphilic cytoplasm with a round to ovoid nuclei; rarely, squamous metaplasia can occur [6,7]. The monophasic has Infiltrative borders, rarely show hyalinization or myxoid change and nuclear palisading may be seen [6]. The Cytological features of the monophasic subtype include monotonous cells with scant amphiphilic cytoplasm, ovoid to spindled vesicular nuclei, evenly dispersed chromatin, and inconspicuous nucleoli [6,7]. It can be Poorly differentiated which shows Highly cellular, round cells with hyperchromatic nuclei and frequent mitotic activity and necrosis

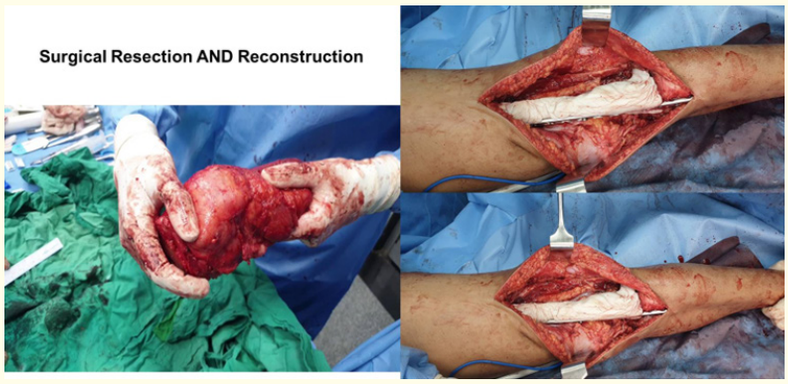

Figure 4

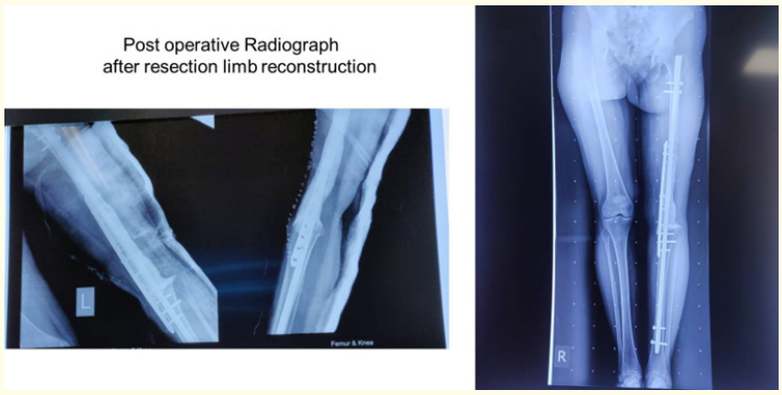

Figure 5

on microscopy and has characteristic features of Focal staghorn (or hemangio-pericytic), branching vascular pattern with mast cells and focal calcification can be seen in 33% of cases [7,8]. Positive IHC makers in Synovial sarcoma are TLE1 (80-90%), Cytokeratin, EMA, BCL2, CD99, NY-ESO1, and SS18-SSX FUSION (95% sensitive and 100% specific). Molecular/cytogenetic testing shows t (X; 18) (p11.2; q11): *SYT-SSX1* fusion in 90%, can detect via RT-PCR, t (X; 18) (p11.21; q11): *SYT-SSX2* fusion, *p16INK4A* gene deletion in 74% helps to distinguish poorly differentiated synovial sarcoma from monophasic and biphasic subtypes [7].

The treatment involves staging the tumor with help of imaging and biopsy and follows principles of limb salvage [8,9]. Preoperatively Neoadjuvant Chemotherapy is given followed by surgical Resection with adequate margins and reconstruction followed by adjuvant chemotherapy like any other musculoskeletal neoplasm [9]. The main importance is given to early detection, careful suspicion to prevent misdiagnosis and inappropriate treatment.

Conclusion

This is important to recognize that soft tissue sarcoma specially intra-articular synovial sarcoma may be missed on normal radiographs and mistaken for other lesions like chronic synovitis, PVNS which leads to misdiagnosis, inappropriate treatment and lead to spread, amputation, and mortality. Orthopedic surgeons must keep a high suspicion for dealing with lesions of unknown etiology and should have a low threshold for seeking an opinion of a musculoskeletal oncologist to assist in concluding appropriate differential diagnosis before intervention. These precautions can prevent complications and improve patient survival rates.

Bibliography

1. Kransdorf MJ. "Malignant soft-tissue tumors in a large referral population: distribution of diagnoses by age, sex, and location". *AJR American Journal of Roentgenology* 164.1 (1995): 129-134.

2. Friedman MV., *et al*. "Intra-articular synovial sarcoma". *Skeletal Radiology* 42.6 (2013): 859-867.

3. Weiss S and Goldblum J. "Enzinger and Weiss's soft tissue tumors". Amsterdam, NL: Elsevier (2008).

4. Bui-Mansfield LT and O'Brien SD. "Magnetic resonance appearance of intra-articular synovial sarcoma: case reports and review of the literature". *Journal of Computer Assisted Tomography* 32.4 (2008): 640-644.

5. Kransdorf MJ., *et al*. "Soft-tissue masses: diagnosis using MR imaging". *AJR American Journal of Roentgenology* 1530.3 (1989): 541-547.

6. Spillane AJ., *et al*. "Synovial sarcoma: a clinicopathologic, staging, and prognostic assessment". *Journal of Clinical Oncology* 18.22 (2000): 3794-3803.

7. Murphey MD., *et al*. "From the archives of the AFIP: imaging of synovial sarcoma with radiologic-pathologic correlation". *Radiographics* 26.5 (2006): 1543-1565.

8. Capanna R., *et al*. "Surgical technique: extraarticular knee resection with prosthesis-proximal tibia-extensor apparatus allograft for tumors invading the knee". *Clinical Orthopaedics and Related Research* 469.10 (2011): 2905-2914.

9. Bergovec M., *et al*. "Intraarticular synovial sarcoma of the knee rising from a lateral meniscus - a case report". *Orthopaedics and Traumatology: Surgery and Research* 104.2 (2018): 227-230.