Rapid Progressive Synovial Sarcoma of Right Knee: A Rare Case Presentation

Rhyan Darma Saputra[1,2*], Dita Anggara Kusuma[1,2], Zeno Aquarista Baharano[1]

[1] *Department of Orthopedic and Traumatology, Oncology Division, Dr Moewardi General Hospital, Surakarta, Indonesia*
[2] *Department of Orthopedic and Traumatology, Faculty of Medicine Sebelas Maret University, Surakarta, Indonesia*

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Abstract

Background: Synovial sarcoma is a rare malignant soft tissue tumor that commonly emerges in the para-articular area, with the knee as the predominant location. The preferred intervention for this tumor is still currently debated

Case description: We presented a 29-year-old male with a very large mass in the right knee. The physical examination revealed impairment of the right lower extremities, particularly the knee. MRI confirmed the extent of the pathology, but the first pathology analysis was inconclusive. A wide resection was performed without prior neoadjuvant therapy. The second pathology analysis from the resected tissue confirmed the diagnosis of biphasic-type synovial sarcoma. The patient was able to walk with no assistance, had an increased range of motion, and the utmost preservation of the limb.

Conclusions: We presented a successful case of wide resection of synovial sarcoma with a promising clinical outcome. The complexity of the case leads to the need for a tailored approach that should be limited to musculoskeletal oncologists.

Keywords: Synovial Sarcoma; Wide Resection; Soft Tissue Tumor; Case Report

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Introduction

Synovial sarcoma is a rare malignant soft tissue tumor of uncertain differentiation. It represents 5 to 10% of all soft tissue sarcomas [1, 2]. The translocation of chromosomes X and 18, which produce SS18:SSX fusion proteins, characterizes synovial sarcoma [2]. Approximately 80% of synovial sarcomas occur in the extremities, commonly in the para-articular area. The knee is the most common location. However, true intra-articular origins are extremely rare, accounting for less than 5% of cases [3].

This sarcoma typically presents at a younger age, occurring in adolescents and young adults with an average age of 30 upon diagnosis, and affects both sexes equally [3, 4]. It is an aggressive malignancy with a high metastatic potential of around 50% in the adult population [5]. As with other soft tissue sarcomas, wide surgical resection to achieve microscopic negative margins, along with perioperative radiotherapy, remains the cornerstone of treatment [2]. Synovial sarcoma has a cancer-specific 5-year survival rate of ~60% in adults. The overall adult 5-year survival rate is 76% for localized tumors, but plummets to 10% if metastatic upon diagnosis [5]. Informed consent was acquired for this case to be written and published, covering all elements included in this paper. We presented this rare case in accordance with the SCARE 2023 guideline [6].

Case Presentation

A 29-year-old male was referred to the orthopedic oncology department at our institution with a large,

*** Corresponding author**: Rhyan Darma Saputra, Sp.OT (k) Musculoskeletal Oncology, M.Kes.
Department of Orthopedic and Traumatology, Oncology Division, Dr Moewardi Province General Hospital, Central Java, 57126, Indonesia AND Department of Orthopedic and Traumatology, Faculty of Medicine Sebelas Maret University, Central Java, 57126, Indonesia
Tel.: +628121659895 Email: rhyands.dr.spot@gmail.com

firm mass in his anterior-medial right knee that had been growing steadily over the previous 6 months. The mass severely interfered with his daily activities. There is no familial tumor or other significant history.

On physical examination, a hard mass on the patient's right knee was found with pain upon palpation. The knee is fixated in a straight position at 12-degree flexion, active movement is virtually impossible, and only 5 degrees of passive movement of the knee is possible. Ankle movement is limited with its position fixated at 40-degree eversion. Fortunately, active movement of the toes and ankle was preserved. There is no neurovascular compromise in the crural and foot region, with normal sensitivity to pinprick sensation and a palpable dorsalis pedis artery. Skin ulceration with necrotic debris and continuous bleeding occurred at the inferior part of the mass (Figure 1A, 1B).

To further investigate the lesion, magnetic resonance imaging (MRI) was performed (Figure 2A-2C). The imaging revealed a large mass spanning almost the medial aspect of the entire knee. The tumor adheres to and causes erosion of the distal epiphysis of the femur, proximal tibia, and fibula. Infiltration of soft tissue is found, including the joint capsule, lateral meniscus, posterior cruciate ligament (PCL), part of the medial vastus muscle, Hoffa's fat pads, and gastrocnemius and semitendinosus muscle with its tendon. A core biopsy was performed but showed inconclusive results. A chest X-ray was performed to investigate possible metastasis, resulting in no lung metastasis.

We decided to perform surgery without any neoadjuvant chemotherapy or preoperative radiotherapy. The patient was positioned supine under general anesthesia, the mass was excised via a medial approach, and identification of the tumor border was performed. Wide excision of the tumor was performed by opening the joint capsule and deliberately sacrificing the medial collateral ligament (MCL). A tissue sample was taken for pathologic analysis.

Macroscopically, the resected tumor was multinodular and encapsulated by a fibrous pseudocapsule with dimensions of approximately 20x15x17 cm (Figure 3A-3C). The microscopic examination revealed the typical appearance of biphasic-type synovial sarcoma (Figure 3D); a

Fig. 1

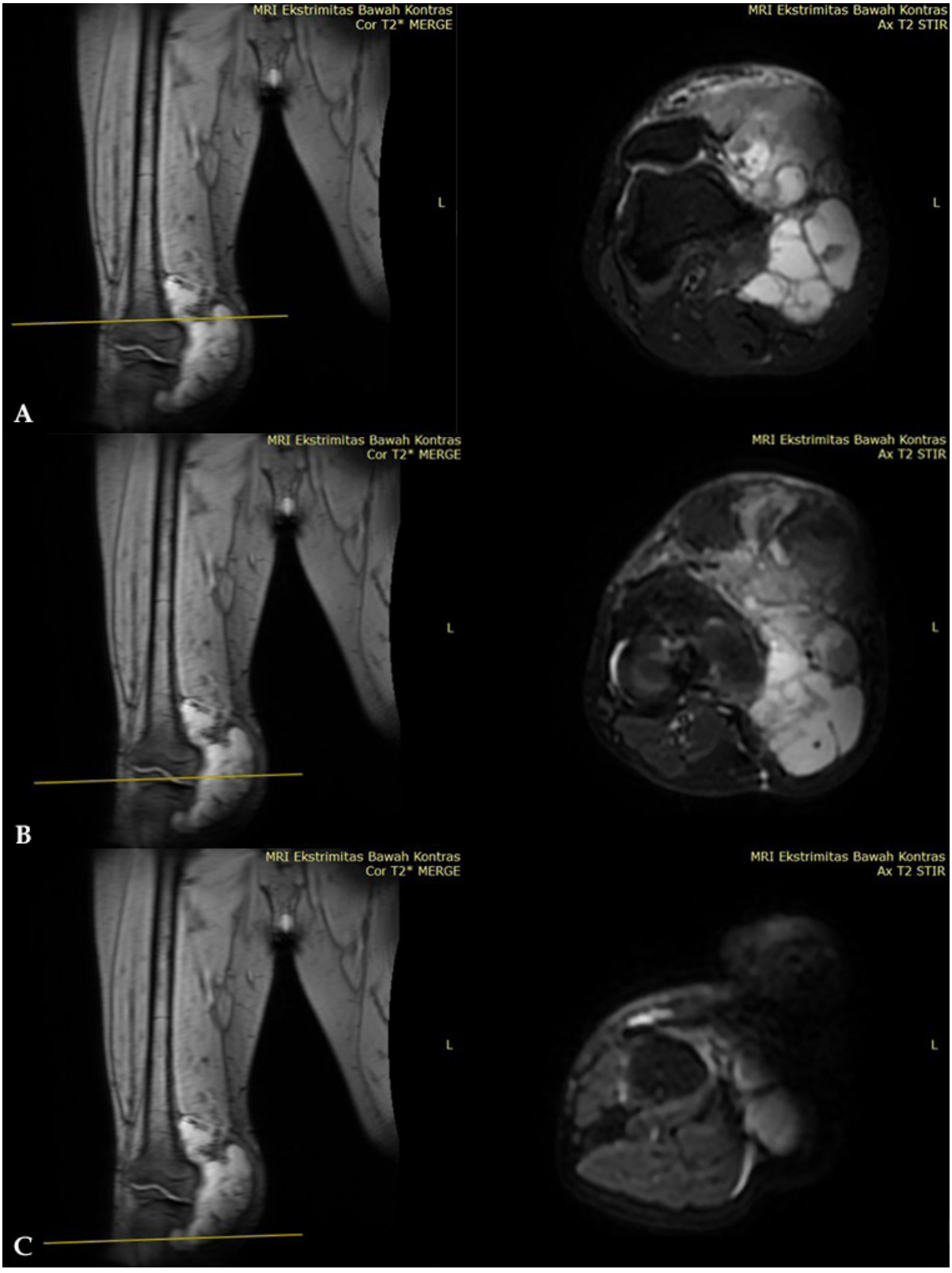
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Fig. 2

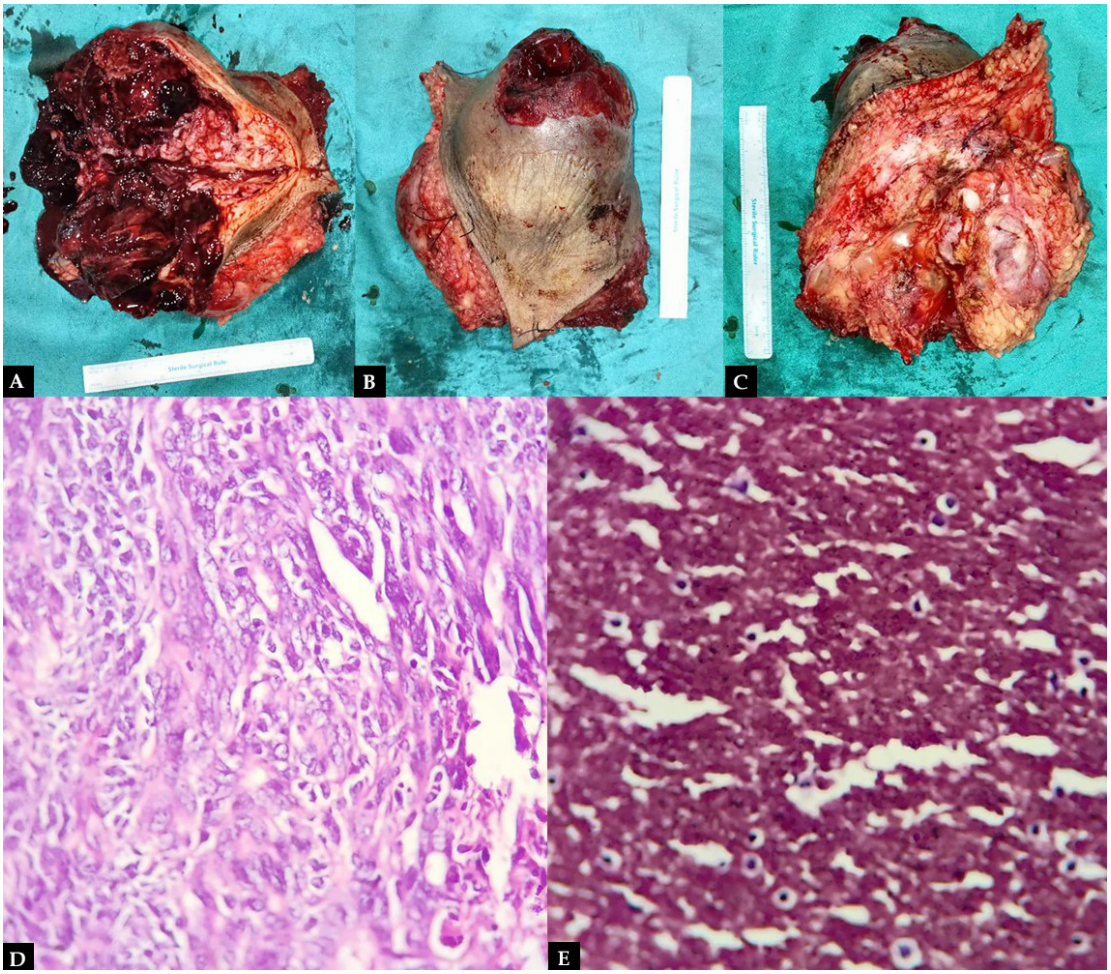

Fig. 3

mesenchymal tumor with proliferation of cells with atypical nuclei, spindle appearance, and several glandular structures (Figure 3E).

The post-operative recovery was relatively fast, and the patient had good clinical outcomes: full-weight bearing and mobile without aid, with an active range of motion of 0° - 0° - 105°. The post-operative radiograph showed satisfactory results (Figure 4A, 4B).

Discussion

The evidence of neoadjuvant therapy for adults with synovial sarcoma is still contradictory [2, 7]. A retrospective study of 112 patients showed that neoadjuvant therapy significantly contributes to the prognosis of patients with grade 3 sarcomas but is not effective for low-grade or grade 2 sarcomas [7]. Conversely, another study suggests that neoadjuvant therapy does not prevent metastasis after resection and that the prognosis is intrinsic to the tumor itself. Furthermore, it implies that current cytotoxic drugs are only poorly efficient in preventing metastasis [8]. Currently, as we progress in understanding the molecular and immunological characteristics of synovial sarcoma, advanced therapies have been proposed [1, 9].

In this case, we performed a wide resection while maintaining a good final clinical outcome. A wide resection surgery with tumor-free margins is crucial in treating soft tissue sarcoma, as retaining any part of a joint capsule conveys a risk of recurrence [10]. Currently, no guideline states the recommended resection margin. For superficial tumors, 1-2 cm is considered adequate. For a tumor that infiltrates deeply adjacent to a neurovascular structure or bone (as in this case), the periosteum or epineurium is utilized as the resection margin. It is important to note that microscopic positive margins might still occur and cause local recurrence after the wide resection is performed; hence, radiotherapy is essential [1, 11]. In our case, it was not performed due to the high cost. A long-term follow-up is planned for this patient.

The lack of standardization and recommendations

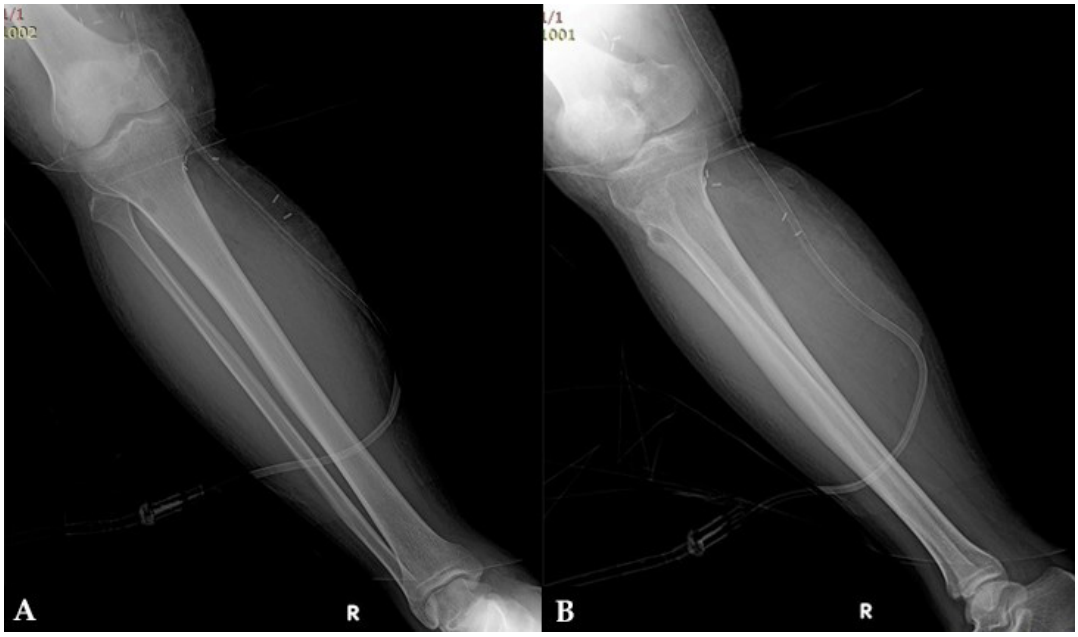

Fig. 4

across the literature regarding the resection margin causes heterogeneity in intervention measures. A musculoskeletal oncologist should manage sarcomas, particularly synovial sarcoma, as each patient needs tailored consideration in determining the therapeutic approach and an acceptable margin in surgery [1, 11, 12].

Conclusion

We presented a case of successful wide resection of synovial sarcoma. It is possible to maintain a good clinical outcome from a wide resection. The management of this tumor should be done by an expert in musculoskeletal malignancy, as it requires tailored judgment for each patient, especially in determining the resection margin of the tumor.

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Conflict of interest

None to disclose.

Author contribution

All authors contributed equally in writing this paper. All writers contributed equally to writing this paper. All writers had read and approved the final manuscript before submission.

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