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# Rare Case of Spindle Cell Sarcoma of the Shoulder Requiring Surgical Disarticulation in an 18-Year-Old Male: a Case Report.

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## ABSTRACT

Spindle cell sarcoma typically affects adults between the ages of 30 and 50 years. This report presents a rare case of spindle cell sarcoma in an 18-year-old male. Although limb-sparing surgeries are generally preferred, the aggressive nature of the tumor in this patient required surgical disarticulation. The patient presented a 6-month history of a painful, rapidly growing swelling over his right shoulder. On examination, the mass was large, ulcerated, and fungating. Further diagnostic investigations were conducted; an MRI revealed a substantial, multi-lobulated, spindle-shaped lesion with heterogeneous enhancement, measuring 13.2 x 13.4 x 15.1 cm (AP x TS x CC), which encased the neurovascular bundle and exhibited multiple enlarged ipsilateral axillary lymph nodes. Histopathology confirmed spindle cell sarcoma. Owing to neurovascular involvement and local invasion, a multidisciplinary team proceeded with shoulder disarticulation. This case highlights the potential for aggressive disease progression in younger patients and the challenges in achieving limb preservation in advanced soft-tissue sarcomas.

**Keywords:** Spindle cell sarcoma; Soft tissue sarcoma; Shoulder disarticulation; Young adult; Neurovascular involvement; Case report

## INTRODUCTION

Sarcomas are a rare group of malignancies arising from mesenchymal tissue, accounting for less than 1% of all adult malignancies [1]. Primary spindle cell sarcoma is an exceptionally rare malignancy and remains one of the

least frequently reported soft tissue tumors [2]. Owing to their rarity, limited data exist regarding their clinical presentation, epidemiological patterns, and optimal management strategies [3].

Spindle cell sarcoma is classified under undifferentiated Soft Tissue Sarcomas (STS). Prognosis is largely influenced by tumor grade and size, with higher-grade and larger tumors associated with an increased risk of metastasis [4]. Sarcomas have a higher incidence in males compared to females [5]. Spindle cell sarcomas most commonly occur between the ages of 30 and 50 years, with the lower limb being the most frequent site of involvement [6]. Sarcomas involving the upper extremities are rare, accounting for approximately 11.2% of all soft tissue sarcomas [7].

The term "Spindle cell" refers to the elongated, spindle-shaped appearance of the tumor cells, which influences the course of treatment compared to sarcomas exhibiting epithelioid morphology [8]. Spindle cell sarcomas are typically diagnosed preoperatively based on patient history and physical examination, with Magnetic Resonance Imaging (MRI) playing a crucial role in assessing the extent and characteristics of the tumor [9].

The preferred surgical approach for soft tissue sarcomas is radical resection, with limb-sparing procedures being favored whenever oncologically feasible [10]. In the majority of cases, approximately one-third of patients experience local recurrence, typically within 18 months of initial treatment. The lungs are the most frequent site of metastasis. Patients with unresectable metastatic disease generally have a poor prognosis and are managed with systemic chemotherapy [11]. According to one retrospective study, the high relapse rate of spindle cell sarcoma means that an initial isolated local recurrence of Soft Tissue Sarcoma (STS) significantly increases the risk of subsequent local recurrences [12].

## CASE PRESENTATION

An 18-year-old male, with no known co-morbidities, presented to the orthopedic outpatient department with a mass on his right shoulder that had been gradually increasing in size over 6 months. On examination, there was a large, firm, exophytic, fungating mass measuring approximately 15 x 13 cm over the anterior aspect of the shoulder. It was irregular, ulcerated, and with areas of active bleeding. The overlying skin was erythematous and hyperpigmented. (Figure 1). The surrounding tissue appeared edematous.

**Figure 1:** shows an exophytic, fungating mass on the anterior aspect of the right shoulder.

Neurovascular assessment of the limb revealed compromised vascular status: the capillary refill time exceeded 2 seconds, and the radial pulse was weak. The sensory examination was unremarkable. Motor function assessment revealed reduced elbow flexion, while movements at the wrist and fingers were intact.

Magnetic Resonance Imaging (MRI) of the right shoulder with contrast (Figure 2) demonstrated a heterogeneously enhancing abnormal signal intensity mass centered over the right axilla. The lesion exhibited multiple cystic areas within the mass, along with some non-enhancing areas, suggesting internal areas of necrosis. There was a marrow signal abnormality within the proximal humerus, predominantly involving the surgical neck and proximal shaft, with associated underlying bone remodeling and cortical thinning. Multiple enlarged lymph nodes were noted in the ipsilateral axilla.

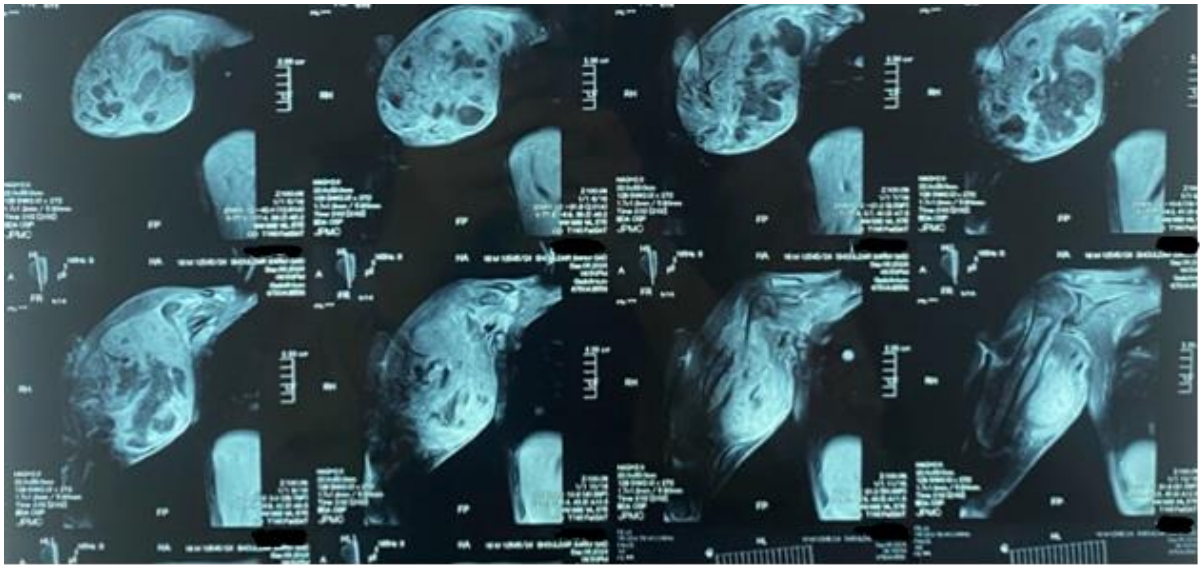

**Figure 2:** Magnetic resonance imaging of the affected region.

An incisional biopsy confirmed the diagnosis of spindle cell sarcoma. Histopathological examination revealed fibrovascular tissues showing a morphological profile of a spindle cell lesion with foci of clear cell changes and associated multiple fascicles of skeletal muscle. Unfortunately, immunohistochemical evaluation could not be performed. After obtaining consent, the patient underwent surgical disarticulation of the right shoulder (Figure 3,4). The procedure was performed by a team of orthopedic surgeons, with intraoperative consultation from a thoracic surgeon to assess for possible invasion into the chest cavity. Intraoperatively, the neurovascular bundle was found to be compromised.

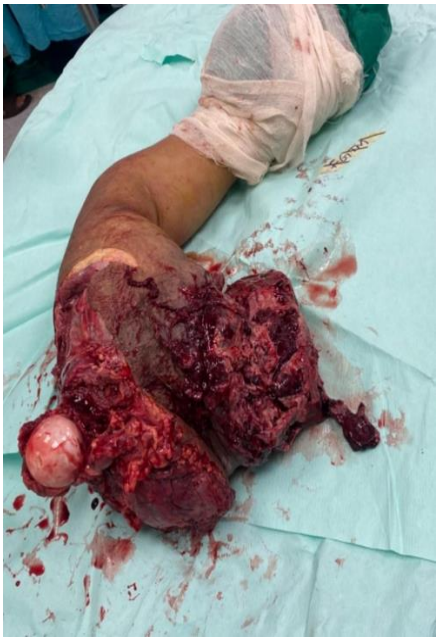

**Figure 3:** Surgical removal of the tumour

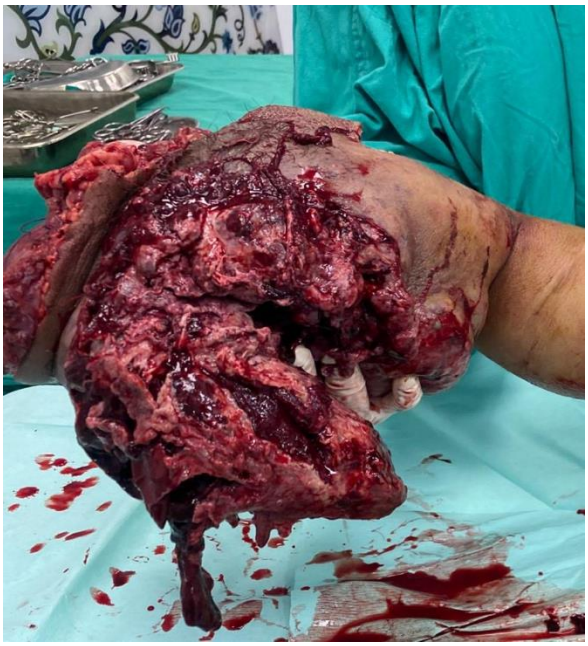

**Figure 4:** total width and length of the tumor

The patient was monitored postoperatively for recovery and any complications. Two units of packed red cells were transfused during the hospital stay. The post-operative medication regimen included intravenous Tramadol for analgesia, Ceftriaxone, and Metronidazole for antimicrobial coverage. The patient was referred to the oncology department for consideration of adjuvant therapy and further multidisciplinary management.

## DISCUSSION

Epidemiological data collected over time have demonstrated a steady and significant increase in the incidence of soft tissue sarcomas, highlighting a growing concern in the global cancer burden [13]. Spindle cell sarcomas are

considered one of the most common soft tissue sarcomas affecting extremities, second only to mesenchymal cell sarcomas [14]. There are various factors that contribute to the development of spindle cell sarcoma, including genetic predisposition, previous radiotherapy, and, in some cases, injury or chronic inflammation in patients with an existing susceptibility to this malignancy [15].

These malignancies are most prevalent in economically developed regions; however, significant increases in incidence have also been observed in Central Asia, Central Europe, and Southern Sub-Saharan Africa, largely attributed to growing age as well as overall population growth [16]. The rarity and diversity of Soft Tissue Sarcomas (STS) have made understanding their metabolism challenging. Traditional views separating glycolytic and oxidative tumors are being reconsidered, as STS cells often display mixed metabolic profiles that enhance adaptability. New models show these tumors can alter their metabolism, affecting energy use. Their metabolic balance depends on key substances like amino acids, NAD+/NADH, lactate, and vitamin B5, which influence tumor growth and development [17].

This case report describes an 18-year-old male presenting with a large, exophytic, ulcerated mass over his right shoulder, progressively increasing in size over the time period of 6 months. The mass measured approximately 15 cm, with areas of necrosis and active bleeding. The average age for spindle cell sarcoma is reported to be 61 years, with the tumor typically affecting middle-aged older adults [18]. The mean size of soft tissue sarcomas at presentation is around 10 cm on average [19]. The unusually young age of our patient, combined with the large tumor size, makes this a rare and clinically challenging case.

Ultrasound is usually the first tool used to check for soft tissue sarcomas, while MRI is the main method for examining the details of the tumor. PET/CT and newer MRI techniques are sometimes used as extra tools to help with staging the cancer or for detecting recurrences [20]. Immunohistochemical tests, such as β-catenin nuclear staining, Ki-67 staining, and CD34 antibody testing, play a crucial role in making a definitive diagnosis [21]. An MRI scan was chosen due to its superior soft tissue contrast and ability to accurately delineate the extent of tumor involvement. Additionally, the range of diagnostic tests was limited by the patient's low socio-economic background. In our case, immunohistochemical tests could not be performed due to limited resources. The absence of IHC poses a challenge in confirming the exact tumor subtype and guiding targeted management.

Treating soft tissue sarcomas is both diagnostically and therapeutically complex. For localized disease, surgery is the primary treatment. In high-risk cases, a multimodal approach, such as neoadjuvant chemotherapy with or without regional hyperthermia, may help shrink the tumor and target metastases before surgery [22]. The primary treatment for spindle cell sarcoma of the extremities is limb-sparing surgery, which has an overall success rate of approximately 70% worldwide [23]. Significant progress has been made over the past two decades in developing targeted therapies for soft tissue sarcoma. Advances in understanding cellular markers and pathways involved in sarcoma development have led to the creation and approval of several new treatments [24]. Multiple studies have confirmed that post-operative radiotherapy after wide local excision or function-preserving surgery improves local disease control, though it does not affect overall survival or reduce distant metastases [25]. According to one study, there is no significant difference in overall survival rates between patients undergoing limb-salvage surgery and those treated with amputation or disarticulation [26].

Surgical disarticulation of the right shoulder was performed for our 18-year-old patient. The post-operative period remained uneventful, with appropriate analgesic and antimicrobial therapy administered. In this case,

shoulder disarticulation was deemed necessary, as attempting limb-salvage surgery would have compromised oncological margins and adequate disease control. Intra-operatively, the neurovascular bundle was found to be encased by the tumor, precluding the possibility of achieving clear surgical margins with limb-sparing resection. Furthermore, extensive tissue necrosis and active hemorrhage were noted. Complete resection of the sarcoma without compromising limb function was deemed unfeasible. Therefore, surgical disarticulation was considered the most appropriate and oncologically sound option, given the extensive size, local invasion, and neurovascular involvement of the tumor.

## CONCLUSION

This case highlights the importance of early diagnosis and timely intervention in soft tissue sarcomas, particularly in regions with a high poverty index, where delayed presentation can lead to radical surgical management. It also highlights the urgent need for improved healthcare accessibility to enhance oncological outcomes in resource-limited settings.

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