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A Rare Tumor in a Rare Localization: Elbow Synovial Sarcoma

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Abstract **Case Report**

Synovial sarcoma are extremely rare malignant tumors of soft extra-skeletal tissue accounting for less than 1% of all malignant tumors. Despite their name, they do not arise from synovial tissue, and their pathogenesis remains unknown. The knee and ankle are the most common sites of occurrence. Due to unclear risk factors, no screening recommendations currently exist. They are characterized by an unpredictable course; hence, multidisciplinary management must be initiated at an early stage in order to improve prognosis. The mainstay of treatment is wide surgical excision, which can often be challenging, requiring the sacrifice of noble structures, while reconstructive surgery can be laborious. The presence of pulmonary metastasis is an indication of poor prognosis. Herein, we report a rare case of a localized elbow synovial sarcoma in a 48-year-old woman with no comorbidity. She underwent surgery for complete tumor removal and had an uncomplicated post-operative follow-up.

Keywords: Synovial Sarcoma, Soft Tissue Sarcoma, Elbow Tumor.

INTRODUCTION

Synovial sarcoma (SS) are extremely rare malignant tumors of soft extra-skeletal tissue accounting for less than 1% of all malignant tumors. They are part of the diverse group of mesenchymal malignancies known as sarcomas and they account for about 8% of these malignancies. They are mainly found in the lower limbs, specifically in the knee and ankle regions. They are high grade tumors and are characterised by local invasion and a propensity to metastasise. They are more common in adolescents and young adults, with a sex ratio (3:1) in favour of men. Despite their name, they do not arise from synovial tissue although they exhibit histological characteristics resembling synovial tissues. As with most soft tissue sarcomas, their pathogenesis is still unknown and there are no well-established risk factors; therefore, no screening program has been evaluated or could be recommended [1-3].

The diagnosis of SS is based on a combination of clinical and radiological evidence and confirmed by histopathological examination. There are three histopathological forms: monophasic with spindle cells, biphasic with a double epithelial and spindle cell contingent and finally the undifferentiated form. High-grade tumors with high metastatic potential, their treatment involves locoregional control: conservative treatment is possible, combining several therapeutic methods (surgery supplemented by radiotherapy and possibly chemotherapy). Tumor control can only be achieved by wide surgical excision, with or without radiotherapy. The rate of local recurrence varies between 20 to 30% in the literature [2].

In this paper, we report a rare case of a localized elbow synovial sarcoma in a 48-year-old woman with no comorbidity in which tumor control was obtain by wide surgical excision without adjuvant radiotherapy no chemotherapy.

CASE REPORT

A 48-year-old woman with no relevant past medical history came to consult in our department for a painless mass of the right elbow. She reported that the mass appeared 10 years ago and its size has been increasing progressively with no history of bodyweight lost. Physical examination revealed a mass located on the lateral aspect of the elbow. This was a non-pulsatile, soft and painless spherical mass with a regular surface measuring 4cm x 3cm (Figure n°1), and mobile with respect to the superficial plane. Locoregional ganglionic regions were free. A magnetic resonance imaging revealed a well-defined extra-skeletal mass in the lateral aspect of the elbow (Figures n°2 A and B) without signs of locoregional invasion. Computed tomography in search of distance metastasis was normal. Initial incisional biopsy revealed synovial sarcoma prompting the patient to undergo definitive surgery for mass removal.

The tumor was accessed and removed via incisions around it borders (Figure n°3A). After carefully dissecting the soft tissues, the ulnar nerve was identified and protected. A wide excision of the tumor was performed, involving part of the capitellum (Figure n°3 C, E and F), necessitating the reconstruction of the lateral collateral ligament of the elbow (Figure n°3C). For this, the palmaris longus tendon from the homolateral forearm was used for ligamentoplasty. To maintain the elbow flexed at a 90° angle, fixation with a Kirschner's wire passing through the olecranon into the humeral medullary canal was performed. The wound was thoroughly irrigated and closed under a negative suction drain. A posterior splint was applied at a 90° angle. To prevent edema, the elbow was kept elevated on a pillow.

The patient's initial post-operative course was uncomplicated, and she was discharged on oral analgesics with a follow-up appointment scheduled for two weeks. Histopathological findings confirmed a monophasic synovial sarcoma with no evidence of local invasion (see Figure n°4). At the three-week post-operative follow-up, the kirschner's wire was removed and the patient began an 8-week gradual functional rehabilitation program with no weight bearing. A gradual load-bearing rehabilitation program was initiated at the three-month follow-up. By the seventh month follow-up, the patient had regained full range of motion of her elbows. At the last follow-up, two years' post-operation, the patient had a good general state with no tumor recurrence.

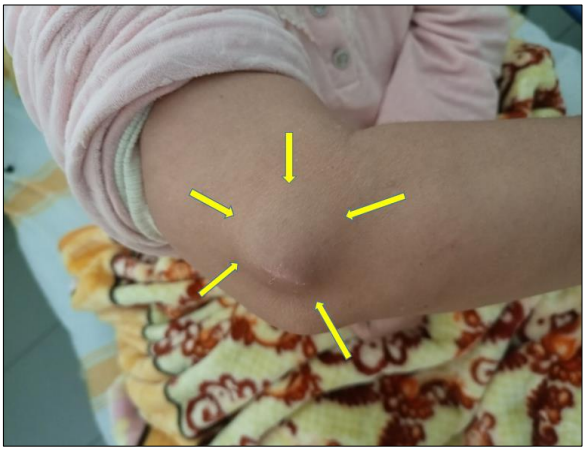

Figure 1: A clinical image of patient's elbow. The yellow arrows on the image are showing the mass on the lateral aspect of the elbow

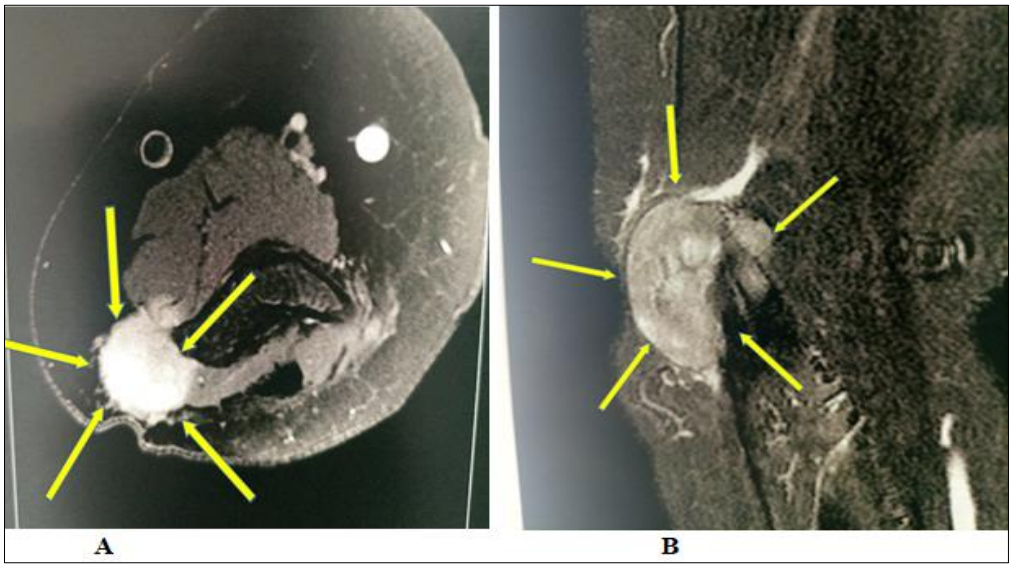

Figure 2: (A) A transvers section of magnetic resonance image of the elbow. The yellow arrows in the image are showing a well-defined extra-skeletal mass. (B) A sagittal section of magnetic resonance image of the elbow. The yellow arrow in the image are showing a well-defined extra-skeletal mass

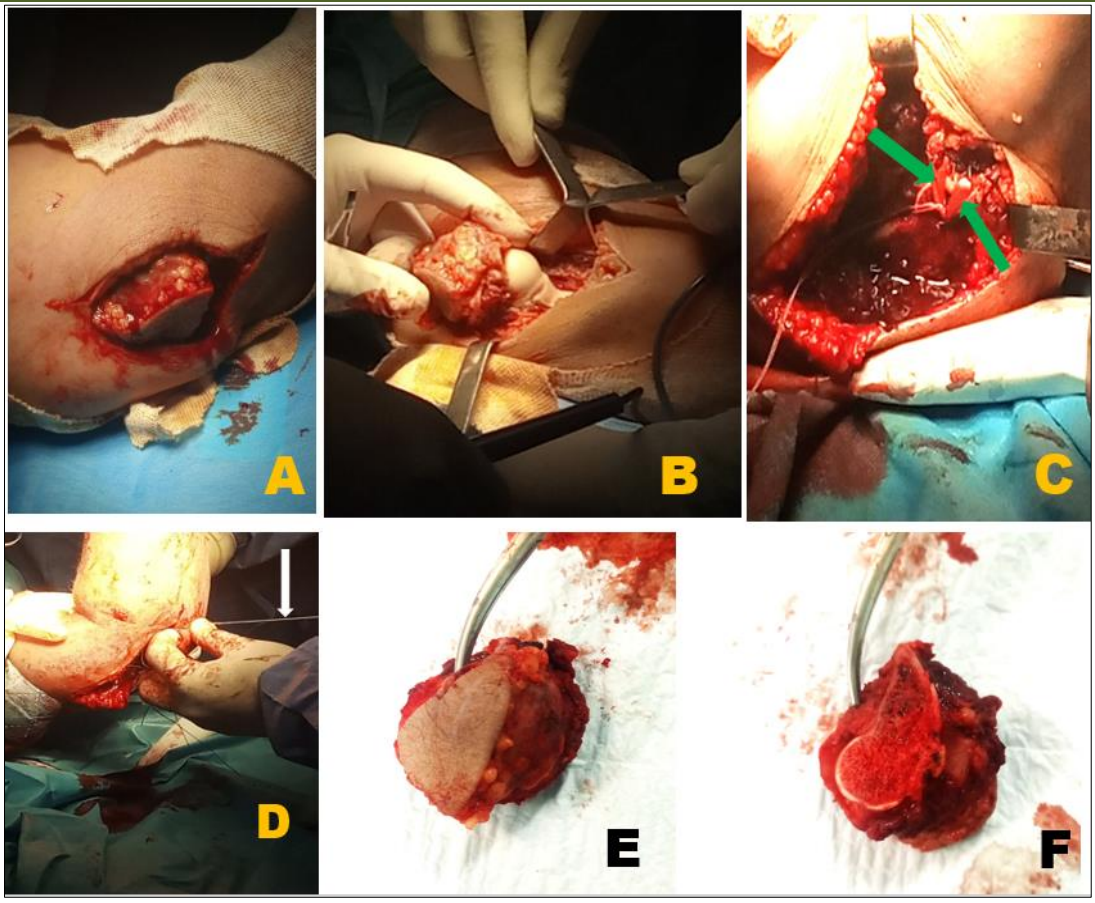

Figure 3: Intra-operative images of wide excision. The mass was totally removes beyond its margin including part of capitellum. (A) image showing access to the mass (B) image showing the mass, capitellum and trochlea of the humerus (C) green arrows in the image showing ligamentoplasty (D) white arrow in the image showing fixation with a Kirschner's wire in order to maintain the elbow flexed at an angle of 90° (E and F) resected mass (E) image showing wide excision of mass with part of capitellum

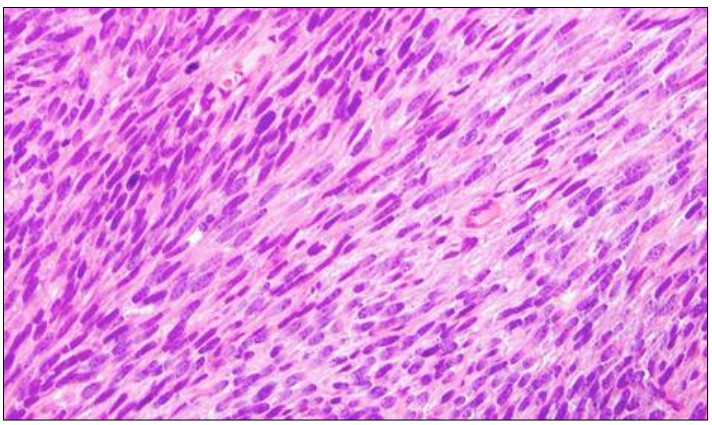

Figure 4: A histological section of the resected mass. Image showing a microscopic feature of a monophasic synovial sarcoma variant

DISCUSSION

In this case report, we describe the clinical presentation and management of a rare localization of synovial sarcoma in a 48-year-old woman with no comorbidities. The synovial sarcoma (SS) was localized at her right elbow.

SS is part of the diverse group of mesenchymal malignancies known as sarcomas. Sarcomas are uncommon malignant tumors of non-epithelial, extra-skeletal tissue in the body, constituting less than 1% of all malignant tumors and 2% of all cancer-related deaths. SS represents the most prevalent form of soft tissue sarcomas, accounting for approximately 8% of all soft tissue sarcomas. It is primarily found in the lower limbs,

specifically in the knee and ankle regions. Although it can manifest at any age, it is more frequently observed in adolescents and young adults, with a mean age of 25 years. The majority of authors accept a male predominance in SS cases [1-3].

While SS exhibit histological characteristics resembling synovial tissue, they do not originate from such tissue. They are high-grade tumors, characterised by local invasion and a propensity for metastasis. The prognosis for SS is poor, primarily due to local recurrence and, notably, early metastasis. The lung constitutes the preferred site for distant metastasis (30 to 70% of SS), with the presence of lung metastases being an indicator of poor prognosis. As with most soft tissue sarcomas, the pathogenesis of SS remains unknown, and there are no well-established risk factors. Consequently, no screening program has been evaluated or can be recommended [2, 3].

The clinical presentation is typically slow progressing, insidious, and pauci-symptomatic tumor, characterized by a single nonspecific soft mass adhering to deep planes near major joints of the limbs, often mimicking benign lesions, which contributes to delayed diagnosis [4]. Magnetic resonance imaging (MRI) plays a crucial role in the diagnostic process. Like for all soft tissue tumors, it remains the gold standard for assessing locoregional extension and providing accurate tissue characterization. While the appearance of synovial sarcoma (SS) on MRI is not specific, the diagnosis is suspected when a soft tissue mass adhering to a tendon or joint capsule, well-defined in 91% of cases, is observed [3]. Computed tomography (CT) is useful for researching distant metastases, particularly pulmonary metastases. Initial incisional biopsy for histological analysis is of paramount importance to determine the definitive management strategy. Final diagnosis is based on histopathological findings which determine one of the three histopathological forms: monophasic with spindle cells, biphasic with a double epithelial and spindle cell contingent and finally the undifferentiated form.

SS are characterized by an unpredictable course; hence, multidisciplinary management must be initiated at an early stage in order to improve prognosis. Achieving carcinological treatment can often be challenging, requiring the sacrifice of noble structures, while reconstructive surgery can be laborious. Radical treatment, when combined with adjuvant radiotherapy and chemotherapy, can improve the patient's quality of life [1].

In this case, the clinical presentation was a slow progressing, insidious, and pauci-symptomatic tumor, characterize by a single soft mass on a major joint, consistent with previous literature [4]. The patient reported a mass that appeared ten years ago on her right elbow, progressively increasing in size with no history of body-weight loss. Physical examination revealed healthy patient with a painless spherical mass, 4cm x 3cm on her elbow. MRI confirmed an extra-skeletal mass of the elbow without signs of locoregional invasion, while CT scans showed no distant metastasis. Initial biopsy confirmed synovial sarcoma, leading to definitive surgery for mass removal. Carcinological treatment involved wide excision surgery, sacrificing the capitellum of the humerus and the lateral collateral ligament, necessitating laborious ligamentoplasty. Definitive histopathological diagnosis was that of monophasic synovial sarcoma with no evidence of local invasion. Adjuvant radiotherapy and chemotherapy were unnecessary due to the absence of local invasion and distant metastasis.

CONCLUSION

Synovial sarcomas are high-grade tumors of extra-skeletal soft tissue, characterized by local invasion and a propensity for metastasis, predominantly affecting adolescents and young adults. They exhibit an unpredictable course, chiefly due to local recurrence and early pulmonary metastasis, resulting in a poor prognosis. Achieving tumor control typically involves wide excision surgery, potentially supplemented by adjuvant radiotherapy and chemotherapy. However, achieving carcinological treatment can be challenging, necessitating the sacrifice of noble structures, and reconstructive surgery may be laborious. Their primary localization on major joints of limbs requires tailored rehabilitation therapy post-operatively. Thus, early initiation of a multidisciplinary approach is imperative for improving prognosis and optimizing treatment outcomes.

Highlights

- Clinically synovial sarcomas are typically slow progressing, insidious, and pauci-symptomatic malignant tumors, characterized by a single nonspecific soft mass adhering to deep planes of major joints of lower limbs, often mimicking benign lesions;
- Despite its name, synovial sarcoma does not arise from synovial tissue;
- They are high-grade tumors, characterised by local invasion and a propensity for metastasis particularly pulmonary metastasis;
- The main stay of treatment is wide surgical excision.

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Consent: Written informed consent was obtained from the patient for publication of this case report including the images used.

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