Case report

A case report on Synovial sarcoma of the foot: Diagnosis and treatment

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ABSTRACT

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| **Aims:** To present a rare case of synovial sarcoma (SS) arising in the foot of a young adult male and highlight the diagnostic challenges, treatment approach, and prognosis associated with this aggressive malignancy.  **Presentation of case:** A 23-year-old Caucasian male with a history of polydactyly presented with a progressively painful mass in the third toe of the left foot. MRI revealed a 4 cm multilobulated soft tissue mass encasing the first phalanx. Biopsy with pathological examination confirmed synovial sarcoma, and the patient underwent metatarsal-level amputation with clear margins. Two years postoperatively, the patient experienced a regional relapse in the gluteal area and developed pulmonary metastases. Despite initiation of chemotherapy, the disease rapidly progressed, and the patient died within a month of relapse.  **Discussion:** Synovial sarcoma is a rare soft tissue sarcoma, typically affecting young adults and often arising in the extremities. It frequently presents with nonspecific symptoms and can mimic benign conditions, delaying diagnosis. MRI is the imaging modality of choice, and definitive diagnosis relies on histopathological analysis. While surgery with negative margins remains the cornerstone of treatment, recurrence and metastasis are common, particularly in larger tumors.  **Conclusion:** This case illustrates the aggressive nature of synovial sarcoma despite adequate initial management. Clinicians should maintain a high index of suspicion for SS in persistent foot masses, and ongoing research is vital to improve outcomes through better systemic therapies and relapse prevention strategies. |

*Keywords: Synovial sarcoma, Lower limb, Prognosis, Survival.*

1. INTRODUCTION

Synovial sarcoma (SS) is a relatively rare soft tissue sarcoma (STS). It accounts for 5 to 10% of all STS diagnoses [1,2]. It is a unique subtype of STS as it occurs in young patients with a median age of 39 years and affects both sexes equally [3]. SS can develop anywhere in the body with the extremities being the most common site. The lower extremity is the most involved site, followed by the upper extremity and the head and neck region [4,5]. The clinical presentation of synovial sarcomas differs from the typical large and rapidly growing painless mass of other soft tissue sarcomas. It usually presents as a slow growing mass with possible pain and swelling [6,7]. The disease is localized in the majority of cases, but up to 13% of patients have distant metastases at the time of diagnosis [3]. Survival rates are variable according to the current literature, affected mainly by local and metastatic relapses. Synovial sarcoma is unique in this matter with relapses that occur much later than other subtypes of STS [6].

2. CASE PRESENTATION

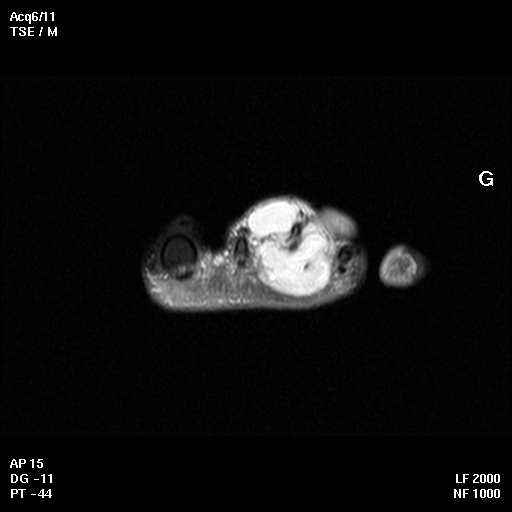
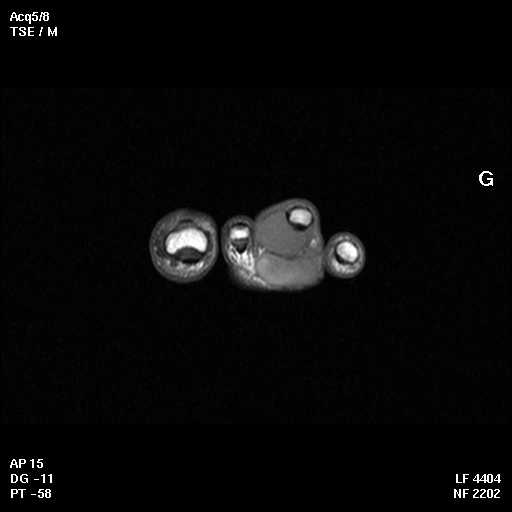
A 23-year-old Caucasian male with past medical history of polydactyly of the left hand and foot presented with a mass of the third toe of the left foot. The mass had been growing slowly over the previous 2 years. The patient stated that the mass had become progressively more painful with a recent rapid increase in volume over the last seven months. The patient also reported important discomfort when wearing shoes due to the size and tenderness of the mass. The patient denied any trauma, systemic symptoms, or constitutional complaints such as fever or weight loss. On physical exam, the skin was normal on the dorsal aspect of the foot and presented a small, ulcerated lesion on the plantar side. Inspection revealed a firm, non-mobile, tender mass of the third toe at the level of the first and second phalanges extending to the metatarsal region, measuring approximately 4 x 3 cm. Distal pulses and neurological examination of the foot were normal (Figure 1). Standard foot X-ray showed normal bone structure. MRI of the left foot demonstrated a well-defined, multilobulated soft tissue mass of 4 cm, encasing the first phalanx of the third toe displacing the second and fourth toes. There were no cortical bone lesions of the phalanx and cancellous bone signal was normal. The mass showed iso-intensity compared to muscle on T1-weighted images and hyperintensity on T2 with an intense and heterogenous enhancement after gadolinium injection. The flexor and extensor tendons of the third toe were simply displaced by the tumor and showed normal MRI signals (Figure 2). Open incisional biopsy was performed. Histology examination revealed a neoplasm composed of spindle cells moderately atypical with an abnormal mitotic activity.

Amputation at the level of the metatarsal bones was performed. Local flaps from the lateral and medial aspects of the foot were used to close the skin (Figure 3 and 4). The histopathologic exam confirmed the prior diagnosis and showed clear surgical margins. Two years following the initial treatment, the patient developed a regional relapse, characterized by a soft tissue mass in the left gluteal region. Further investigation with CT scans identified bilateral lung nodules, indicative of distant metastasis. Despite commencing chemotherapy, the patient's condition rapidly deteriorated, and they died within the subsequent month.

**A close-up of a foot

Description automatically generated**

**Figure 1:** Clinical aspect of the mass of the third toe of the foot displacing the second and fourth toes.



**Figure 2:** MRI images of the foot showing a well-defined, multilobulated soft tissue mass of 4 cm, encasing the first phalanx of the third toe displacing the second and fourth toes.



**Figure 3:** Resected piece of the amputation of the five toes to the metatarsal level with macroscopically intact margins.

A foot with stitches on it

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**Figure 4:** Skin closure was performed by two plantar flaps; medial and lateral covering the bone completely with good soft tissue padding.

3. discussion

Synovial sarcoma is a rare subtype of Soft tissue sarcomas [8]. STSs are a heterogeneous group of rare cancers originating from mesenchymal tissues [9,10]. While the term 'synovial sarcoma' suggests a connection to the synovium, this is a misnomer. The true cellular origin of SS remains unclear, and these tumors do not arise from synovial tissue. While SS can occur across all ages, it characteristically affects young adults, with peak incidence in the 30s and a median age of diagnosis around 30 [11].

Synovial sarcoma predominantly arises in the soft tissues of the extremities, particularly near joint capsules and tendon sheaths of the foot, knee, or ankle. However, it can also manifest in other locations, including the trunk, head and neck, abdomen, pelvis, mediastinum, pleura, lungs, and, rarely, bone [11–14]. SS is considered to be the most common STS of the foot [15–17].

Similar to other STSs, SS often presents with nonspecific symptoms like swelling or pain due to tissue compression [6]. These symptoms can mimic benign conditions such as trauma, myositis, bursitis, or tendonitis, leading to potential misdiagnosis. Notably, patients may experience prolonged pain at the tumor site before swelling becomes evident [6].

Magnetic resonance imaging (MRI) is the gold standard for diagnosing synovial sarcomas, similarly to other STSs. It defines the local extent of the soft tissue mass with excellent visualization of surrounding tissue [6]. SSs are typically a heterogenous mass with low intensity on T1-weighted images and high intensity on T2 sequences with enhancement after contrast [18].

Diagnosis relies on biopsy and pathological evaluation. Open incisional biopsies have been considered to be the gold standard for STSs to define the sarcoma subtype and tumor grade [19]. SS presents as one of three histological subtypes: monophasic, biphasic and poorly differentiated [20]. Monophasic is the most frequent histological variant composed of monomorphic spindle cells with moderate cytologic atypia organized in fascicles [20,21].

Most patients present with localized disease at the time of diagnosis. The rates of initial metastatic disease in the literature range from 6 to 18% [3,6,22,23]. The most common metastatic site being the lungs [3,24].

The cornerstone of SS treatment continues to be surgical excision ensuring negative margins, supplemented by radiotherapy with or without chemotherapy based on patient and tumor-specific factors. In the absence of specific guidelines defining ideal negative margins for SS, the surgical approach mirrors that employed for other STSs [6]. Historically, amputation was frequently performed; however, advancements in adjuvant therapy and cross-sectional imaging have enabled limb-salvage surgery for most patients. While primary amputation is seldom needed for localized limb SS, it is the right initial treatment for a few cases due to specific medical factors and patient wishes [25]. In the lower limbs amputation is usually indicated when inadequate function of the limb would result from complete tumor clearance or when composite tissue is involved in the sarcoma [25].

Radiation therapy is recommended in the neoadjuvant or adjuvant setting for tumors larger than 5 cm or when a close surgical margin is anticipated to preserve major neurovascular structures or bone [26]. It has been demonstrated that radiotherapy helps to improve local control and may have overall survival benefits [27].

Five-year survival rates are variable in the literature, ranging from 59 to 75% [6]. Several prognostic factors have been thoroughly evaluated including: patient age, tumor size, grade, location and negative surgical margins [11,28,29]. Deshmukh et al. demonstrated that tumor size was a main prognostic factor and predicted worse overall survival when exceeding 5 cm [15]. Patient age at the time of diagnosis is also considered a main prognostic factor and survival rates are better among children. Studies using registry data (Sultan et al. [30] ; Smolle et al. [31] ) consistently demonstrate superior five-year survival rates in children and adolescents (83% and 89%, respectively) compared to adults (62% and 75%, respectively). Vlenterie et al. [11] further support this trend, showing a clear inverse relationship between age and survival, regardless of other factors.

4. Conclusion

In summary, this case report details the initial presentation of synovial sarcoma in the foot of a young adult male, which often presents as a gradually growing mass. The diagnostic process, aided by MRI, led to surgical resection with limb salvage as the primary treatment. However, the regional and distant relapse underscores the aggressive potential of synovial sarcoma and the challenges in achieving long-term control even with initial local treatment. This case emphasizes the importance of considering synovial sarcoma in the differential diagnosis of foot masses and highlights the critical need for continued research into more effective systemic therapies and strategies to prevent both local and distant recurrence in this challenging malignancy, regardless of the primary tumor location. Given the aggressive nature of synovial sarcoma, for newly diagnosed patients, cytogenetic with molecular profiling is important. Genetic specific alterations may inform oncologist decision-making around adjuvant therapy options. Tailoring treatment based on these findings holds the potential to improve patient prognosis and optimize therapeutic outcomes.

Consent

All authors declare that ‘written informed consent was obtained from the patient’s next of kin for publication of this case report and accompanying images.

Ethical approval

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

Disclaimer (Artificial intelligence)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

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