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Case report

A rare case and unusual localization of a poorly differentiated giant synovial sarcoma of the ankle: Case report and literature review

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ABSTRACT

Introduction: Synovial sarcoma is a rare and aggressive soft tissue tumor that affects the para-articular regions of the large joints. It originates from mesenchymal cells with synovial differentiation. Its management remains surgical with wide excision.

Case presentation: We report a case of synovial sarcoma with rare localization that occurred in a 37-year-old man on the inner side of the right ankle, and we recall the clinical, radiological, and histological signs that allowed us to suggest the diagnosis as well as multidisciplinary management.

Discussion: Synovial sarcoma is a highly aggressive soft tissue tumor with a high risk of spreading. It is a deceptive tumor in some clinical and morphological aspects that may indicate benignity. Although non-specific, MRI is critical for guiding diagnosis, identifying prognostic criteria, and ensuring post-treatment follow-up.

Conclusion: This rare case underlines the importance of evoking a synovial sarcoma in front of a mass of soft parts of the benign aspect of the ankle, as well as adapting the treatment in order to prevent possible metastases, especially in the lungs.

1. Introduction

Synovial sarcoma accounts for 7 to 8% of malignant sarcomas [1]. Although it is most common in adults, adolescents and children account for 30% of reported cases. This tumor frequently appears as a slow-growing mass. The lower and upper limbs, particularly the para-articular regions of the large joints, are the most affected. It is a primary mesenchymal tumor with the same organization as synovial tissue, but it does not have a synovial origin [2]. We report a rare case of synovial sarcoma in a 37-year-old man who developed a giant form with unusual localization in his ankle, as well as the clinical, radiological, and histological signs that allowed us to make the diagnosis and the multidisciplinary management. This manuscript has been reported in line with SCARE's 2020 Criteria [3].

2. Case presentation

This is the observation of a 37-year-old alcoholic and chronic cannabis user who was weaned 10 years ago and has no pathological history. He consulted regarding a tumor on the inside of his right ankle. A swelling next to the internal malleolus has revealed it for the past two years. After 10 months of evolution, the tumor's size increased and became ulcerative, accompanied by periodic bleeding. Faced with the ulcerative and hemorrhagic aspects of the tumor, the patient sought treatment at the University Hospital Center. An ulcerative and budding mass occupying the entire medial surface of the right ankle, measuring approximately 150 × 150 × 150 mm, was discovered during a clinical examination. The skin was endured, raised 6 cm with necrosis and an inflammatory peripheral halo, as well as an itchy rash (Fig. 1). The rest of the body exam was normal.

A right ankle X-ray revealed a cloudy soft tissue mass opposite the medial malleolus with no calcifications and slight demineralization of the various surrounding bone structures (Fig. 2).

Axial magnetic resonance imaging (MRI) DP, sagittal T1 FSE and DP, coronal DP and T1 FATSAT in the three planes with Gadolinium injection revealed a voluminous tumor process at the expense of the soft parts and opposite the internal malleolus of the right ankle, multi-partitioned, hypersignal DP and hyposignal T1 with hypointense levels in T2 and

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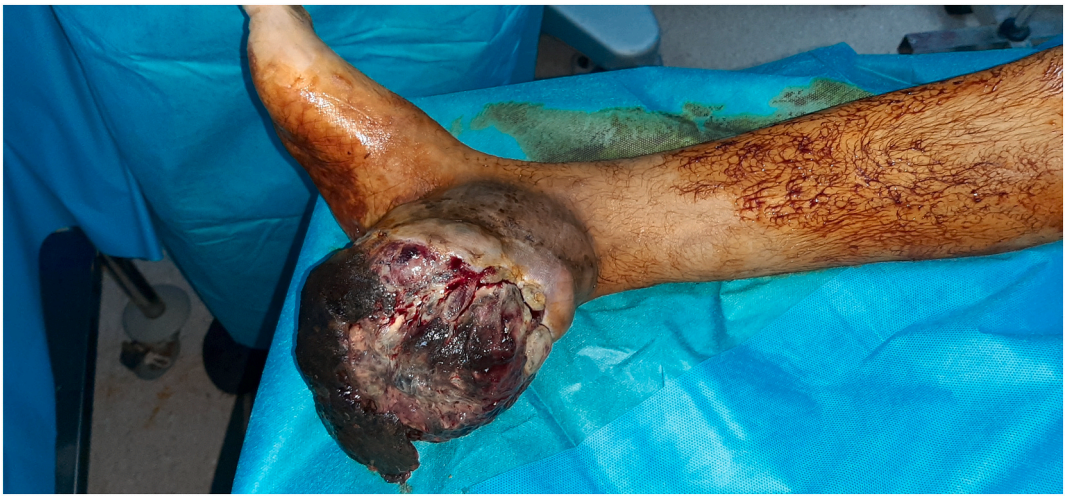

Fig. 1. Right leg with a large ulcerating tumor with evident tumor necrosis situated at the medial aspect of the right ankle joint.

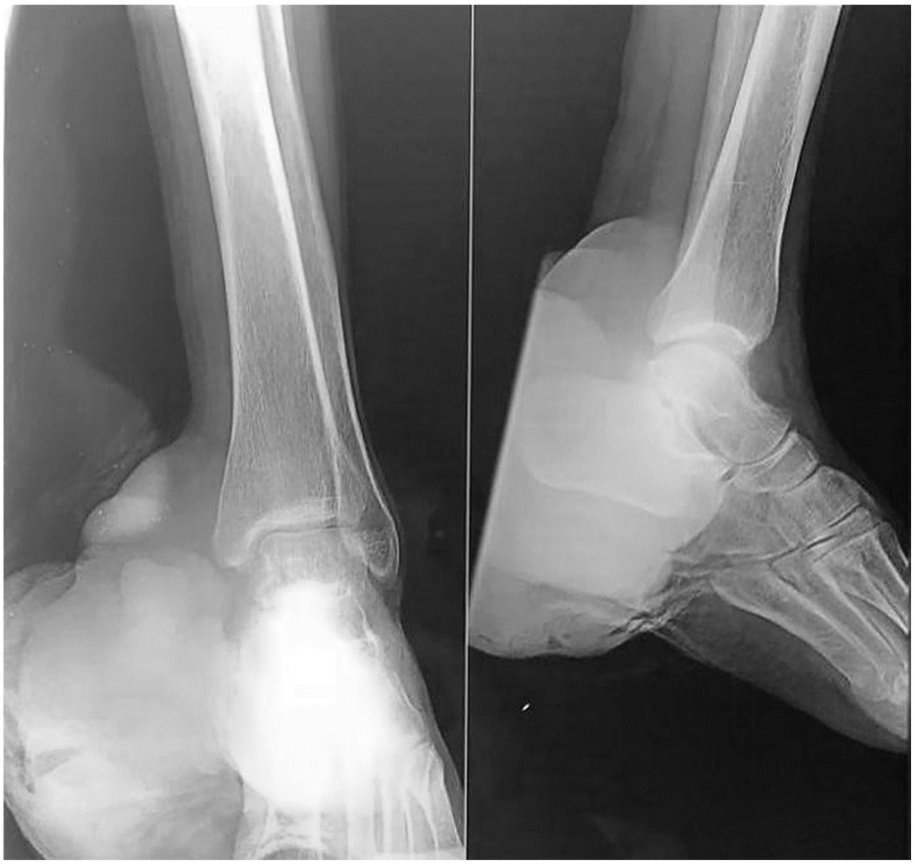

Fig. 2. A standard X-ray of the right ankle shows a tumor mass with no visible joint involvement.

isointense in T1, is enhanced after injection. It is lobulated and heterogeneous, overflowing and infiltrating the subcutaneous fat and buds at the opposite medial skin level. Furthermore, there are no abnormalities in the bone signal or musculo-ligamentous lesions related to the joint space (Fig. 3).

Faced with invasion of the posterior tibial pedicle, a CT angiography was ordered to thoroughly investigate a tumoral process and rule out a vascular origin of the lesion, specifically an arteriovenous malformation.

A surgical biopsy revealed a malignant spindle cell tumor infiltrating the dermis and hypodermis, as well as subacute and chronic ulcerative and suppurative inflammation, indicating poorly differentiated synovial sarcoma.

The abdominal ultrasound and computed tomography (CT) scans were both normal.

The patient came in 3 weeks after the biopsy in shock, with a deterioration in general condition caused by hemorrhage and suppuration of the mass. As a result, the patient underwent a distal trans-tibial amputation after the indication for radical treatment was raised. He benefited from a vascular filling and a blood transfusion after surgery. There were no complications during the post-operative period. The biopsy and the

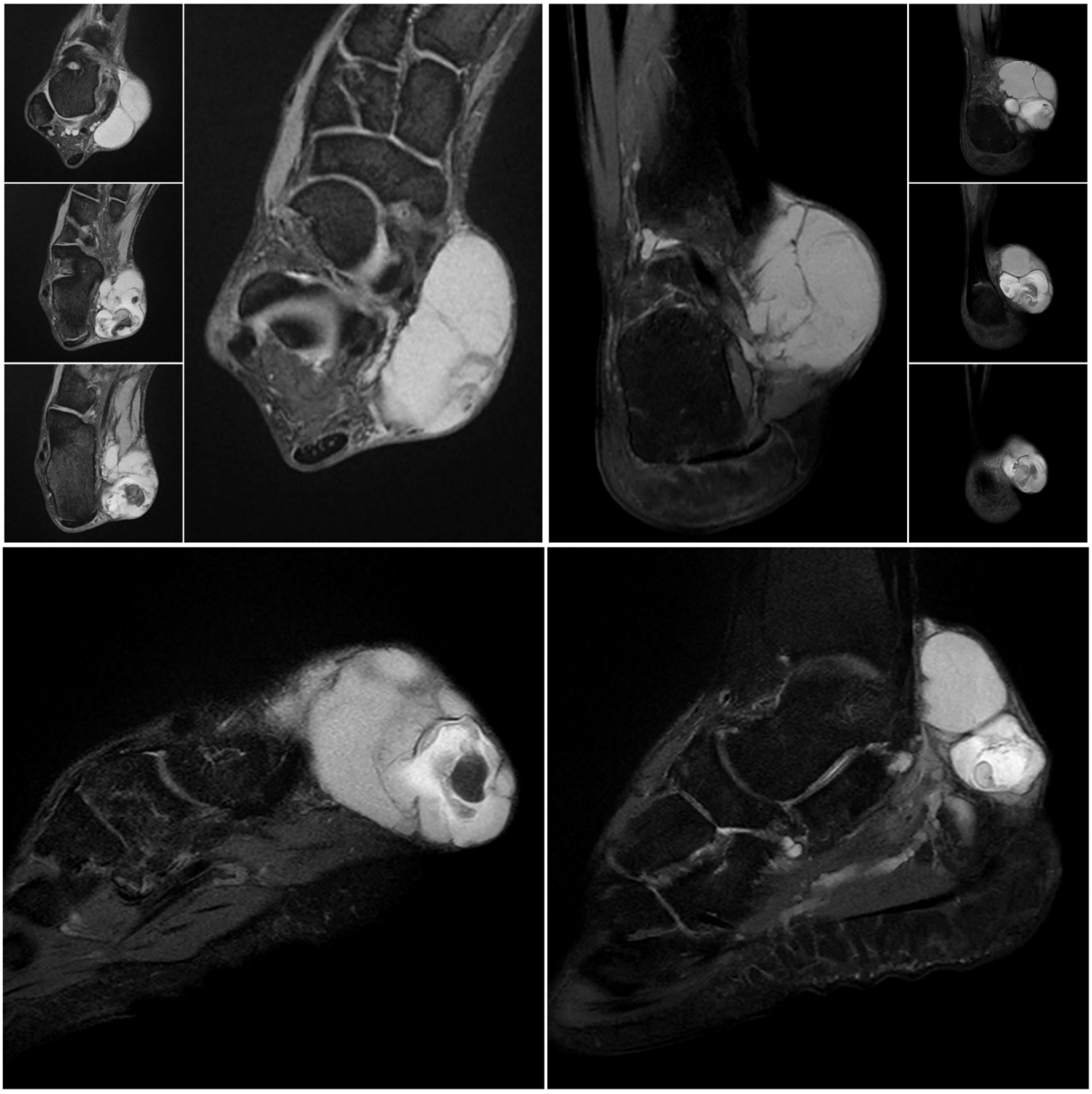

Fig. 3. MRI sections showing the multi-partitioned tumor mass at the level of the internal part of the ankle as well as its local invasion.

surgery were performed by an experimented surgeon from the department. Consent of patient was obtained and perspectives were painless limb and radical removal of tumor and infection.

At the last follow-up (2 years after surgery), abdominal ultrasound and computed tomography (CT) scans was performed and doesn't reveal any metastasis. The patient was pleased, and an examination of the stump revealed no signs of infection or other complications. He benefited from a walking prosthesis.

3. Discussion

Synovial sarcoma is currently defined as an aggressive primary mesenchymal malignancy that mimics the histological characteristics of synovial tissue. It accounts for approximately 8% of all soft tissue sarcomas [4], can occur at any age, with a slight male predominance, and can occur at any age (three men for two women). It is most common in young patients; 73% of cases are reported in patients aged 20 to 50, and 30% of patients are under the age of 20 at the time of diagnosis [2]. It is usually described as peri-articular, within tendons, bursae, capsules, and fasciae, but it is never a joint tumor, despite the fact that it is frequently classified as such [5].

In fact, it is derived from mesenchymal cells undergoing synovial differentiation or a tumor derived from synovial elements in the intermuscular fascia. 90% of synovial sarcomas are found in the extremities, with 60% found in the lower limbs; the three sites of predilection are the foot, knee, and thigh, with ankle localization being uncommon. The size of the lesion is primarily determined by its topography [2]. 85% of tumors are at least 5 cm in diameter, and 50% are in contact with bone, which is invaded in 21% of cases. The tumor's epicenter is within 7 cm of a joint in 63% of cases [5]. Signs of vasculo-nervous compression are less common, but they are more common in young adults. The average age is 25 years, and the majority of authors agree on male predominance [5].

Clinically, the tumor grows insidiously and asymptomatically; it most often manifests as a single nonspecific soft mass, most often deep and adherent to adjacent structures, slowly increasing in size despite its high malignancy, frequently mimicking a benign process, and becoming progressively painful, as was the case in our patient. In 35% of cases, the standard x-ray shows a mass of soft tissues of water density with

calcifications within it. Adjacent bone is affected in 20% of cases, either as a pressure response with osteosclerosis or as bone invasion with osteolysis and cortical erosions [5].

The best imaging tool for assessing neurovascular, bone, and soft tissue extension is magnetic resonance imaging (MRI) [1]. The appearance of synovial sarcoma on MRI is not specific, but the diagnosis is suggested by a mass of soft tissues that adheres to a tendon or a joint capsule [6]. The shape is usually round or multiloculate [7], and it is surrounded by a pseudocapsule that mimics a benign process. In T1, the signal of the mass is similar to that of the muscle, but it is heterogeneous in T2. Small synovial sarcomas typically have a homogeneous signal, leading to the incorrect conclusion that the lesion is benign; the presence of a lobulated character or septa is an element suggestive of the diagnosis, but the contrast enhancement is nonspecific [8].

The presence of a hemorrhagic component (40% of cases), liquid-liquid levels (10 to 25% of cases), and the presence of three signal intensities in T2 with saturation of the signal fat (blood, fluid, fibrosis, or calcifications) [2] are also suggestive of the diagnosis. Our patient had triple signal areas and liquid-liquid levels.

The treatment of synovial sarcoma is based on surgical resection, which must be carcinological and includes large, monobloc, and extra-compartmental resections. This requirement, according to our observations, implied a distal trans-tibial amputation. Radiotherapy is used to prevent amputation in cases of incomplete excision or local recurrence. The role of chemotherapy in patients with metastatic forms with a response rate of around 50% is discussed [9].

Synovial sarcoma has a poor prognosis, which is dominated by local recurrences and, in particular, early metastases. As a result, 30 to 75% of synovial sarcomas will develop pulmonary localization [10]. Even if the majority of deaths are caused by this attack, the prognosis is still bleak.

4. Conclusion

This case report describes a rare evolution of a giant and poorly differentiated synovial sarcoma in an unusual location. Management should be multidisciplinary, and treatment should be primarily surgical, with chemotherapy or radiotherapy used if excision is incomplete. Because of the large size and spread of the tumor in our case, we chose a radical treatment. The prognosis is determined by the timing of the treatment, its type, and the stage of the tumor.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Ethical approval

N/A. This study is exempt from ethical approval from the institution.

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Author contribution

Omar Fadili: study concept, data collection, analysis and interpretation, writing the paper.

Abdellah Chrak: managed the analyses, and the correction of the manuscript.

Mohamed Lafani: managed the analyses, and the correction of the manuscript.

Oussama El Adaoui: managed the analyses, and the correction of the manuscript.

Yassir El Andaloussi: managed the analyses, and the correction of the manuscript.

Mustapha Fadili: managed the analyses, and the correction of the manuscript.

Guarantor

Omar Fadili.

Research registration

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Declaration of competing interest

None.

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