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Bone Undifferentiated Pleomorphic Sarcoma – A Case Report

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Abstract

Bone sarcomas are a rare group of tumors arising mostly in the extremities, being the undifferentiated pleomorphic sarcoma (UPS), also known as malignant fibrous histiocytoma, the most common. UPS exhibits a wide spectrum of clinical behavior and a diagnosis challenge. We report a clinical case of a proximal humeral UPS that presented with a non-recognized pathological fracture, initially treated with open reduction and internal fixation. Suboptimal postoperative management of the patient, leading to a diagnosis delay, occurred related to its hematoid transformation, a variant of aggressive high-grade sarcoma associated with a worse prognosis. Pulmonary embolic complication, external axillary vein compression, rapidly tumor growth and diffuse metastization were some characteristics. Abrupt disease development makes important to recognize the clinical features of those tumors, so better outcomes can be obtained.

Keywords: Undifferentiated pleomorphic sarcoma; Pathological fracture; Shoulder

Introduction

Sarcomas account for less than 2% of all primary malignant bone tumors. These are a heterogeneous group of rare tumors arising in tissues of mesenchymal or ectodermal origin and may occur anywhere in the body [1,2]. About 50% to 60% occur in the extremities [3,4], with only 15% arising in the upper limb [2]. The most common sarcoma found in the extremities is the undifferentiated pleomorphic sarcoma (UPS), also known as malignant fibrous histiocytoma [3,5]. UPS is a high-grade and aggressive histiocytic lesion with no specific line of differentiation [6,7] and is common among adults between 60 and 70 years of age [8,9]. Definite diagnosis is made by histologic evaluation [5].

Case Report

A 72-year-old woman, with a previous history of hypertension, presented in the emergency room complaining of significant pain and functional disability of her right upper limb, following direct trauma after a fall from her own height, resulting in a two-part Neer proximal humeral fracture, as the x-rays (Figure 1) and CT scan (Figure 2) revealed. The patient was treated with open reduction and internal fixation (ORIF) with a locked plate and cemented screws - Philos Augmentation Synthes® (Figure 3), indicated for osteoporotic bone.

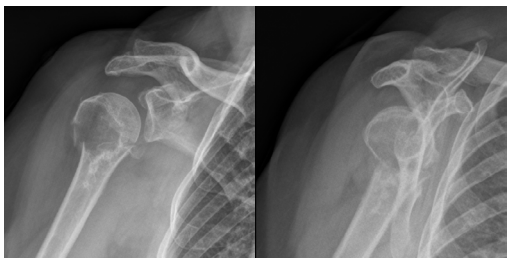

Figure 1: X-rays in the emergency room.

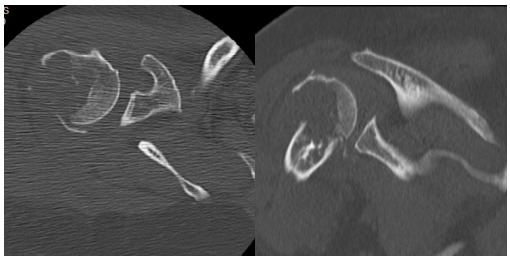

Figure 2: CT scan in the emergency room.

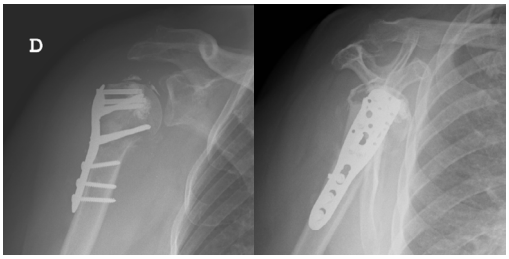

Figure 3: X-rays after open reduction and internal fixation.

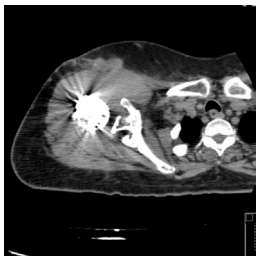

Figure 4: CT scan showing the periarticular soft tissue collection two months after fracture fixation.

In the postoperative period, despite intensive physiotherapy, the patient-maintained pain and swelling of the shoulder, with no associated inflammatory signs or incision drainage. Two months later, an articular CT scan showed a poorly delimited periarticular soft tissue collection involving the articular space, not previously present (Figure

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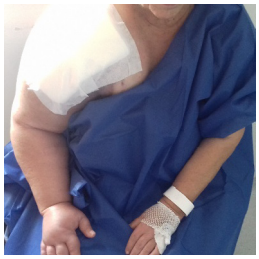

Figure 5: Picture of the affected upper limb.

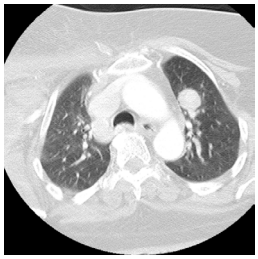

Figure 6: Isolated left lung superior lobe nodule.

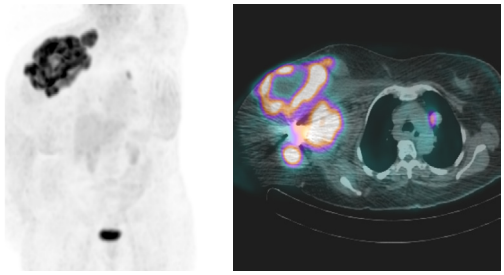

Figure 7: PET scan showing an intense metabolism in the right shoulder and left lung nodule.

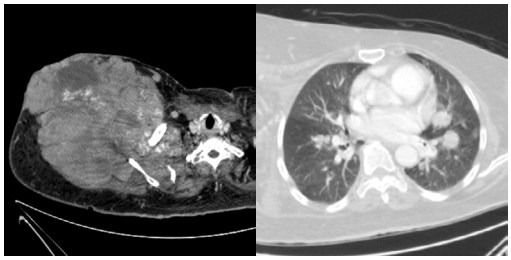

Figure 8: Restaging CT scan revealing local and disseminated disease progression.

4). At that time, with the suspected shoulder collection, the patient underwent an exploratory arthrotomy, debridement and periarticular tissue biopsy, being the main finding an extensive hematoma. During the surgery, the patient suffered a pulmonary embolism with full recovery. After this second procedure, shoulder pain and swelling intensified, progressing to a massive edema involving the entire upper limb (Figure 5). The postoperative chest CT angiogram, beyond the pulmonary embolism area, showed a 2 cm left lung nodule located on the superior lobe (Figure 6) and extrinsic compression of the right axillary vein by the shoulder soft tissue collection. These results arose the suspicion of a neoplastic etiology, but the workup that followed failed to identify any other primary location. The PET scan showed an intense glycolytic metabolism in the right shoulder and an 18-FDG avidity of the left lung nodule, supporting the hypothesis stated (Figure 7). Bronchofibroscopy and transthoracic needle biopsy of the lung were normal. The periarticular tissue biopsy report described a highly cellular smear, with pleomorphic spindle cells and scattered mitosis; associated multinucleated giant cells were present; but necrosis was not - these aspects supported an UPS diagnosis. One month later, during the hospital stay, the restaging CT scan revealed multiple bilateral lung lesions and axillary adenopathies suggesting disseminated disease (Figure 8). The patient was proposed for palliative measures and died a few days later.

Discussion

Extremities sarcomas are rare and challenging neoplasms [3]. In the latest World Health Organization classification, several groups of pleomorphic sarcomas were differentiated, and it is widely accepted that accurate histological subtyping in soft tissue sarcomas is clinically relevant [1]. Behavior is best predicted by the histologic grade, which is determined by the degree of differentiation, mitotic index, degree of cellularity, necrosis when present, vascularity and degree of nuclear anaplasia [3,4]. UPS is an unclassifiable category defined as a group of pleomorphic sarcomas in which any attempt to disclose their line of differentiation has failed [1].

UPS exhibits a wide spectrum of clinical behavior [4]. This patient presented to us with an initially non-recognized pathological proximal humeral fracture with an associated lytic component (Figures 1 and 2), whose malignancy was not suspected until a major embolic episode happened, which, coupled with the discovery of the left lung nodule, clarified the neoplastic etiology of this clinical picture. The most common site of metastasis are the lungs [7], and approximately 40% of patients with high-grade sarcomas develop pulmonary metastases even with surgically local tumor control [1]. According to Kontogeorgakos et al. metastatic disease developed at a mean time of 7 months in 57.3% of the patients [5]. Two months after initial presentation (pathological fracture) this patient had one lung metastasis; the following month, multiple bilateral metastases were evident, showing abrupt disease development. Significant tumor growth may lead to extrinsic compression of neighboring structures, as happened in this case. The large shoulder mass originated massive edema of the entire upper limb by axillary vein compression.

Conclusion

Sarcomas, usually present as a single, often large mass, with areas of necrosis and hemorrhage in their substance. The soft tissue sarcoma most commonly associated with hematoma formation is high-grade UPS [5]. Suboptimal patient management can occur when malignant tumors with internal hemorrhage masquerade as simple hematomas [10], and, for that reason, a delay in diagnosis is common [3]. The hematoid transformation in sarcomas is a distinct variant of aggressive high-grade sarcomas [11], associated with a worse prognosis [5].

Recognized UPS prognostic risk factors include histological grade, tumor size, depth and proximal location, all influencing survival and local recurrence [11,12]. Among these, histological grade and tumor size are widely considered the most significant. Large size predisposes to distant metastases [4]. Considering the very proximal upper extremity sarcoma and the presence of an isolated lung metastasis, forequarter amputation was the first-choice treatment for this patient, but the presence of nodal disease and diffuse metastatic disease on the restaging CT scan, obliged to a treatment plan redefinition, towards palliative measures only.

Bone tumors are not frequent in our daily practice, and sarcomas are even more rare. Nevertheless, it's important to recognize the clinical features of this type of lesions for a faster diagnosis and better outcomes.

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