CASE REPORTS

Undifferentiated Pleomorphic Sarcoma. From Supposition to Certainty: a Case Report

Mihai MEHEDINTU-IONESCU, Gabriela ALEXA, Horia TOADER, Costinela COCIU

Abstract

Second Opinion is a difficult problem for every doctor put in this situation. In this position, we must not let ourselves be influenced, in any way, by the judgment of the first clinician, who formed a diagnosis. But not judging the "evidence" that led to the formation of the first diagnosis is difficult. This case presentation reveals the influence of a diagnosis assumed to be correct from the first interpretation. The patient, from the rural area, presents herself to the doctor following a trauma from falling down the stairs. A soft tissue ultrasound is performed and it is interpreted to be a hematoma. Afterwards, the lesion stagnates in size without progressing towards resorption. After repeating the ultrasound, its interpretation is influenced by the first ultrasound investigation supporting the same diagnosis. As a result, the patient is not guided to make a radical therapeutic decision. The presentation in our clinic was decided by the patient for a "second opinion" and, as a result, all the investigations carried out did not take into account the results previously stated by the patient during the clinical examination in the specialized outpatient clinic. The tumor was one with unimpressive dimensions but disturbing in appearance, without influencing the functionality of the forearm. The patient does not complain of pain or paresthesia in the area occupied by the tumor. She was clinging to deep plans and immovable in front of them.

Keywords: tumor, pleiomorph, soft tissue ultrasound, second opinion.

Rezumat

Second Opinion este o problema dificila pentru fiecare medic pus in acesta situatie. In aceasta postura nu trebuie sa ne lasam influentati, in nicio masura, de judecata primului clinician, ce a format un diagnostic. Insa a nu judeca "probele" ce au dus la formarea primului diagnostic este dificil. Acesta prezentare de caz releva influenta unui diagnostic presupus a fi corect inca de la prima interpretare. Pacienta, din mediul rural, se prezinta la medic in urma unui traumatism prin cadere pe scari. Se efectueaza o ecografie tesuturi moi si se interpreteaza a fi un hematom. Ulterior, leziunea stagneaza ca dimensiuni fara a avea o evolutie spre rezorbtie. Dupa repetarea ecografiei, interpretarea acesteia este influentata de prima investigatie ecografica sustinand acelasi diagnostic. Drept urmare, pacienta nu este indrumata spre a lua o decizie terapeutica radicala. Prezentarea in clinica noastra a fost decisa de pacienta pentru un "second opinion" si, drept urmare, toate investigatiile efectuate nu au luat in considerare rezultatele anterior enuntate de catre pacienta la examenul clinic in ambulatoriul de specialitate. Tumora era una cu dimensiuni neimpresionante dar care deranja ca aspect, fara insa a influenta functionalitate antebratului. Pacinta nu acuza dureri sau parestezii in teritoriul ocupat de tumora. Era aderenta de planurile profunde si imobila fata de acestea.

Cuvinte cheie: tumora, pleiomorf, ecografie parti moi , second opinion.

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INTRODUCTION

Soft-tissue sarcoma represent a group of rare, high-grade malignant tumors that originates in mesenchymal stem cells population and presents similar phenotype to fibroblast cells rather than histiocyte, contrary to what was thought in previous decades, before using immunohistochemistry and monoclonal antibodies in their documentation.[1,4]

According to the multitude of histologic subtypes and molecular patterns, they often show heterogenic clinical and prognostic features, which leads to unique treatment solutions, requiring multidisciplinary team approach, involving oncologists, orthopedic and plastic surgeons, radiologists and pathologists.[5]

The most common histologic subtypes of soft-tissue sarcoma include liposarcoma, synovial sarcoma, leiomyosarcoma and undifferentiated pleomorphic sarcoma (UPS), whilst approximately 50 % of their total occur in the extremities, followed by the trunk. Is characteristically a tumor of late decades of life, with two-thirds occurring in white males, diagnosis in children being extremely rare.[2,3,6]

Undifferentiated pleomorphic sarcoma, formerly known as malignant fibrous histiocytoma, originates frequently in deep tissues of the extremities, it is characterized by aggressive tumoral behavior, early local recurrency and high distant metastatic rates, frequently in the first year since primary diagnosed.[4,5,12]

Imaging differential diagnosis, with ultrasound as the first-line investigation and MRI, is crucial for proper therapeutic management and surgical approach of UPS.[7] It is documented that preoperative suspicion of sarcoma by recognizing "red flag" symptoms and clinical signs, followed by ordering advanced imaging analysis showed significant association with complete excision of the tumor.[9,11] Pseudo-sarcomas that may mimic a UPS regarding clinical features and imaging appearance are mainly represented by myositis ossificans, synovial chondroma, elastofibroma, nodular fasciitis, hematoma, infection.[7,8] Identifying malignant or benign nature of STS proves to be challenging by imaging alone given their nonspecific appearance, therefore, biopsy tissue sampling guided by imaging findings represents the gold standard for definitive diagnosis before surgical approach.[11]

Prognosis depends on tumor size and location, distant metastasis and grade assessment by morphology, necrosis and mitotic activity analysis in the FNCLCC grading system. General 5-year survival is documented around 65-70%, with increased, metastatic-free survival for UPS located in limbs or trunk.[4,12]

PATIENT AND OBSERVATION

Clinical findings

A 60 years old female patient referred to our clinic with a rapidly enlarging tumor after repeated trauma to the elbow region. The clinical exam revealed a painless firm mass located on the extensor digiti muscle, adherent and with no mobility in relation to the deep plane tissues. There was no limited ability to extend the digits at the moment of the assessment.

Diagnostic assessment

All routine investigation were done, early imagistic findings pointing to a hematoma as a diagnosis. Two ultrasounds and an elbow MRI were performed.

First ultrasound showed a 1,12 cm X 1,10 cm X 0,707 cm resorbing hematoma, close to the joint, without a Doppler vascular signal. Three months apart the MRI came with the same conclusion, describing a larger collection, 1,5X 3 cm, with modified fluid signal, suggesting a resorbing hematoma localized partially in the deep fat subcutaneous tissue and partially intramuscularly, on the level of extensor carpi ulnaris muscle, having moderate mass effect on extensor digitorum muscle. The ultrasound performed before surgery described a 3 X 2X 4 cm tumor developed in the thickness of common extensor muscle, multiloculated, extended in the subcutaneous tissue, with peripheral vascularization and contact with radial brachii muscle and radial nerve.

THERAPEUTIC INTERVENTION

Corroborating clinical findings with imaging results, we decided to perform the excision of the tumor under regional anesthesia (peripheral nerve block). Intraoperative evaluation of the tumor revealed it was incapsulated in the extensor digitorum communis muscle. While attentively dissecting the tissues, we couldn't observe any important vascular or nervous elements affected. The tumor was excised and redirected to the pathological anatomy department.

Histopathological exam determined the nature of the tumor as undifferentiated pleomorphic sarcoma, grade III FNCLCC, without lymphatic or vascular

invasion, whilst immunohistochemistry studies didn't identify any specific tumoral markers.

DISCUSSIONS

The clinical characteristics of the investigated mass, firmness, adherence and lack of mobility in relation to deep plane tissues, suggests a high probability for a malignant lesion.

Considering the ultrasound and MRI presented at admission, revealing a resorbing hematoma, we can rise the suspicion of an interpretation error by lack of clinical and imaging exams corroboration, that would have excluded this diagnosis.

Although the lesion is described by the patient as a consequence of repeated trauma by falling from the same height, there was no associated pain, nor alteration on the appearance of the muscular plane and no limited ability on active or passive mobility of the area. This, in corroboration with the MRI exam , which would have shown calcified masses in the muscle plane, excludes myositis as a diagnosis with high certainty. With inflammatory biologic markers being negative, we can also exclude myositis ossificans as a diagnosis.

All clinical findings were suggesting a malignant tumor, so the surgical intervention could've been preceded by biopsy. This approach would've postponed surgery with at least 21 days and would've had a negative impact on survival. The decision to pursue surgical management is made in agreement with the patient, meaning excision of the tumor while preserving function, sensitivity and architecture of the involved area.

CONCLUSIONS

From the beginning of our case investigation, firmness and adherence of the investigated mass suggested malignancy.

All imaging exams performed before referring to our clinic followed the evolution of a post-traumatic lesion, assuming it to be a hematoma.

Exclusion of differential diagnosis by corroborating imaging and clinical findings would have saved time for early therapeutic conduct.

Histopathological diagnosis has multiple therapeutic limitations, leading the patient towards radical surgery with amputation and recovery with prosthesis or palliative care, that aims preserving life quality.

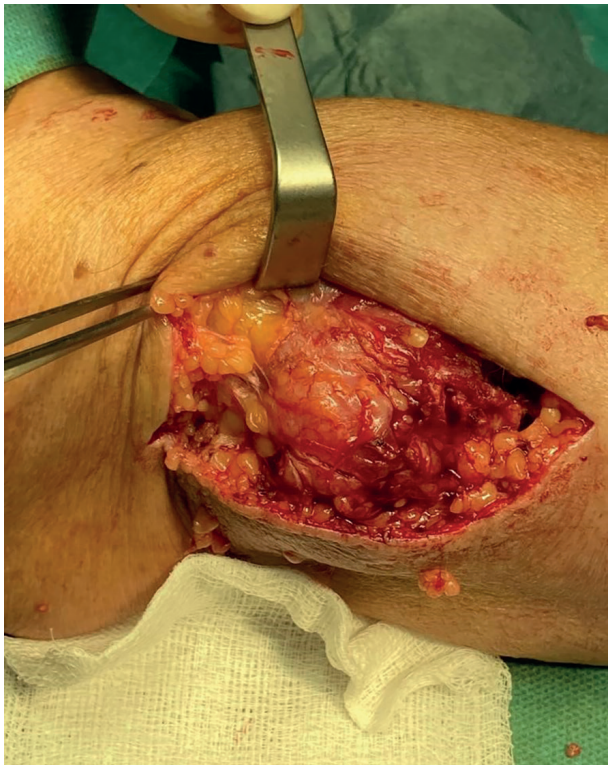

Figure 1. Tumor identification

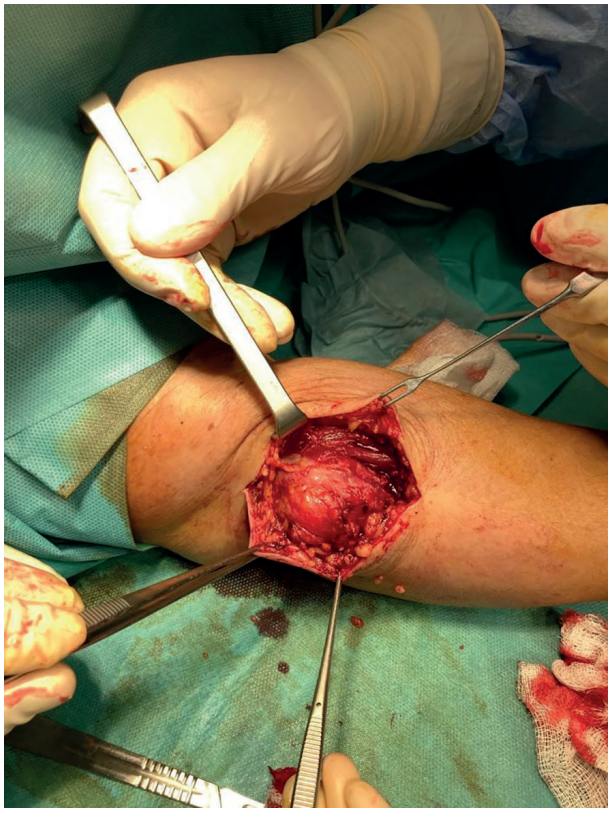

Figure 2. Tumor dissection

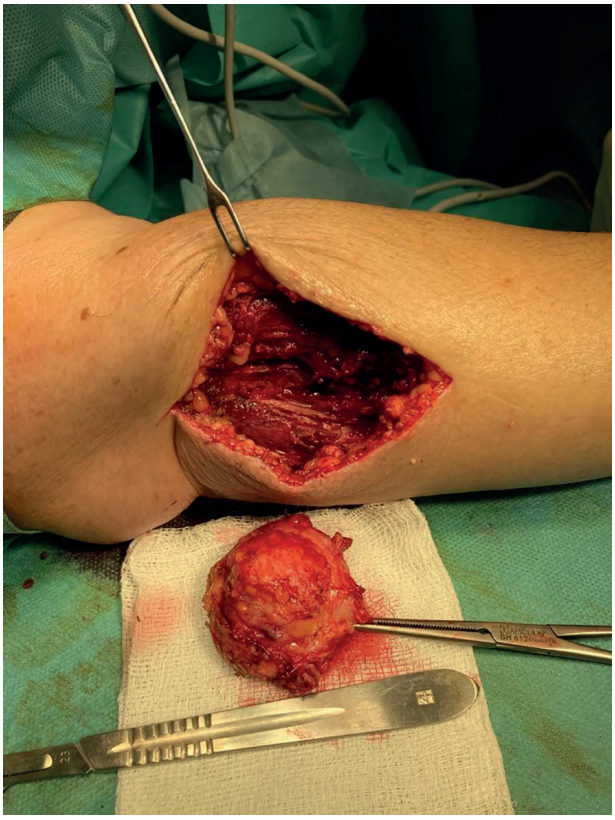

Figure 3. Frame sizes

All individuals who meet the authorship criteria are listed as authors, and all authors certify that they participated sufficiently in work to take public responsibility for the content, including participation in the concept, design, analysis, writing, or revision of the manuscript. Each author certifies that this material has not been and will not be submitted or published in any other publication. The authors' names listed below attest that they have NO affiliation or involvement with any organization or entity with any financial (such as honoraria; scholarships; employment; consulting, etc.) and non-financial (such as professional or personal) interests, relationships, affiliations, etc.) in the topic or materials discussed in the manuscript. All the medical deontology outlined in the legislation was respected and all the clinical elements of the patient respected the protocols in force in the clinic where the medical act took place. The patient was informed of the possibility of exposing the case in an article.

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