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Sarcoma the great “masquerader” hematoma/deep vein thrombosis manifestation



J. Valverde*, M. Vinagreiro, P. Gouveia, P. Koch, V. Soares, T. Gomes

Rua Dr. Eduardo Torres, 4464-513 Senhora da Hora, Portugal

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ABSTRACT

INTRODUCTION: The clinical presentation of patients with soft-tissue sarcoma is highly variable. Most patients present with a painless mass, typically one that is increasing in size, and few have systemic symptoms such as fever, weight loss, or malaise. Soft tissue sarcomas can initially present as, or even be misdiagnosed as, deep venous thrombosis (DVT), leading to a late diagnosis.

CASE REPORT: A 51-year-old woman presented to the hospital with complaints of pain and swelling in her left thigh, interpreted as an infected hematoma with an associated deep vein thrombosis and treated accordingly. The patient presented to our emergency department two more times. In the last visit and due to an unresolving clinical scenario a MRI and surgical biopsies were made that confirmed a sarcoma diagnosis.

DISCUSSION: When a patient presents with an expanding, nontraumatic mass simulating a haematoma, several other differential diagnoses should be considered including aneurysm, bleeding tendency, chronic expanding haematoma and soft-tissue sarcoma. The growth of the tumor undetected while being treated for the DVT and then posteriorly for the hematoma, was without a doubt dismal to the patient, so earlier diagnosis would have been preferable.

CONCLUSION: When a patient presents with an unusual history of hematoma in the extremities, it is necessary to consider the possibility of a malignant soft tissue tumor.

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1. Introduction

Sarcomas are neoplasms arising from connective tissue elements of the body. Approximately 80% arise in soft tissue, while the remainders originate in bone. Soft tissue sarcomas can develop from fat, muscle, nerves, fibrous tissues, blood vessels, or deep skin tissues. Almost 55% of all soft tissue sarcomas are found in the extremities, especially in the lower limb [1,2].

The incidence of soft tissue sarcoma represents approximately 1% of all malignancies [3]. About 10,600 new cases of soft-tissue sarcoma and 3800 deaths were estimated to have been occurred in the United States in 2009 [4].

There are predisposing factors that make a person more likely to develop soft tissue sarcomas like genetic syndromes (neurofibromatosis, Gardner syndrome, Li-Fraumeni syndrome, retinoblastoma), radiation exposure and damaged lymph system [5].

The clinical presentation of patients with soft-tissue sarcoma is highly variable. Most patients present with a painless mass, typically one that is increasing in size, and few have systemic symptoms

such as fever, weight loss, or malaise. Though not common, pain or tenderness can occur and, when it does, the possibility of malignancy is greater [6]. Soft tissue sarcomas can initially present as, or even be misdiagnosed as, deep venous thrombosis (DVT), leading to a late diagnosis. Spontaneous intramuscular hematomas in the extremities are very rare and should be approached with a high degree of clinical suspicion.

There are 3 pitfalls: in some cases of sarcomas a history of trauma is reported by patients, spontaneous hematoma could reveal high grade sarcomas, and hematomas, in some instances, can develop slow expansion.

Poor prognostic factors in soft-tissue sarcomas are primary metastasis, high-grade tumors, several histological entities, tumor of head and neck or deep trunk, large size, resection margin positive or uncertain and older age [7,8].

We report on one patient with soft-tissue sarcoma of the thigh initially diagnosed and treated as hematoma. A detailed study of the patient's history, clinical course, and diagnostic images, as well as the discussion of the clinical features of soft-tissue sarcomas mimicking hematomas are presented to facilitate the correct diagnosis.

* Corresponding author.

E-mail address: jorgenunovalverde@gmail.com (J. Valverde).



Fig. 1. - CT scan – 15 x 6 x 10 cm cystic lesion.



Fig 2. - CT scan – 15 x 6 x 10 cm cystic lesion.

2. Case report

A 51-year-old woman presented to the hospital with complaints of pain and swelling in her left thigh. The patient had a history of three previous episodes of deep vein thrombosis eight months before presenting to our emergency department (ED), since then, she has been on oral anticoagulation therapy. The patient had no previous history of trauma or bleeding disorder. There was no family history of venous thromboembolism.

On physical examination, a tender, firm swelling with some redness and local warmth was found in the patient's left thigh. Blood coagulation was normal. Computed tomography scan of the lower limbs demonstrated thrombosis of the left popliteal vein in the adductor canal with a large luminal thrombus, and revealed a cystic 15 × 6 × 10 cm lesion, filling with a homogeneous fluid, in the deep posterior muscle compartment of the lower thigh, without septums and without solid vascularized components, suggesting a para-articular cyst (Figs. 1 and 2).



Fig. 3. (CT scan).

Due to the persistence of symptoms, the patient was taken to the operating room for hematoma evacuation. Nine days later she left the hospital restarting the oral anticoagulation therapy.

However, the patient returned three weeks later with worsening of the swelling and pain. INR test was high (4.3). She subsequently underwent antibiotherapy and hematoma evacuation surgery. The specimens were sent to pathologic analysis. She left the hospital under therapeutic doses of subcutaneous enoxaparin (80 mg/day).

Five days later the patient was admitted to the hospital reporting the same symptoms and was submitted to a third surgery, with fasciotomy, to remove the hematoma. Subsequently, a wound vacuum assisted closure (VAC) was placed for drainage. Microbiological analysis was negative. Magnetic resonance (MR) imaging demonstrated an extensive mass in the posteromedial muscle compartment of the left lower thigh, measuring 20 × 18 × 11 cm, filling with blood and necrotic tissue, extending into the anterior and external compartment. The lesion surrounded the femur without evidence of bone infiltration but some periosteal reaction was observed. The mass was in close proximity to the deep femoral vessels and compressed the superficial vessels. The lesion was associated with multiple lymphadenopathy in the homolateral inguinal region. This was the first time a diagnosis of sarcoma was suggested (Fig. 3).

The presence of distant metastasis was evaluated with thoracic, abdominal and pelvic computed tomography scan and, although there was no evidence of distant metastasis, there were multiple enlarged lymph nodes in the inguinal and obturator regions with suspicious features.

The histologic examination revealed a spindle cell pattern composed of cells with large, irregular and pleomorphic nuclei, with high mitotic rate and atypical mitosis. On immunohistochemistry, the tumor was positive for vimentin, α -smooth muscle actin, h-caldesmon and S-100 protein, and was negative for CD34, CD117 and AE1/AE3. A diagnosis of leiomyosarcoma was made based on these histological findings.

The patient was then transferred to Portuguese Institute for Oncology where chemotherapy treatment and thigh amputation were preformed.

3. Discussion

Leiomyosarcomas originating from peripheral veins, including saphenous, femoral, iliac and popliteal veins, have been sporadically described [9].

In the present case, the most significant problem was diagnostic delay, resulting in delayed treatment. When reviewing the case and stratifying the initial DVT risk according to SAGES, this was a woman of 55 years of age, obese (BMI > 35), without any other risk factors. If the Wells Clinical Score is taken into account, the patient had no risk factors.

Painful swollen leg is a common clinical scenario for a wide range of pathology. The patient presented herself in the emergency department, with three previous diagnosis of DVT, under warfarin (since the first diagnosis), with a localized painful swelling of the thigh, without any trauma history and with a normal INR. A CT scan was made and a thrombus of the popliteal vein with an associated hematoma was the proposed diagnosis.

The initial management in the majority of cases is to start anticoagulation and proceed to a venous duplex scan, as the priority is to rule out a DVT. One must keep in mind that only one third of the first episodes of a venous thrombosis are spontaneous [10].

When a patient presents with an expanding, nontraumatic mass simulating a haematoma, several other differential diagnoses should be considered including aneurysm, bleeding tendency, chronic expanding haematoma and soft-tissue sarcoma [11].

Tumours are a rare but important differential diagnosis in such patients. Sixty percent of soft tissue sarcomas arise in the extremities, 70% occur in the lower limb and mostly in the thigh. As a rule of thumb any mass over 5 cm in size arising beneath the level of deep fascia should be considered a sarcoma unless proven otherwise [12].

In retrospective, probably the first manifestation of the leiomyosarcoma was the initial DVT, but as the patient was observed in another institution data is lacking.

The first time she was evaluated by our team in the emergency department this differential diagnosis (sarcoma) wasn't considered. The patient was examined, a CT reported a thigh hematoma with a thrombosis of the left popliteal vein, with fever and an increased white blood cell count, the treatment offered was as if we were treating an infected hematoma. After being discharged the patient returned, 3 weeks later, this time with altered coagulation, which could justify the thigh hematoma recurrence, a surgical drainage was repeated. The non-resolving hematoma and the growing thigh lump raised a red flag and tissue was sent to be examined as well as a MR imaging was made. Preoperative examinations such as CT and MR imaging sometimes cannot easily distinguish a tumor thrombus from a blood clot [13]. MRI has become a powerful tool for diagnosing bone and soft-tissue masses because it shows the location of a mass and offers clues regarding intralésional characteristics such as myxoid change, age of the haematoma [14,15], and amount of necrosis [16]. However, diagnosis can be challenging when a significant portion of the tumor is occupied by hematoma without a solid mass lesion [17] as was the case with this patient in an early stage.

The obtained diagnosis was a soft tissue sarcoma, more specifically a leiomyosarcoma. Large series have showed disease-specific survival rates for extremity soft tissue sarcoma of 73–79% at 5 years [18–20]. Independent prognostic factors include age >50 years, histologic grade, tumor size >5 cm, deep vs superficial tumor, proximal vs distal location, and positive microscopic margins on resection

[18–20]. Of these factors, high grade and large size are the primary predictors of mortality [21].

The growth of the tumor undetected while being treated for the DVT and then posteriorly for the hematoma, was without a doubt dismal to the patient, so earlier diagnosis would have been preferable.

4. Conclusion

When a patient presents with an unusual history of hematoma in the extremities, it is necessary to consider the possibility of a malignant soft tissue tumor. Investigate the patient's history of trauma, bleeding disorders, anti-platelet or anticoagulation medication, clinical course, and MRI findings. Moreover, prompt biopsies are recommended to facilitate correct diagnosis. This article serves a purpose that is to once more, alert to the caveat of the soft tissue tumor masquerading as a "spontaneous" hematomas and/or DVT and the potential severe consequences of a late diagnosis.

Conflict of interest

No conflicts of interest.

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Ethical approval

This article was approved by the general surgery department, and clinical director of Hospital Pedro Hispano.

Consent

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

Author contributions

Jorge Valverde – Author.
Margarida Vinagreiro – Co-author, revision.
Pedro Gouveia – Revision and approval.
Pedro Koch – Revision and approval.
Virginia Soares – Revision and approval.
Taveira Gomes – Revision and approval.

Guarantor

Jorge Valverde.

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