



Synovial Sarcoma of the Right Hand. Case Report

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Abstract

Synovial sarcoma presents as a well-defined entity in its clinical and morphological conditions, its articular location is uncommon, it affects areas with no apparent relation to synovial structures, and it accounts for 5 to 10% of soft tissue sarcomas. Appearing in 66% of cases in soft tissues of the extremities, with a higher incidence in the lower ones close to the large joints, the location in the hands and feet is more infrequent and presents a favorable clinical evolution, appearing between 15 and 40 years of age. We present the case of an adult woman with a tumor mass in the palm of her right hand who underwent an excisional biopsy that reported a diagnosis of synovial sarcoma, closure by second intention and radiotherapy.

Keywords: Synovial Sarcoma (SS); Hand; Immunohistochemistry

Introduction

Synovial sarcoma presents as a well-defined entity in its clinical and morphological aspects, its articular location is infrequent because it affects areas with no apparent relationship with the synovial structures [1-4]. These affected areas are most commonly located in the extremities in 60%-70% [1,4,5] with a close relationship to the sheaths, bursae and joint capsules are rare in the hands and feet [3].

In the first descriptions of the sarcoma, its histological similarity to the developing synovium was pointed out, but there is no evidence that this tumor originates from the synovium, nor that it differs in this regard. The first report of this entity was made by Simon [6-8]. In 1927 Smith used the term synovium [3], while Lejars and Rubens-Duval [3] preferred synovial endothelioma. In 1934 Sabrazes used the term synovial sarcoma [6,7], It is now known that the immunohistochemical and ultrastructural differences between synovial sarcoma and normal synovial sarcoma are such that most authors consider the name synovial sarcoma to be a name derived from the first published works, and that its name should be changed.

Synovial sarcoma accounts for 5% to 10% of all soft tissue sarcomas [3-6,9] and is most common in adolescents and young adults aged 15 to 40 years, with an average age of 34 years, is more common in men than women, with a male/female ratio of 1.2:1, No predilection for a specific breed [3]. Other authors argue that there is no predominance in terms of sex [6]. This is a rare mesenchymal neoplasm. It is the fourth most common soft tissue tumor after liposarcoma, malignant fibrous histiocytoma and rhabdomyosarcoma [9]. It is characterized by a translocation of the X chromosome and 18, its name is a historical accident since the tumor does not originate from or differ from the synovium [4].

- **Clinical:** The most typical form of presentation consists of the appearance of a painless juxtaarticular mass or with mild pain, occasionally with many months of evolution until its definitive diagnosis (up to more than a year) [4-6,9]. In some cases, a history of trauma has been reported in 20-25% of cases [9].
- **Radiology:** Plain radiography is suitable for assessing the condition of the adjacent bone. Computed Tomography and Magnetic Resonance Imaging are essential in determining the

location of the tumor in relation to fascial planes, bones, vessels, nerves and organs. PET scan (positron emission tomography) is a staging technique [9,10].

- **Macroscopically:** It is characterized by a mass between 3 and 10 cm in diameter, it is usually circumscribed and can be multilobular or multicystic [4].
- **Histologically:** They are divided into two categories, monophasic and biphasic. The biphasic has both spindle and epithelial cells and the monophasic has only spindle cells or exceptional epithelial cells [3-6,11].

The treatment is multidisciplinary by a team of experts in various techniques, in a reference center. The biopsy will be performed by an experienced surgeon and should not interfere with the final surgery. Radiotherapy treatment is very important, it can be post-operative, intraoperative or preoperative (when the tumour cannot be removed because it is too deep or in a location that makes it inoperable, when the margins obtained after surgery are not sufficient or to favour the operation in a second time) [10].

Synovial sarcomas are treated aggressively with limb-sparing surgery and often chemotherapy. Survival at 5 years ranges from 25 to 62% and only 11 to 30% live 10 years or more. Metastases are most often located in the lung, bone, and occasionally lymph nodes [5].

We propose to present a case of synovial sarcoma located in the palm of the right hand and to conduct a discussion on the subject.

Presentation of the case

A 57-year-old female patient, with a history of controlled arterial hypertension, presented a small mass at the level of the palm of the right hand proximal between 4 and 5 fingers, painless, slow-growing, she was treated in her health area where a biopsy was performed, which reported palmar fibromatosis, presented recurrence of the lesion and arrived at our center where she was admitted for study and treatment.

Physical exam

A lobular mass in soft tissues of approximately 5cm located in the palm of the right hand, irregular with a firm consistency, with slight local heat and darker in color than the rest of the hand, small ulcerated central lesion, with limitation of finger movements (Figure 1a and 1b).

Presence of soft, mobile, non-painful axillary lymphadenopathy.

Sensitivity preserved.

It has no collateral circulation.

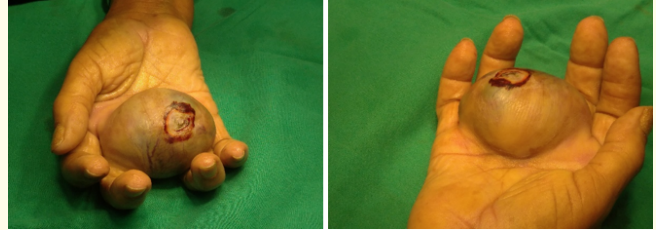


Figure 1: a Caudal view. b Cephalic view.

Lab tests

Hemoglobin 10.8 x 10⁹/L
 Alkaline phosphatase: 133 U/L
 Hematocrit: 0.31%
 Rheumatoid factor: 35 IU/ml
 Leukocytes: 7.8x10⁹ /L
 Serology (VDRL): non-reactive
 Platelets: 563x10⁹ /L
 HIV: negative
 Glicemia: 4,8 mmol/L
 Creatinina: 115 umol/L
 Erythrocyte sedimentation rate: 83 mm/h
 TGP: 9 A/L
 PCR: 21mg/L
 TGO: 17 U/L

Imaging studies

Plain x-rays of the anteroposterior and lateral right hand were performed, as well as a chest x-ray.

Computed tomography (CT) scan of the right hand: a mass of well-defined heterogeneous structure of 5 x 5 cm is visualized that extends on its inner surface to subcutaneous cellular tissue and fat, no bone alterations with density of up to 27 U are observed.

T.A.C of the thorax and abdomen: no pathological mediastinal lymphadenopathy, no pericardial or pleural effusion, no acute pleuropulmonary lesions.

Spondyloarthritis in the dorsal spine, not secondary bone lesions. Lumbosacral spine spondyloarthritis with protrusion of the disc between L5/S1 causing compression of the dural sac, slight lumbar scoliosis.

No secondary lesions to the liver, adrenal, or kidneys. No abdominal or inguinal lymphadenopathy.

Abdominal ultrasound: no metastatic lesions were present.

Scintigraphic bone survey: increased uptake of the radiopharmaceutical in the right hand and diffuse in the proximal metacarpophalangeal and interphalangeal joint of 3 and 4 right fingers. (Radiopharmaceutical: technetium ⁹⁹ MDP).

Conduct

Excisional biopsy of the lesion and closure by second intention with the use of pigskin as a biological dressing (Figure 2a, b, c).

Anatomical pathology result: (2020-B169) Compatible with biphasic synovial sarcoma with areas of necrosis and hemorrhage, very close to the area of deep section.

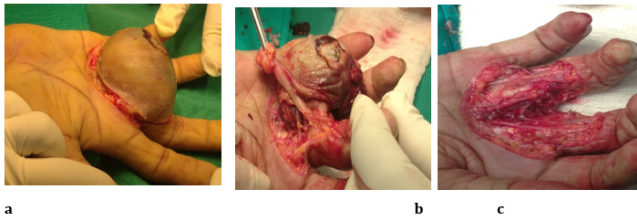


Figure 2

After the healing of the lesion, an interconsultation with Oncology is carried out to evaluate the possibility of radiant treatment, which is carried out in this institution, applying cobalt therapy in 25 sessions, evolving with secondary injuries due to burns to this procedure that were treated with local dressings, healing completely (Figure 3 a, b).

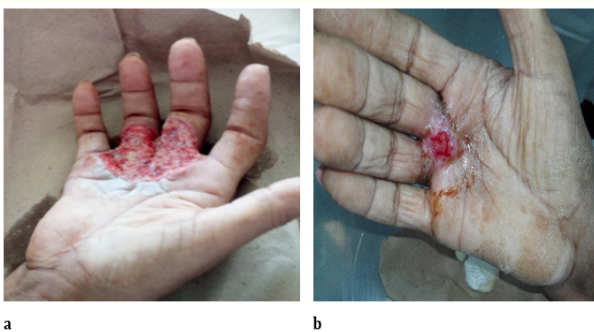


Figure 3

Once the lesion healed, the patient was referred to the Rehabilitation Service to achieve the extension of the fingers and wrist and fine movements of the hand with kinesiotherapy and Occupational Therapy, Phonophoresis with sodium heparin cream was used to treat post-surgical fibrosis and soft tissue retraction.

Image of the hand after radiation and rehabilitation treatment (Figure 4 a, b).



Figure 4

Discussion

Synovial sarcoma does not originate, as its name suggests, from synovial tissue, it presents uncertain differentiation lacking counterpart precision from normal tissue, its name comes from the first reports in the literature, due to its frequent pararticular location and microscopic similarities with developing joint tissue. The clinic, morphology, and genetics of synovial sarcoma are different from other sarcomas [12].

Synovial sarcoma can be classified into two subtypes: monophasic and biphasic. Recently, a third one has been described, which is poorly differentiated from the histological point of view [13]. The biphasic SS subtype presented by the patient is composed of a biphasic cellular structure formed by indentations or acinous structures and lined with cells with an epithelioid appearance with mucoid material formation, fusiform and epithelial [1,3].

The pre-treatment and post-treatment staging of the surgical patient defines the treatment strategy. The classification of clinical TNM allows differentiating stages of SS according to local invasion, tumor size, absence or presence of lymph node metastases, and distant metastases [13].

- Group I - Tumors are completely removed with microscopic negative margins
- Group II - Wide resection of tumors with microscopic residual disease and/or regional lymph node spread.
- Group III - Gross residual disease after incomplete resection or biopsy.
- Group IV - Onset metastasis [13,14].

The prognosis of patients with SS is related to the possibility of complete removal, the size of the tumor, and local invasion. Patients with small tumors that can be completely removed at the time of diagnosis have an excellent prognosis. For tumors larger than 5 cm, the risk of developing distant metastases is higher. The Paediatric series (Ladenstein 1993, Pappo 1994, Ferrari 1999, Okcu 2003, Brecht, in press) observed an 80% survival rate for patients in group I-II of the IRS, but about 60-70% of cases > 5 cm. The survival rate of patients with tumors that are unresectable at the time of diagnosis (group III IRS) is between 50 and 70%, which is clearly lower for cases in the neck-head, lung and abdomen region. Patients with distant metastases have a very poor prognosis [14].

In this case, after the biopsy, the patient is classified in group I. This represents a longer survival at 5 years and a lower probability of metastasis.

The ideal treatment for localized disease is surgical resection. Radiotherapy has been shown to play an important role in improving local control after incomplete resection, as SS is a high-grade condition, especially in tumors larger than 5 cm. The role of chemotherapy is still unclear. Unfortunately, due to the diversity and rarity of these tumors, it has not been possible to accumulate an adequate number of patients to compare treatments through randomized trials, however, it is possible to affirm that surgical resection with or without adjuvant radiotherapy and/or chemotherapy based on doxorubicin and ifosfamide are the current pillars of treatment [13,14].

Conclusions

Synovial sarcoma is a malignant tumor that can appear in several locations that, with an accurate diagnosis, classification and multimodal treatment are essential to achieve favorable results and outcome. As it was a soft tissue sarcoma located in the hand, the possibility of salvage surgery with oncological treatment was assessed, which facilitated better incorporation of the patient into the activities of daily living due to the importance of its conserva-

tion, also allowing a favorable psychological evolution. It should be strictly monitored for the possibility of secondary lesions or local recurrence, guaranteeing greater chances of survival.

Conflict of Interest

The authors of this paper certify that there are no conflicts of interest in the realization and review of this topic.

An informed consent form was signed by the patient and her family member after verbal explanation by the attending physician and the Peripheral Tumours Service staff.

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