Leiomyosarcoma of the Hand

Leiomyosarkom der Hand

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Key words
- tumour
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Schlüsselwörter
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Abstract

Soft-tissue sarcomas of the hand are rare and the devastating effect of an undiagnosed sarcoma warrants clinical vigilance. We present the case of an unsuspected leiomyosarcoma localised in the hand in order to underline (i) the rarity of the disease in this site, (ii) the role of adequate surgical treatment in the first step, (iii) the relationship with adjuvant treatments, lymph node metastasis and the poor prognosis of this tumour.

Zusammenfassung

Weichteilsarkome der Hand sind selten und die verheerenden Folgen eines nicht-diagnostizierten Sarkoms mahnen zu klinischer Wachsamkeit. Wir stellen den Fall eines unerwarteten Leiomyosarkoms, dar um folgendes zu unterstreichen: 1) die Seltenheit der Krankheit an dieser Stelle 2) die Bedeutung der adäquaten chirurgischen Behandlung im Anfangsstadium 3) das Verhältnis zu Zusatzbehandlungen, Lymphknotenmetastasen und die schlechte Prognose dieses Tumors.

Introduction

Leiomyosarcoma is an uncommon tumor but in contrast to other kinds of soft tissue sarcomas and with regard to the many histological types, its frequency in adults is not low (ranging between 10–20%) [1,2]. Leiomyosarcomas can be classified into somatic, vascular, osseous lesions [2], and other forms such as uterine (highly aggressive tumor) and cutaneous leiomyosarcoma (excellent prognosis relative to other sarcomas), both with a different prognosis and treatment algorithm compared with the somatic subtype [3]. Hand sarcomas are very rare (1% of all soft tissue sarcomas) and leiomyosarcoma is rarely observed compared to epithelioid and synovial sarcomas, rhabdomyosarcomas and malignant fibrous histiocytoma [1,4,5].

The prognosis for patients with leiomyosarcoma appears to be on an intermediate level between that for patients with liposarcoma and those with undifferentiated pleomorphic sarcoma (HGUPS) [1,3]. Disease-specific mortality is usually related to the development of metastatic disease; however, local effects of the tumor can cause substantial patient morbidity. Reports of variables concerning cases of survival showed that tumor size, grade, anatomic site and surgical margins appear to be the most important factors [1,5]. The presence of tumor necrosis and vascular invasion may also be conducive to a poorer prognosis [3].

We present the case of an unsuspected leiomyosarcoma localized on the hand which was treated incorrectly in the first step and its poor prognosis.

Case Report

A 59-year-old male without concomitant diseases had a 6-year history of evolution of a painless mass localized between 2nd and 3rd fingers on his right hand. In January 2008 the lesion grew significantly to about the size of an olive, without presenting any other symptoms. On July 2008 the patient was treated with surgical excision (lesion was 1.5 cm of diameter) in another center without additional studies. The histological diagnosis was leiomyosarcoma, with only marginal margins. Because of the diagnosis, the patient was referred to our institute for further evaluation and/or treatment. An experienced pathologist reviewed the samples and histological slides and confirmed the diagnosis. Local (CT and MRI) and

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systemic (CT body scan) studies were performed. Considering the absence of previous adequate imaging studies it was not possible to correctly identify the stage of the tumor according to the Enneking classification, even if we supposed it most probably to be a stage IIB. In similar cases, our approach for treatment consists of conservative re-excision and radiotherapy. In this case, considering that no pathological tissue was found on CT and MRI and that the patient refused further surgery, chemotherapy cycles with epirubicin and ifosfamide were started without surgical intervention. External radiotherapy was not performed considering that low efficacy has been reported in patients with soft tissue sarcomas of the hand and the high risk of complications.

Local recurrence was observed on May 2009 at the second interdigital region (**Fig. 1**) and double ray amputation was performed on the affected hand (**Fig. 2**), with a histological confirmation of stage IIB high grade leiomyosarcoma (**Fig. 3**). Immunohistochemistry showed that cells were positive for caldesmon and smooth muscle actin, whereas CD34 was negative (**Fig. 3**). Resection margins of the specimen were free of disease.

The postoperative clinical course of the wound was regular with a good healing and an early mobilization of the hand. During the postoperative follow-up, a CT scan of the lung revealed small lesions in both lungs and ipsilateral nodules in the axilla. Chemotherapy was performed for 4 cycles with gemcitabine and docetaxel until October 2009. Next lung evaluations showed a progression of pulmonary metastases. In February 2010 an axillary lymphadenectomy was performed with histological confirmation of metastatic lesions. The patient received further cycles of chemotherapy with trabectedin, although lung metastases increased in number and size. New local recurrences were identified at the surgical wound and at the mid forearm level. They were removed in January 2011 with histological confirmation of sarcoma. During follow-up, and in spite of established chemotherapy, a progression of disease with bilateral lung metastasis growth and breathing insufficiency was observed. Palliative treatment with analgesia and oxygen therapy was introduced for the last months. Finally the patient died of the disease.

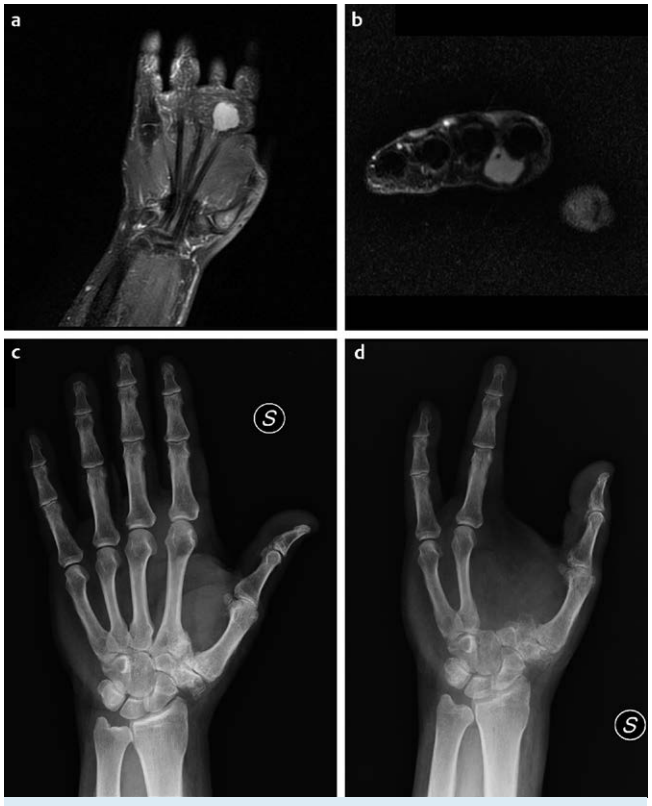

Fig. 1 Preoperative evaluation with MRI **a, b** and X-ray **c** and plain radiograph after double-ray amputation **d**.

Discussion

Soft tissue sarcomas are rare tumors, and the incidence in acral extremities is not frequent either [1,4–6]. To the best of our knowledge, there are less than 10 previous published cases in the literature about leiomyosarcoma of the hand [1,5].

Most hand tumors have a benign nature, but nevertheless a detailed study of the lesion, including accurate imaging and biopsy, should be performed [1,4,5]. In the case of a malignant tumor, an incomplete and poor study of the lesion associated with inappropriate treatment is the prerequisite for subsequent evolution and progression of the disease [1]. Hand preservation is possible in most patients with malignant hand tumors [7], although the type of resection selected (whether en bloc soft tissue resections without digit sacrifice or partial hand amputation) depends to a great degree on the size and location of the tumor. In the literature there is no evidence that amputations or segmental resections are superior to limb-preserving resections when negative margins are achieved [1,5,7]. In cases where resections with adequate margins are not possible, good results have been re-

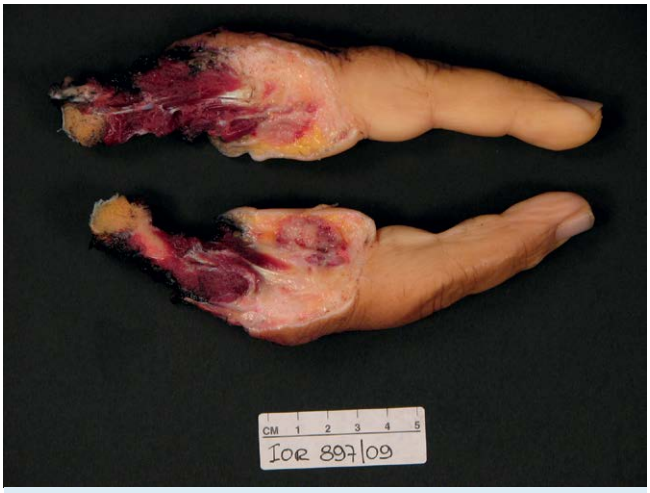

Fig. 2 Gross appearance of the soft tissue leiomyosarcoma constituted of a homogeneous, grayish cut surface, with a well-delimitated margin.

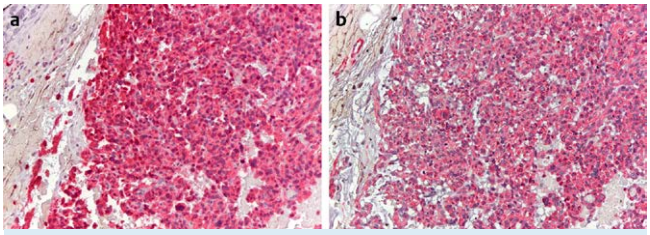

Fig. 3 The lesion was composed of spindle cells focally longitudinally oriented in intersecting fascicles, with eosinophilic cytoplasm, associated with pleomorphic areas. Immunohistochemical features showed that neoplastic cells were positive for caldesmon **a** and smooth muscle actin **b**.

ported with aggressive surgical treatments (amputation or segmental resection and hand replantation) for leiomyosarcomas of the hand and distal forearm as first treatment [1,5,8]. In our case, the first marginal excision of the lesion and the subsequent local recurrence compromised the possibility of limb-salvage surgery, forcing us to perform a double ray amputation of the hand. Likewise, the lack of experience and references in the literature, due to the rarity of this type of malignant tumors, precludes a standardized treatment. The knowledge about the treatment is derived from limited series and single reports [1,5–8]. In our case, the first surgical treatment was performed with inadequate margins but the disease had a rapid growth and progression in a short time, even if it was calm for many years. In addition the tumor did not respond to chemotherapy and it was neither possible to modify the prognosis, nor the course of the disease. In fact, as reported, the tumor had a torpid and practically constant progression despite systemic treatment.

It is well-known that patients with high-grade extremity soft tissue sarcomas are at significant risk for distant recurrence and death from metastatic disease. About chemotherapy, the heterogeneity of the different histological soft tissue sarcoma subtypes suggests that histology-driven treatment is ideal [9]. Since peculiar chemosensitivity towards alternative drugs was described for different metastatic subtypes in second or further lines, the modern concept of 'histology-driven chemotherapy' has been accepted and employed: gemicitabine ± dacarbazine, trabectedin and taxanes are used, respectively, in patients with leiomyosarcoma, solitary fibrous tumor, myxoid/round cell liposarcoma, and angiosarcoma [10]. Leiomyosarcomas are particularly sensitive to the combination of gemcitabine and docetaxel (which is currently the standard of care) and trabectedin in the form of long disease stabilization [2]. Nonetheless, given the rarity of disease, evidence upon which to base histologically-specific neoadjuvant or postoperative adjuvant therapy guidelines is still lacking. However, in the presence of unresectable metastasis the systemic treatment should always be considered palliative [2].

The occurrence of lymph node metastasis, which is an infrequent event in the natural history of leiomyosarcomas, has demonstrated its aggressiveness and has been associated with a poor prognosis [11]. Radiation therapy is an option for adjuvant treatment of soft tissue tumors [12], even in patients who are not candidates for re-excision [7], but important complications have been reported in the hand, such as tissue shrinkage, loss of function and reduction of the range of motion [13]. Moreover low efficacy has been reported in patients with positive resection margins for soft tissue sarcomas of the hand [1]. Radiotherapy was not performed in our case due to these reasons. Concerning prognosis, certainly these outcomes are sometimes unavoidable, but we think that a first adequate treatment at a specialized center is one of the most important prognostic factors.

In conclusion, a primary inadequate surgical treatment for sarcomas of the extremities is associated with a poor prognosis even with systemic treatment and subsequent ablative surgical procedures. This report underscores that care must be taken when approaching proliferative lesions of the hand, even if they appear to be slow growing.

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Conflict of interest: None

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