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Recent advance

Soft tissue sarcomas of the forearm, wrist and hand

Tumeurs malignes des parties molles de l'avant-bras, du poignet et de la main

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ABSTRACT

Soft tissue sarcoma of the forearm, wrist and hand are rare. Their benign appearance leads often to primary inadequate treatment. Due to the complex anatomy of the hand and forearm, they are challenging to treat. The two goals are to obtain wide resection of the primary tumor while preserving function. Limb-sparing surgery is now the cornerstone for the treatment of most sarcomas of the forearm, hand and wrist. To achieve optimal oncological and functional outcomes, the surgical excision should be associated with early reconstructive procedures and a multidisciplinary meeting to define the treatment strategy including adjuvant medical treatments. This article outlines the current principles and presents the results of the treatment of soft tissue sarcomas with emphasis on to particularities related to their forearm, wrist and hand location.

RÉSUMÉ

Les tumeurs malignes des parties molles de l'avant-bras, du poignet et de la main sont des tumeurs rares, souvent d'apparence bénigne et régulièrement initialement négligées. De par la complexité de l'anatomie de la main et de l'avant-bras, leur prise en charge est difficile et représente un vrai challenge thérapeutique. L'objectif à atteindre est de concilier à la fois une exigence carcinologique et une exigence fonctionnelle d'importance majeure à l'avant-bras, au poignet et à la main. La chirurgie d'exérèse conservatrice est aujourd'hui la pierre angulaire du traitement de la plupart des sarcomes à l'avant-bras, au poignet et à la main. Pour permettre un résultat carcinologique et fonctionnel optimal, cette exérèse chirurgicale doit être associée à une chirurgie reconstructrice précoce et à une prise en charge multidisciplinaire, afin de définir un projet thérapeutique global incluant les traitements médicaux adjuvants. Cet article précise les principes actuels et les résultats de prise en charge des tumeurs malignes des parties molles en insistant sur les aspects spécifiques des localisations à l'avant-bras, au poignet et à la main.

1. Introduction

Malignant tumors of the soft tissues, or soft tissue sarcomas (STS), are rare malignant tumors. They make up less than 1% of newly diagnosed cancers each year [1]. Their incidence is 2–3 cases per 100,000 [2] and there are an estimated 3000 to 4000 new cases each year in France [3,4]. About 60% of these STS develop in the limbs, of which 15% to 25% affect the upper limb and only 5% to 10% affect the hand or wrist [5–7]. STS are histologically heterogeneous, with about 50 different histological kinds described [8]. In the upper limb, the most common STS are synovial sarcomas, epithelioid sarcomas and malignant histiocytofibromas [5,9] (Table 1).

These tumors in the forearm, wrist and hand are challenging to treat. Optimal treatment for the cancer should be balanced with minimal impact on function.

Since the end of the 1970s, radical treatment by amputation is no longer the reference surgical treatment for these lesions [10,11]. Limb-sparing excision and reconstructive surgery are now

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Table 1
The most common histological types of soft tissue sarcomas in the hand, wrist and forearm.

Malignant histiocytofibroma (undifferentiated pleomorphic sarcoma)
Synovial sarcoma
Epithelioid sarcoma
Clear-cell sarcoma
Fibrosarcoma
Leiomyosarcoma
Liposarcoma
Malignant peripheral nerve sheath tumor
Dermatofibrosarcoma protuberans
Myxoinflammatory fibroblastic sarcoma

used in the hand and wrist [12–14]. To achieve the best possible functional and cancer-related outcomes, this surgical excision must be combined with early reconstructive surgery and multidisciplinary care, in order to define the overall treatment plan including adjuvant medical treatments (chemotherapy and radiation therapy). The current principles of the various steps of the care of these STS are presented in succession, with specific features related to the forearm, hand and wrist.

2. Diagnosis

The clinical presentation of STS is often misleading and can appear trivial [15]. In most cases, a subcutaneous mass is discovered by chance, sometimes during the final healing phases of an injury; it is often painless and without associated symptoms. In this location, the tumors are usually small – less than 4 cm – in size [5]. The chronicity of the mass is not evidence of its benign nature and rapid progression is more suggestive a high-grade tumor. It is important to remember that in the hand and wrist, there are more benign than malignant tumors. However, any atypical "suspicious" mass must receive comprehensive care, including a full clinical examination and imaging work-up. If malignancy is still suspected after this assessment, a primary biopsy is recommended.

The clinical examination must qualify the mass (size, adhesion, hardness, cutaneous invasion, etc.), evaluate the local consequences (functional, vascular, neurological) and in general, palpate the lymph nodes (certain tumors target the lymph nodes: epithelioid sarcomas, clear-cell sarcomas, synovial sarcomas, rhabdomyosarcoma) and look for familial disease that is often associated with a tumor (e.g. malignant tumor of the peripheral nerve sheaths and type I neurofibromatosis, Li-Fraumeni syndrome, etc.) or even exposure to carcinogens (herbicides, pesticides, radiation) [15].

The imaging work-up must include standard X-ray views to rule out bone tumors or invasion of neighboring bones.

Ultrasound imaging, which is commonly available and non-irradiating, now has very extensive indications for the assessment of forearm, wrist and hand masses. While ultrasonography only amounts to a scout scan, it makes it possible to evaluate the liquid or solid, homogeneous or heterogeneous, superficial or deep nature of the mass. When correlated with the clinical findings, it is often able to confirm the benign nature of most suspicious masses in these locations. If the results of US are atypical, not well correlated with the clinical findings and do not allow malignancy to be ruled out, MRI is required before any treatment is initiated. Ultrasonography is also indicated in the supplemental assessment of STS to look for associated adenopathy.

MRI is now the gold standard examination for STS. The MRI must be performed before any biopsy is done [15] and consists of several T1- and T2-weighted sequences, with and without contrast. In each sequence, orthogonal planes [16] (ideally with an axial plane) must be visible to allow better surgical targeting of the tumor margins and anatomical structures and to facilitate preoperative planning of the tumor excision margins [17]. While MRI often orients the diagnosis [18], it has no certitude and cannot substitute for a biopsy.

Local CT scan of the STS region is only relevant if MRI cannot be performed, or if bone invasion is present.

Chest and abdomen/pelvis CT with contrast is the gold standard examination for disease staging. Metastasis typically impacts the lungs, infrequently the soft tissues or abdomen in certain histological STS types (myxoid liposarcoma, clear-cell sarcoma) and even more rarely the lymph nodes (clear-cell sarcoma, epithelioid sarcoma, synovial sarcoma, rhabdomyosarcoma, angiosarcoma). In certain histological STS types, a positron emission tomography (PET) scan, which has growing indications for STS [19–21], can be proposed in the context of disease staging or of recurrences. However, this examination is not currently recommended in the context of STS care [20].

Lastly, vascular imaging (angiography, CT angiography, MR angiography) can be indicated, particularly for tumors requiring complex reconstruction.

Carrying out this complete diagnostic strategy will reduce the probability of diagnostic errors and inappropriate surgical treatments. Also, a complete preoperative imaging work-up will make surgical revisions easier to perform, if needed.

3. Treatment

The treatment principles for STS in the hand, wrist and forearm are widely described in the literature and do not differ from the treatment of STS in other locations [3,15,22–24]. There are however some distinctive features in the hand and wrist.

Because of their small size and more superficial nature, STS of the hand, wrist and forearm are often operated on right away without appropriate consideration of the possibility of malignancy, with or without an incomplete, or even inappropriate preoperative work-up. This was the case for 38% of patients in the Pradhan study [25] and 68% of patients in the Bray study [26]. And yet, the impact of the initial surgery on the risks of residual or recurrent disease it is widely reported in the literature [27–30], along with the importance of care in a specialized cancer center [31–33] and a review of the case in a multidisciplinary meeting (MDM) before treatment is initiated [22].

3.1. Biopsy

The first phase of care is the biopsy. It will be used to confirm the benign or malignant nature of the mass, classify the tumor, plan the type of surgery needed in an optimal manner and in advanced STS cases, to consider performing neoadjuvant preoperative treatments (chemotherapy, isolated limb perfusion, radiotherapy) [3].

Not every soft tissue mass in the hand and forearm require a biopsy. There are many more benign than malignant tumors. If the clinical and radiological findings are typical (synovial cyst, paraarticular mucoid cyst, superficial lipoma, giant cell tumor of the synovial sheaths, etc.), an excision biopsy can be performed right away. However, if there is even the slightest clinical or radiological suspicion on the potential malignancy of a mass, primary biopsy is justified. There are no formal clinical or imaging criteria for malignancy. A long-standing mass is not always benign. Conversely, a rapidly growing mass is not always malignant [15]. However, a mass that has rapidly grown, of more than 3 cm in the upper limb, adhering to the superficial or deep layers, that is poorly defined or subfascial, must be suspected for malignancy. Similarly, on imaging, a heterogeneous mass that is poorly systematized, that takes up contrast agent on MRI and is intramuscular, must be suspected for malignancy. Under these conditions, a primary biopsy is indicated.