Surgical management of soft tissue sarcoma

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Abstract

Soft tissue sarcomas (STS) are a heterogeneous group of tumours and surgery is the principal treatment. The majority occur in the limbs and trunk wall and this article refers to management of these anatomical sites. STS is managed within a specialist multidisciplinary team and an individualized treatment plan is produced for each patient. Histological, anatomical and patient factors will all influence the strategy. In localized disease, the surgical aim is to achieve a wide excision with negative margin and preserve function. Radiotherapy may be indicated to reduce the risk of local recurrence and can be given pre- and/or post-operatively. Planned marginal excision combined with radiotherapy is an acceptable approach in order to preserve critical structures. Neo-adjuvant radiotherapy can reduce tumour burden and improve operability. However, amputation is sometimes the only treatment option. Reconstructive surgery may be required to achieve wound healing and restore function. The approach to management of local recurrence is much the same as for primary disease. Advanced disease is predominantly treated by systemic therapy but metastatectomy is utilized in some circumstances. Follow-up is required to monitor for recurrence, metastasis and complications of treatment.

Keywords margin; radiotherapy; reconstruction; sarcoma; surgery

Introduction

Soft tissue sarcomas (STS) are rare malignant mesenchymal tumours arising in fat, muscle, nerve, blood vessels and other connective tissues. The World Health Organization classification of STS includes more than 50 different sub-types, which are classified by tissue of origin and biological potential. Whilst the term 'STS' refers to malignant tumours, there are also some intermediate soft tissue tumours that may be managed under the sphere of STS due to locally aggressive or rarely metastasizing behaviours.

STS are a heterogeneous group of tumours with variable clinical, prognostic and therapeutic features. They can occur anywhere with the extremities being the most common site, followed by the trunk.[1] Of the peripheral and truncal tumours, two thirds arise beneath the deep fascia and have an average diameter of 9–11 cm at diagnosis; the remainder are more superficially located with an average diameter of 5–6 cm at presentation.[2,3] Retroperitoneal, intra-abdominal, gynaecological and pelvic tumours are often large at time of presentation and require management by a specialist sarcoma surgeon with the appropriate site-specific expertize. The management of these is outside the scope of this article.

Guidelines for management of STS are available from various sources[4–7] The principal treatment of primary STS is surgical excision often combined with adjunctive radiotherapy. Chemotherapy or other pharmacological agents are also utilized for specific tumour sub-types and within therapeutic trials.

The surgical management of peripheral and trunk wall STS is described in this article.

Diagnosis

Whilst this article does not focus on diagnosis, it is important to establish the diagnosis prior to treatment and manage the patient in a multidisciplinary team (MDT) setting. Image-guided core needle biopsy following MRI is the standard pathway. Occasionally incisional and rarely excisional biopsy is required and performed under the guidance of the MDT. Unplanned excision of a potentially malignant soft tissue mass is undesirable.

Surgical planning

Definitive resection of an STS should be performed at a designated STS treatment centre by an STS surgeon or a surgeon with tumour site-specific skills in consultation with the sarcoma MDT.

Treatment recommendations will be made by the MDT for patients who have followed the prescribed referral pathway. The MDT will typically have access to the following information:

- clinical features and functional status (history and examination by STS surgeon)
- MRI with contrast (performed in conjunction with and reported by STS radiologist)
- core needle biopsy (ideally image-guided, performed by STS service, processed by a specialist sarcoma pathologist)
- staging CT chest (further imaging sometimes indicated depending on tumour site and type).

All of these factors will influence the surgical procedure and its timing.

The aim of surgical resection is to achieve oncological clearance with minimal morbidity, hence maximizing long-term survival, avoiding local recurrence and preserving function. An algorithm of basic principles for treatment of localized STS is outlined in Figure 1.

Where possible the aim should be to achieve a wide excision with negative surgical margin. In some cases this is possible without a significant impact on function. When this is not possible consideration should be given to the impact of resecting

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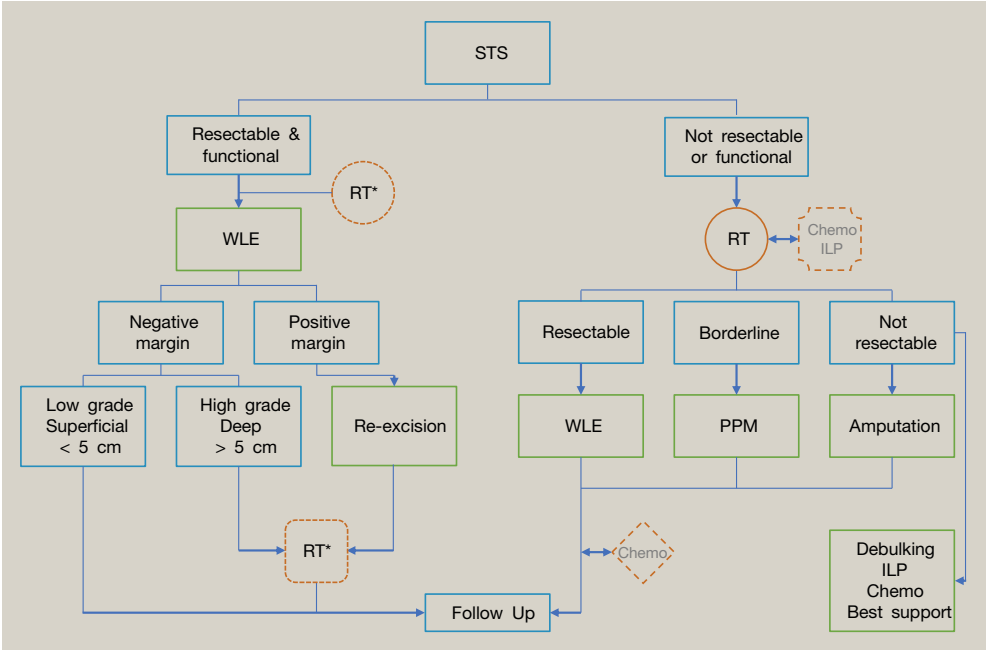

Figure 1 Algorithm for management of localized soft tissue sarcoma (STS). *Radiotherapy is given either pre-operatively or post-operatively in resectable STS that are high-grade, deep or large. Chemotherapy may be used pre-operatively in chemo-sensitive STS sub-types that are difficult to resect. Isolated limb perfusion is also a pre-operative treatment option for limb STS that are difficult to resect. Post-operative chemotherapy may be offered in selected high-grade STS. RT, radiotherapy; WLE, wide local excision; Chemo, chemotherapy; ILP, isolated limb perfusion; PPM, planned positive margin.

structures, the possibilities for reconstruction, the prognostic implication of a more conservative excision and suitability for neo-adjuvant or adjuvant treatments, along with patient wishes. Although primary amputation is now unusual, it must still be considered according to patient preference and if reconstruction of a functional limb is not possible or sensible. It must also be remembered that some STS are not resectable and in addition patient factors such as fitness for surgery or advanced disseminated disease may result in a decision not to operate.

Margins

Surgical

There remains a lack of consensus regarding the desirable excision margin in STS. Enneking et al. first described a model for sarcoma resection in 1981[8] based on the anatomical and pathological features of sarcoma growth (Figure 2). This took account of the potential presence of microscopic tumour in the reactive zone, or 'pseudocapsule' around the tumour, and the possibility of microsatellites within adjacent normal tissues. Excision was classified into four categories:

- intralesional: the tumour is breached macroscopically
- marginal: the pseudocapsule forms at least part of the periphery of the specimen
- wide: non-reactive normal tissue forms the entire periphery of the specimen
- radical: all normal tissue of the anatomical site is removed (e.g. fascial compartment and its sheath).

Other groups have attempted to quantify the amount of normal tissue required for a 'safe' margin in wide excision and in doing so realized the importance of tissue composition. Rydholm and Rooser observed that local recurrence was higher in STS that were excised with a wide margin of muscle or areolar tissue compared with those that had a wide margin of fascia or subcutaneous tissue.[9] Kawaguchi et al. analysed the width and quality of tissue comprising the margin and suggested that smaller margins were acceptable in the presence of a barrier such as periosteum or fascia.[10] In addition specific STS sub-types (eg. myxofibrosarcoma) have high risk of local recurrence despite wide margin excision, whilst others have low risk despite marginal excision.[11] Therefore both tumour biology and the inherent resistance of tissues to invasion also influence the planned margin.

Although excision margins of up to 5 cm have been suggested for STS, the introduction of adjuvant therapy has changed practice. The effect of radiotherapy on microscopic tumour has allowed substantial reduction in the surgical margin with some

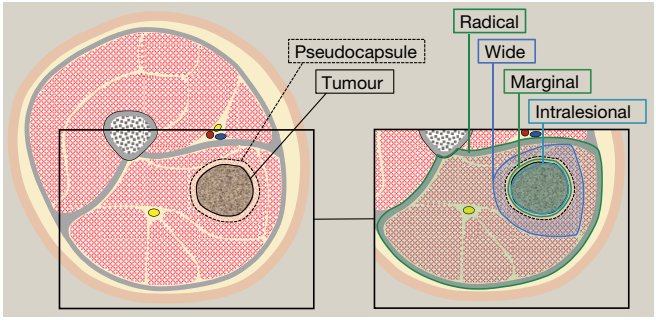

Figure 2 Schematic diagram of Enneking's description[8] of soft tissue sarcoma excision. Cross-section of thigh showing tumour in adductor magnus with surrounding reactive zone or 'pseudocapsule'. Four different classes of surgical excision are depicted, indicating tissue excised with each type of resection.