Management of extremity soft tissue sarcoma after unplanned incomplete resection: experience of a regional musculoskeletal tumour centre

This study reviews the presentation and management of extremity soft tissue sarcoma after unplanned incomplete resection in a musculoskeletal tumour centre in Hong Kong. Medical records of 18 patients who were referred to our centre for further management from January 1995 to May 2001 after inadequate tumour excision were reviewed. Fourteen patients had been referred from private clinics and four from public hospitals. At initial presentation, 10 patients had lesions exceeding 5 cm, nine had a tumour deep in the subfascial plane, and eight had tumours that had recently increased in size. Sixteen had no preoperative radiological assessment or biopsy performed before excision. All except two patients needed additional skin and muscle reconstruction in a subsequent re-resection, and 12 required postoperative radiotherapy. Two patients subsequently developed distant metastases, and one patient died of an unrelated cause. No amputations were required, and no major complications arose from second surgery. Physicians’ alertness towards the possible malignancy of soft tissue masses in extremities is important to avoid a potentially mutilating second resection. Well-planned re-resection in a specialised tumour centre can achieve satisfactory local control of disease.

Introduction

Soft tissue sarcomas of the extremity are mesenchymal tumours arising from connective tissue elements; such tumours feature local invasiveness, as well as a high propensity to local recurrence and distant metastasis. Fifty percent of patients with sarcoma eventually die of the disease. However, these tumours are rare and have an average annual incidence of 169 cases, according to the figures reported to the Hong Kong Cancer Registry from 1993 to 2000, with a slight male preponderance. The rarity of such tumours could, in turn, lead to an inadequate awareness of diagnosis and thus improper excision. This ‘unplanned incomplete resection’, defined as excisional biopsy only or unplanned resection of the lesion without prior information from preoperative imaging and without regard for the
necessity to resect the lesion with a margin of normal tissue,\textsuperscript{3} will lead to higher local recurrence\textsuperscript{4,5} and may jeopardise overall survival.\textsuperscript{6,7} Subsequent ‘well-planned resection’ with adjuvant radiotherapy is indicated to improve overall local control.\textsuperscript{8-11} However, a second operation frequently leads to further extensive and potentially mutilating surgery.\textsuperscript{12} This study aims at analysing the initial features of presentation of soft tissue sarcomas and the possible harm incurred by primary unplanned surgery in terms of skin and muscle reconstruction, and the need for additional adjuvant therapy (ie brachytherapy, external beam radiotherapy, or chemotherapy) after surgery.

**Methods**

From January 1995 to May 2001, 76 patients with soft tissue sarcoma were operated on in the Department of Orthopaedics and Traumatology, Queen Elizabeth Hospital—a designated musculoskeletal tumour centre of the Hospital Authority. Of the patients, 18 had had prior operations and had been referred to our centre for further management because of incomplete primary excision. Case records of these patients, who were followed up for an average of 42 months (range, 23-100 months), were reviewed retrospectively.

All patients had been jointly assessed in a multi-disciplinary sarcoma clinic by dedicated tumour surgeons, clinical oncologists, radiologists, and pathologists to delineate the plan of further management. Our protocol was to perform complete wide resection of the previously operated site if technically feasible, and to add adjuvant radiotherapy in the form of external beam radiotherapy with or without perioperative brachytherapy.\textsuperscript{13,14} Before the second operation, all patients underwent chest radiography, computed tomography (CT) of the thorax, and isotope bone scanning to exclude distant metastasis, as well as magnetic resonance imaging (MRI) of the previous primary tumour site for surgical planning. When possible, pathologists specialising in musculoskeletal tumours reviewed slides of the initial specimen. The demographic data, initial presenting features, nature of primary operation, and subsequent treatment were recorded.

**Results**

**Demographic data**

Table 1 shows the details of all 18 patients included in this study, of whom 10 were male and eight were female. The mean age was 55 years (range, 17-80 years).

**Initial presentation**

Tumours were located in an upper limb in nine cases, a lower limb in five, and the trunk in the remaining four. For 17 patients, a mass had been present for more than 4 weeks (mean, 6.6 months). In eight cases, the leading symptom was an enlarging soft tissue mass. Pain was not a significant symptom in any patient. The mass was larger than 5 cm in 10 patients, and nine tumours were located deep in the fascia.

Overall, seven patients had a subfascial mass of larger than 5 cm at presentation, which was commonly regarded as a feature to suggest malignancy. Only one patient underwent preoperative MRI, which revealed a subfascial mass, but not a subsequent biopsy. Sixteen patients had no biopsy before excision; the remaining two patients underwent fine-needle aspiration biopsy, yielding mesenchymal tumour of uncertain nature in one and inadequate tissue for diagnosis in the other.

**Primary operation and histology**

Two patients underwent marginal excision, whereas 15 had intralesional excision performed and one underwent only piecemeal removal. The surgery in 14 patients was performed in private clinics and that in four were done in public hospitals. Histological studies revealed a diversity of tumours, including dermatofibrosarcoma protuberans (n=5), malignant fibrous histiocytoma (4), undifferentiated sarcoma (2), leiomyosarcoma (1), high-grade myxoid liposarcoma (2), well-differentiated liposarcoma (1), de-differentiated liposarcoma (2), and high-grade myxofibrosarcoma (1).

**Deleterious effects of inadequate primary operation**

All but two patients required additional skin and muscle reconstruction during a subsequent operation. Reconstruction was necessary because of the deleterious effects caused by primary surgery in 10 patients: three cases for oversized scarring, three for the presence of a distant drain site (Fig 1a), one for broad mattress-stitch scarring (Fig 1b), one for extensive postoperative haematoma (Fig 1c), and two for repeated inadequate excision. In addition, adjuvant radiotherapy was indicated in five patients because of incomplete excision during primary surgery.

As a case example, patient No. 7 presented with a 1-year history of a 3-cm subcutaneous mass over the right upper arm. Initial marginal excision was performed under local anaesthesia, and histological examination revealed myxofibrosarcoma. The second excision, with a 1-cm margin, was performed 4 weeks later by the same surgeon, but remained inadequate because the resection margin was still involved. The patient was then referred to our centre for further management. After discussion in the combined sarcoma clinic, wide excision was performed and a transpositional parascapular flap was created to cover the large cutaneous defect (Fig 2a, 2b). Brachytherapy catheters were also inserted during the operation (Fig 2c). Perioperative brachytherapy and external beam irradiation were delivered subsequently. The donor wound was closed primarily. All wound healed uneventfully but left prominent scars in the shoulder and back regions (Fig 2d). There was no local recurrence or distant metastasis detected for this patient after 32 months of follow-up.

**Subsequent management**

The main curative therapy for soft tissue sarcoma is wide excision with 2- to 3-cm margin of grossly normal tissue. In
all 18 patients, adequate margins were not achieved. Hence, subsequent well-planned resection with adjuvant radiotherapy was indicated to improve overall local control. Accordingly, all patients underwent MRI of the previous surgical site to delineate the extent of residual disease. In addition, CT of the thorax and isotope bone scanning were
pathology after repeated resection. Overall, MRI was neither sensitive nor specific enough to accurately detect residual tumour. However, the MRI scan was still necessary in the planning of surgery, because subsequent wide excision needed to include tissue with abnormal signal changes to facilitate complete tumour removal. Tumour staging showed no evidence of distant metastasis in any of the patients at the time of second surgery.

Two patients underwent no further surgery and received radiotherapy alone. In one of these patients (No. 10), remedial wide margin resection would have resulted in major disfiguration of external genitalia, and hence it was recommended against by a gynaecologist. The other patient who had low-grade sarcoma had a clear, although close, margin in the primary operation. Wide-margin excision was achieved in the remaining 16 patients. In two patients, direct wound closure was possible followed by subsequent radiotherapy. Four patients required wide excision with skin grafting and muscle flap creation, whereas a combination of wide excision, skin grafting, muscle flap creation, and radiotherapy was performed in 10 patients. In all patients, the involved limb was preserved after completion of all treatment. All excised tissue was sent for sectioning and histological analysis, which showed that nine of the 18 patients had residual tumour at the primary surgical site.

Follow-up outcomes were obtained at an average interval of 42 months (range, 23-100 months). Until the last follow-up visit, two patients had distant metastasis: the first, patient No. 1, had metastases in the kidney and left leg diagnosed at 9 months and 28 months after re-resection, respectively. He, however, refused further treatment and was still alive at the last follow-up. The second patient (No. 16) had lung metastasis at 31 months after re-resection, and a further treatment plan was being formulated at the last follow-up. Among the 18 patients, only one (No. 6) died because of an unrelated cause at 19 months after surgery; there was no evidence of local recurrence or distal metastasis. The remaining patients survived without evidence of tumour recurrence, high-grade complications affecting daily life, or any neurovascular deficits or wound complications that needed further flap surgery.

Discussion

Soft tissue “swelling” of an extremity is frequently seen in daily practice, and the majority of them are benign lesions. To diagnose sarcoma among these soft tissue swellings requires a level of clinical suspicion. Clinical features suggestive of malignancy include the following: (1) a mass of larger than 5 cm, (2) pain in a previously painless lump, (3) rapid increase in size, and (4) subfascial location of a mass. These suspicious masses should be subjected to further investigation before definitive excision. Magnetic resonance imaging is the most accurate and valuable imaging technique to visualise the anatomy of the lesion and its relationship with surrounding vital structures, such as the neurovascular

Table 2. Magnetic resonance imaging (MRI) and histological results after re-resection

<table>
<thead>
<tr>
<th>MRI</th>
<th>n*</th>
<th>Histological evidence of residual tumour</th>
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<tbody>
<tr>
<td>Positive</td>
<td>2</td>
<td>Yes</td>
<td>2</td>
</tr>
<tr>
<td>Uncertain</td>
<td>4</td>
<td>Yes</td>
<td>1</td>
</tr>
<tr>
<td>Negative</td>
<td>10</td>
<td>Yes</td>
<td>4</td>
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* MRI was not done in two cases because surgery was not performed
bundle, and to allow detailed preoperative planning of resection margins. Biopsy should be performed only after MRI, because the procedure may alter the appearance of the MRI scan. Image-guided percutaneous tru-cut biopsy is currently our preferred method of obtaining tissue for pathology. One study showed an overall diagnostic accuracy of 96% with such a biopsy method, and other studies have demonstrated variable success rates, ranging from 60% to 100%.17 Besides the high diagnostic accuracy, image-guided biopsy also has the added advantages of avoiding damage to nearby vital neurovascular structures and tumour contamination by transversing the tissue plane during the procedure. The morbidity and mortality rates were also lower than those associated with open biopsy.18,19 Communication between the surgeon and radiologist is of utmost importance to ensure a proper biopsy site, so that positions of operative incisions and musculocutaneous flaps are correct. When inadequate tissue is obtained from image-guided needle biopsy, open-biopsy should be performed. Open-biopsy itself is not a minor procedure and should be performed in a specialised centre by a surgeon especially trained in musculoskeletal oncology who will also perform the subsequent resection.20 Once proper investigations have been completed, limb-sparing surgery consisting of wide excision with a 2- to 3-cm margin of grossly normal tissue is the main curative therapy. Adjuvant radiotherapy, in form of external beam radiotherapy with or without brachytherapy should be considered in cases of high-grade sarcoma, because this treatment is very effective in preventing local recurrence in patients after wide local excision.21,22

This study reveals the lack of awareness of potential malignancy in soft tissue swellings despite the presence of one or more clinical clues. The incomplete resection during the primary operation implies that local relapse can be expected in more than 90% of patients without further therapy,23,24 in contrast to the 10% recurrence rate after a primary well-planned wide excision with or without adjuvant radiotherapy.23-26

![Fig 2. Patient No. 7: (a) planned wide excision and design of parascapular flap; (b) transposition of parascapular flap; (c) insertion of brachytherapy catheters; (d) prominent scarring in shoulder and back](image-url)
Radiotherapy alone is not advocated as a remedial approach, as demonstrated in a study by Giuliano and Eilber, in which 90 patients were referred after an excisional biopsy of soft tissue sarcoma; all were treated with adriamycin (doxorubicin) and radiation therapy before re-resection. Despite the intensive preoperative treatment in that study, half of the patients still had gross evidence of residual sarcoma in final pathology. The presence of residual tumour in 50% of re-resected specimens in this study again supports the current recommendation of repeated wide-margin resection after incomplete primary surgery, so as to improve the local control of disease. These secondary surgeries should be performed in musculoskeletal tumour centres by a multidisciplinary team of dedicated tumour surgeons, clinical oncologists, radiologists, and pathologists, all contributing to optimal tumour control.

As demonstrated in this study, re-resection in the majority of cases necessitates more extensive surgery. If the initial management were planned properly, more than half of the patients (10/18) would have avoided such extensive surgical reconstruction and more than a quarter of them (5/18) additional adjuvant radiotherapy. The loss in functional outcome and the emergence of potential complications due to the additional procedures could therefore be avoided.

Conclusion

A high level of clinical suspicion and proper investigation will avoid incomplete resection. Early referral to a specialised musculoskeletal tumour centre for clinically suspicious masses should be encouraged, because diagnostic errors and treatment complications are far less likely if biopsy is performed properly. Subsequent definitive treatment aiming at complete eradication of tumour while sparing the tumour-bearing limb requires the joint effort of various specialties. Re-resection for patients referred after inadequate primary surgery by surgeons specialised in musculoskeletal oncology can still achieve local tumour-free status, thereby avoiding more extensive and mutilating surgery.

References

2. Hong Kong Cancer Registry, Hospital Authority. Soft tissue sarcoma registered in Hong Kong. Hong Kong: Hospital Authority; 2002.