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The diagnosis of soft tissue tumours

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We prospectively analysed methods of diagnosis in 118 patients referred for definitive treatment with documented or presumed soft tissue sarcoma (STS). Of 65 patients with primary STS, 54 were biopsied before referral. Of these, 5 (9%) were biopsied by Tru-cut® biopsy, 17 (32%) by incisional biopsy and 32 (59%) by excisional biopsy. The remaining 11 patients with primary STS, referred without biopsy, were all diagnosed by Tru-cut biopsy. An additional eight patients suspected of having STS were referred without biopsy and were found to have malignant tumours other than STS involving soft tissue by Tru-cut biopsy. Nineteen patients were proved to have benign soft tissue tumours; in 13 presumed to have STS, the diagnosis was unknown at referral. In four of these, biopsy was inappropriate. Of nine submitted to Tru-cut biopsy, an unequivocal diagnosis was made in 5 (56%) and incisional biopsy was required in the other four. Therefore, paradoxically, benign soft tissue tumours may be more difficult to diagnose with Tru-cut biopsy than malignant tumours. This study confirms the high degree of accuracy of Tru-cut biopsy in diagnosing malignant soft tissue tumours and highlights the disadvantages of open biopsy techniques.

The annual incidence of soft tissue sarcoma (STS) is about 1200 in the United Kingdom. As a group, STS represent less than 1% of all malignant tumours. These levels are approximately those of testicular cancer. Soft tissue sarcomas are therefore rare *(1)* and an individual surgeon may see no more than one new case each year. Because of this, the diagnosis is often unsuspected at the time of presentation and an inappropriate biopsy is performed.

Although computed tomography (CT scanning) and magnetic resonance imaging (MRI) are helpful diagnostic techniques, a histological diagnosis must be obtained before definitive treatment. Previously, incisional biopsy was the recommended technique to establish the diagnosis of STS *(2)*. It was argued that incisional biopsy was necessary to obtain adequate amounts of tissue and thereby avoid errors of sampling and interpretation. However, there are significant disadvantages to incision biopsy which include inappropriate siting of the incision and unacceptable rates of wound complications *(3,4)*. As a result of this, the value of Tru-cut® core needle biopsy was evaluated at this unit *(5)* and an accuracy of 98% was reported for the diagnosis of STS *(6)*.

In appropriately selected patients, survival rates achieved by a combination of surgery and radiotherapy resulting in limb conservation, are equivalent to those after radical amputation *(7,8)*. To achieve the best results from limb conservation, appropriate planning of any biopsy is essential so that subsequent resections are not compromised.

Many pathological conditions, including benign tumours and non-neoplastic lesions, may present as a mass in an extremity and be confused with a soft tissue sarcoma *(9)*. Although various diagnostic features of these are well known *(10)*, with the possible exception of a haematoma, all of these conditions will require surgical intervention. Awareness of the possible differential diagnoses, and a suitable biopsy of the lesion will ensure appropriate treatment.

In the current era of surgical audit and increasing subspecialisation, the purpose of the present study was to identify and discuss methods of diagnosis of soft tissue tumours, occurring during a 12-month period at a referral centre for the mangement of soft tissue sarcomas.

Patients and methods

During the 12-month period, 1 October 1989 to 30 September 1990, 130 patients with soft tissue tumours were referred to the surgical section of the Soft Tissue

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Sarcoma Unit, Royal Marsden Hospital (RMH), usually with an established or suspected diagnosis of soft tissue sarcoma. Each patient was registered at presentation and complete demographic, clinical, treatment and pathological details were recorded prospectively by a Data Manager. There were 76 males and 54 females with a median age of 45 years (range 10–82 years).

Several categories of patient were referred. Some patients were referred with a suspected diagnosis of STS without biopsy or with the result of incision biopsy. Many patients were referred having had their STS enucleated because the diagnosis had unfortunately not been suspected clinically. Some patients were referred with recurrent STS after previous treatment. Finally, a small group of patients with known or suspected benign soft tissue tumours were referred.

Of the 130 patients with soft tissue tumours, 12 patients with documented soft tissue sarcomas were referred for opinion only, resulting in 118 patients available for analysis. Of these, 50 (42.4%) patients were referred from general surgeons, 29 (24.6%) from orthopaedic surgeons, 13 (11.0%) from radiotherapists, 10 (8.5%) from medical oncologists and the remaining 16 (13.6%) from miscellaneous sources.

Results

The final broad diagnostic categories for the total series of 118 patients with soft tissue tumours were: primary soft tissue sarcoma in 65 (55.1%) patients; recurrent soft tissue sarcoma in 26 (22.0%); benign soft tissue tumours in 19 (16.1%); and malignant tumours other than STS involving soft tissue in 8 (6.8%).

Thus, of the 118 patients, 91 (77.1%) had STS. The diagnosis of those with recurrent STS was verified by histological review. Of 65 patients with primary disease, biopsy had been performed at the referring hospital in 54 patients, while 11 patients were referred without biopsy on suspicion alone of STS. The techniques of biopsy used at the referring hospital are detailed in Table I, and are compared with the technique of biopsy used at the Royal Marsden Hospital.

Of 17 (12%) of patients undergoing incision biopsy before referral, two developed severe wound complications. In one patient, the wound failed to heal and preoperative radiotherapy had to be abandoned as tumour grew into the incision. In the other patient, the tumour itself became irreversibly infected and the leg had to be amputated above the knee.

The detailed pathological diagnoses of those with soft tissue sarcomas are shown in Table II. Of the 15 patients with leiomyosarcoma, 12 (80%) arose within the retroperitoneum. Of the 16 patients diagnosed by Tru-cut core biopsy (Table I), in no case did the histological diagnosis differ after resection when the whole specimen was available for examination. Of the 91 patients with STS, 51 (56%) arose from the pelvic girdle and lower limb, 22 (24%) from the retroperitoneum, 15 (16%) from the shoulder girdle and upper limb, and 3 (3%) from the head and neck. The median size of the primary soft tissue sarcomas of the limb and limb girdles was 12 cm (range 4–30 cm).

Of the 118 patients, 19 (16.1%) were proved to have benign soft tissue tumours (Table III) and of these, 13 were referred with undiagnosed soft tissue masses. The correct diagnosis was made by Tru-cut biopsy in seven cases, although in two of these a confirmatory incisional biopsy was performed, due to the unusual nature of the lesion (non-specific myositis, myxoma). Tru-cut core biopsy failed in two patients, one patient underwent incisional biopsy for myositis ossificans after failure of Tru-cut biopsy due to heavy calcification. Another

Table I. Methods of diagnosis of primary soft tissue sarcomas ($n = 65$)

Diagnosis before referral		*Diagnosis made at RMH*
Tru-cut core biopsy	5 (9%)	11 (100%)
Incisional biopsy	17 (31%)	0
Excisional biopsy	32 (59%)	0
Total	54	11

Table II. Pathological diagnosis of soft tissue sarcomas

Histology	*Number*
Malignant fibrous histiocytoma	20
Liposarcoma	13
Leiomyosarcoma	15
Rhabdomyosarcoma	5
Malignant peripheral nerve sheath tumour	4
Clear cell sarcoma	3
Fibrosarcoma	1
Haemangiosarcoma	1
Synovial sarcoma	10
Chondrosarcoma—extraskeletal	2
Sarcoma—not otherwise specified	15
Dermatofibrosarcoma protuberans	2
Total	91

Table III. Benign soft tissue lesions

Diagnosis	*Number*	*Diagnosis known at referral*
Lipoma	4	1
Myxoma	2	0
Arteriovenous malformation	3	0
Myositis ossificans	1	0
Non-specific myositis	1	0
Fibromatosis	5	4
Lymphangioma	1	1
Encysted suprapatellar synovitis	1	0
Chronic osteomyelitis	1	0
Total	19	6

patient subsequently shown to have an encysted suprapatellar synovitis required incisional biopsy as repeated Tru-cut biopsies yielded fluid material only. One patient referred with a histological diagnosis of retroperitoneal sarcoma was proved to have chronic osteomyelitis of the pelvis on review of histology and confirmation by CT-guided Tru-cut biopsy. Of four lipomas, two were massive, one a dumb-bell tumour through the greater sciatic notch involving the pelvis and the buttock, and the other within the adductor compartment of the thigh. Both were confirmed on Tru-cut biopsy, one before referral. Another patient had a lipoma encasing the brachial artery, which was not biopsied before operation. In a further three patients biopsy was not performed, as the clinical diagnosis of intramuscular arteriovenous malformation was confirmed using a combination of ultrasound and computed tomography with intravenous contrast.

Eight patients suspected of having STS were referred without biopsy and were found to have malignant tumours other than STS involving soft tissue. The diagnosis was established by Tru-cut biopsy in all patients. These included four patients with extranodal lymphoma of soft tissue without any other focus of disease, three with Ewing's sarcoma of bone presenting as soft tissue masses and one patient with a soft tissue buttock mass which proved to be metastatic adenocarcinoma from a bronchogenic carcinoma subsequently detected by CT scan of the thorax.

Discussion

Soft tissue sarcomas are rare and a high index of clinical suspicion is required before biopsy. An enlarging painless mass, particularly in the proximal limbs and limb girdles, that is deep to the deep fascia, should be considered a soft tissue sarcoma until proved otherwise *(9)*. Regional lymphadenopathy is rare, occurring in about 4% of patients, and indicates an extremely poor prognosis *(11,12)*. No particular age group is affected and the sex incidence is equal. Stigmata of Von Recklinghausen's neurofibromatosis should be sought and a history of previous irradiation and exposure to carcinogens noted.

Patients with soft tissue tumours normally present with a lump but, unfortunately, because of its rarity, soft tissue sarcoma is not always suspected and too often the mass is enucleated on the basis of a clinical diagnosis of lipoma. In this study, of the 54 patients referred for definitive management after biopsy at the referring hospital, 32 (59.3%) had been treated by excisional biopsy (enucleation). This procedure is an unfortunate error but is widespread, as in a collective series of nearly 6000 cases, enucleation to establish the diagnosis was reported in 49% of the cases *(11)*. Enucleation should be avoided in the management of STS because the exact site of the tumour is then unavailable for assessment by CT scanning, which is the basis upon which radiotherapy and definitive surgery are planned *(13,14)*. After enucleation, excision of the tumour bed is usually undertaken as residual tumour will be found in 50% of re-excised specimens *(15)*. We believe that the biopsy method of choice is Tru-cut core biopsy and that this can be performed under local anaesthetic at the time of first presentation. This study and a previous paper *(6)* confirm the high degree of accuracy of core biopsy which provides enough tissue for histological diagnosis and grading. Contrary to the reports of others *(2,4,16)*, large amounts of tissue obtained by incisional and excisional biopsy are unnecessary.

Open incisional biopsy should normally be avoided not only because it is unnecessary but because complications may follow. If performed, care must be taken to site the incision where it can be encompassed easily rather than compromise a subsequent definitive resection. Although a tumour will often act as a natural tissue expander, rapidly growing tumours may compromise the viability of overlying skin, hence an incision placed over the dome of such a mass will be prone to dehiscence and infection, as occurred in this series. Mankin *et al. (3)*, in a series of 329 biopsies, reported a 17.3% complication rate, predominantly involving wound breakdown, haematoma and infection. Furthermore, it has been claimed *(3,17)* that the haematoma accompanying incision biopsy may predispose to local recurrence, but others *(1,18)* have shown in retrospective studies that preliminary biopsy followed by delayed definitive resection does not prejudice survival. 'Punch' incisional biopsy using a trocar and cannula has been advocated as a way of avoiding these complications *(12)*. For these reasons, if open incisional biopsy is required it is usually best planned by the surgeon who will later perform the definitive resection *(4,19)*.

The differential diagnosis of soft tissue tumours should be considered in several categories. Malignant bone tumours involving soft tissue, lymphoma, metastatic adenocarcinoma and a large group of miscellaneous benign soft tissue and bone lesions may all present as a soft tissue mass and mimic soft tissue sarcoma.

Of the nine patients with benign soft tissue lesions submitted to core biopsy, the tissue was inadequate for diagnosis in two cases, and in a further two patients the tissue obtained was suggestive but not diagnostic. Incisional biopsy was therefore required in four patients (44%). Therefore, paradoxically, benign soft tissue lesions are more difficult to diagnose using this technique than malignant lesions.

Eight patients in this series were referred without biopsy with a clinical diagnosis of STS but were proved by Tru-cut biopsy to have malignant tumours other than STS involving soft tissue. Lymphoma and Ewing's sarcoma of bone (and embryonal rhabdomyosarcoma) produce the histological appearance of small round cell tumours but these may be differentiated by immunohistochemical staining *(20)*. In these cases Tru-cut biopsy provided adequate material for this differentiation.

In conclusion, we have highlighted the problems associated with incisional and excisional biopsy of

tumours suspected of being STS. Tru-cut needle biopsy is the preferred technique to establish the diagnosis, and in combination with preoperative CT scanning enables appropriate planning of any subsequent surgery and radiotherapy. An awareness of the differential diagnoses of soft tissue sarcomas is essential.

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Invited comment

This paper describes the diagnostic approach in 118 patients referred to a specialist treatment unit for definitive management. The inadequacy of the initial diagnostic approach for many of the patients is emphasised. A Tru-cut biopsy is the initial approach of choice in a patient with suspected soft tissue sarcoma. This study confirms the high degree of accuracy using this approach and illustrates the disadvantages of open biopsy techniques. Of 54 primary sarcomas referred, a disappointingly high number (32) were referred following enucleation making tumour assessment by computed tomography or magnetic resonance imaging and an appropriate curative surgical approach difficult. If an incisional approach is to be made this should be carried out by the surgeon who will later perform the definitive resection.

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