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Soft tissue sarcomas: are current referral guidelines sufficient?

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ABSTRACT

INTRODUCTION To investigate the adequacy of current early referral guidelines for patients with extremity soft tissue sarcomas.

PATIENTS & METHODS 365 patients with confirmed soft tissue sarcomas were evaluated. Data were collected prospectively and included the length of history and the presence of features in current guidelines suggestive of malignancy (pain, rapidity of growth, depth and tumour size). Statistical analysis included the t-test, ANOVA and the Chi test.

RESULTS Deep tumours were the commonest (306 patients with deep tumours). Pain was the least consistent feature (176 patients with pain). 345 patients with one or more of the guideline features had an average history of 19.86 months, 238 of these were seen after more than 3 months.

CONCLUSION Although the majority of soft tissue sarcomas in our patients had one or more of the clinical guideline features, there was still an unacceptable delay in referring these patients to a specialist unit. The referral guidelines should be modified with special emphasis on depth, which is the most sensitive, followed by size and a history of rapid growth. This combined with increased awareness of these guidelines and a well-advertised, open-access clinic linked to a specialist unit should allow for a more rapid evaluation of soft tissue tumours.

KEYWORDS
Soft tissue – Sarcoma – Guidelines

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Insufficient attention has been given to the symptoms and signs of patients with extremity soft tissue sarcomas. The confusion surrounding these tumours has been compounded by many authors combining soft tissue sarcomas with bone sarcomas. They are two completely different entities.[1] This has led to delays in diagnosis and a failure in many cases to refer patients to specialist units, leading to inappropriate treatment.[2]

Soft tissue sarcomas are rare but up to 2000 patients would be expected to present each year in the UK; of these, 60% would occur in the extremities making them accessible to the examining physician.[3] In order to increase awareness of these tumours and the importance of early specialist input, clinical guidelines have been established describing the features thought to be most commonly associated with malignancy.[4] These features include rapid growth, pain, a diameter of more than 5 cm and location deep to the deep fascia. Despite these guidelines, there are considerable delays in diagnosis, which can adversely affect management and prognosis.

Not all soft tissue sarcomas conform to the guidelines and often those tumours that do not conform may be discounted as they are thought to be benign. The guidelines may be counterproductive and delay referral in some cases.

The purpose of this study was to evaluate the presenting symptoms and signs prospectively, in line with established clinical guidelines, of a cohort of confirmed extremity soft tissue sarcomas presenting to our specialist unit and assess their relative significance in early diagnosis.

Patients & Methods

A total of 365 patients were included in the study; all had a histologically confirmed extremity soft tissue sarcoma. At presentation to our specialist tumour unit, the examining physician completed a standard proforma for each patient. The data collected included the length of history, the presence of pain, rapidity of growth as reported by the patient, and the depth of the tumour. Tumour size was confirmed on surgical specimen.

The features included in the clinical guidelines for early referral of soft tissue tumours were evaluated individually and in combinations to establish the most frequently occurring combinations and their effect on the length of history. Statistical analysis included the *t*-test, ANOVA and the Chi test.

Results

The mean age of the patients was 51.9 years (range, 1–91 years);, 194 were male and 171 female. Of the current guidelines, the depth in relation to the deep fascia was the most consistent, accounting for 306 deep cases (Table 1). Pain was the least reliable of the guidelines, accounting for 176 patients. Looking at the frequency of combinations, those tumours that were deep and more than 5 cm in size accounted for 221 cases; those conforming to all the guidelines accounted for only 92 cases (Table 2).

The average overall delay between the patient becoming aware of an extremity mass and the onset of specialist management (*i.e.* length of history) was 21 months (range, 1–240 months; SD 37.7). Twenty patients (5.4%) had none of the guideline features and had a mean history length of 33.15 months; the remaining 345 patients had one or more of the guideline features with an average history of 19.86 months. The difference in history length between these two groups did not reach statistical significant (*t*-test: $P > 0.1$). If 3 months is accepted as a realistic delay between onset of symptoms and onset of treatment, only 110 patients were seen in the specialist unit within that period, 107 of whom had one or more worrying features (pain, deep, size of more than 5 cm and rapid growth). Of the remainder, 255 patients were seen after more than 3 months, 238 of whom had one or more worrying features.

The individual effect of the guideline feature combinations on the length of history was evaluated (Table 3). The difference between the groups was statistically significant (ANOVA: $P < 0.0001$), revealing that the presence of pain and or a size of more than 5 cm in a rapidly growing deep tumour had the shortest average history, accounting for 160 patients. It is worth noting that pain by itself did not seem to affect the length of history ($P > 0.07$).

Table 1 Frequency of guideline features

Guidelines	Number of patients
Sub-fascial	306
Size > 5 cm	235
Rapid growth	214
Pain	176

Table 2 Combination frequency of guideline features

Groups	Number of patients
Deep + > 5 cm	221
Deep + rapid	186
Deep + pain	165
Rapid + > 5 cm	149
Deep + rapid + > 5 cm	143
Pain + > 5 cm	130
Deep + > 5 cm + pain	126
Rapid + pain	115
Deep + rapid + pain	109
Rapid + pain + > 5 cm	94
Deep + rapid + > 5 cm + pain	92

Discussion

Soft tissue sarcomas are rare and benign soft tissue tumours are common. Often the two present with similar features making clinical diagnosis difficult. Guidelines emphasising aspects that are most commonly associated with malignant tumours can be helpful. However, the present guidelines appear to be misleading, they emphasise certain aspects such as pain although over 50% of the tumours in our patients were painless, and size which is more relevant to prognosis than diagnosis. The right guidelines may have a place. Any lump with a history of rapid growth and situated deep to the deep fascia must be referred and seen promptly.

Rydholm[5] emphasised depth and a size more than 5 cm as well as including 'any lump that was otherwise suspicious' in his Scandinavian guidelines. This resulted in a dramatic increase in referrals to a specialist centre, although no reference was made to the effect on length of history.

Table 3 The effect of guideline features on the length of history

Combinations	No. of cases	Average history (months)
Deep + rapid + pain + < 5 cm	17	6.2
Deep + rapid + > 5 cm + painless	51	6.8
Deep + rapid + pain + > 5 cm	92	7.1
Deep + painless + slow + < 5 cm	20	15.5
Rapid + painless + superficial + < 5 cm	18	19.5
Deep + pain + > 5 cm + slow	34	31
Deep + > 5 cm + painless + slow	44	44.6
Deep + pain + slow + < 5 cm	22	45.1

Combinations with small numbers have been excluded

Guidelines have to be chosen carefully and should be based on features relevant to diagnosis. Depth seems to be the most relevant. Unfortunately, there are a significant number of lumps that are superficial, small and painless, which are malignant. There are also benign tumours, such as desmoid tumours and haemangiomas, which can be very significant locally and cannot be ignored.

Of most concern is that even those tumours conforming to the relevant guidelines have an unacceptably long delay between their initial presentation to the patient and attendance at the specialist unit. This is well in excess of a year in many cases and needs to be reduced. Tumours not conforming to the guidelines seem to be at a further disadvantage with even more delay in referral.

Soft tissue sarcomas are rare and have a variable presentation. The guidelines and the mechanism of referral need to be simplified. The most relevant symptom/sign is their site, deep to the deep fascia. This one feature should be stressed; the emphasis being that such a lump is malignant until proven otherwise. Of course, other features such as a size greater than 5 cm or a history of rapid growth must not be ignored. A patient with a suspicious tumour should be referred to an open-access 'lump and bump' clinic which is part of a specialist sarcoma service. This should expedite referral to the appropriate unit with immediate prognostic benefit.[6]

References

1. Simon MA, Finn HA. Diagnostic strategy for bone and soft tissue tumours. *J Bone Joint Surg Am* 1993; **75**: 622–31.
2. Clasby R, Tilling K, Smith MA, Fletcher CD. Variable management of soft tissue sarcoma: regional audit with implications for specialist care. *Br J Surg* 1997; **84**: 1692–6.
3. Bauer HC, Trovik CS, Alvegard TA, Berlin O, Erlanson M, Gustafson P *et al*. Monitoring referral and treatment in soft tissue sarcoma: a study based on 1,851 patients from the Scandinavian Sarcoma Group Register. *Acta Orthop Scand* 2001; **72**: 150–9.
4. Referral guidelines for suspected cancer, Department of Health, UK <www.doh.gov.uk/cancer/referral.htm>.
5. Rydholm A. Improving management of soft tissue sarcoma (Editorial). *BMJ* 1998; **317**: 93–4.
6. Damron TA, Beauchamp CP, Roughraff BT, Ward WG. Soft tissue lumps and bumps. *J Bone Joint Surg Am* 2003; **85**: 1142–55.