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PRACTICE

EASILY MISSED?

Soft tissue sarcoma

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A 45 year old woman presented to her general practitioner several times over nine months with a gradually enlarging, hard lump in her anterior thigh. An ultrasound scan showed a solid intramuscular mass in the anterior compartment of her thigh, prompting a two week wait referral to the regional sarcoma diagnostic clinic. Magnetic resonance imaging showed a large, heterogeneous intramuscular mass. Core biopsy confirmed a high grade soft tissue sarcoma, and staging computed tomography showed lung metastases. The patient underwent chemotherapy for metastatic disease, followed by resection of her primary tumour, but her disease relapsed and she died 16 months after diagnosis.

What is soft tissue sarcoma?

Soft tissue sarcoma is a heterogeneous group of malignancies. They account for 1% of cancers and have a five year survival of approximately 50%. They can occur at any age (although more than 65% occur in people aged over 50 years[1]) and in any part of the body.

Why is soft tissue sarcoma missed?

Diagnosis of soft tissue sarcoma is often delayed or missed.[2] Delays from presentation to specialist referral can be many months (median 6-14 months).[3 4] Many patient and doctor related factors contribute to delays.[5] Failure to recognise a soft tissue sarcoma is partly due to its relative rarity, with benign lesions (such as lipomas) being 100 times more common. A misconception also exists that malignant soft tissue tumours must be painful and rapidly growing, when they may in fact sometimes grow slowly, be painless, and be located superficially.[6]

Why does soft tissue sarcoma matter?

Delayed presentation results in an increase in tumour size, with poorer prognosis, an increased likelihood of metastases,[7 8] and a lower chance of cure. For every 1 cm increase in size, the chance of cure is reduced by 3-5%.[9]

How is soft tissue sarcoma diagnosed?

Clinical diagnosis

Box 1 outlines red flags that may indicate sarcoma.[10 11]

Investigations

In primary care, ultrasonography can be useful to evaluate superficial lumps,[12] especially if examination findings are equivocal. The National Institute for Health and Care Excellence (NICE) recommends urgent ultrasonography for all unexplained lumps increasing in size, within two weeks in adults and within two days for children and young people.[11] If any red flag sign (box 1) is present, we advise referral for ultrasonography or, if not readily available, referral to a sarcoma centre. If there are two or more red flag signs, consider urgent referral.[9] If three or four red flag signs are present, there is a greater than 80% chance of the lesion being malignant.[9]

If ultrasound findings suggest soft tissue sarcoma, or if they are uncertain and clinical concern persists, NICE recommends urgent referral to a sarcoma diagnostic centre.[11] Referral should not be delayed for investigations.

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This is one of a series of occasional articles highlighting conditions that may be more common than many doctors realise or may be missed at first presentation. The series advisers are Anthony Harnden, professor of primary care, Department of Primary Care Health Sciences, University of Oxford, and Richard Lehman, general practitioner, Banbury

What you need to know

- Consider soft tissue sarcoma with any unexplained lump larger than 4.3 cm (a golf ball) or any lump increasing in size, and undertake urgent ultrasound
- Concerning clinical or ultrasound features of a soft tissue lump should prompt urgent (two week wait) referral to a sarcoma centre

How common are soft tissue sarcomas?

- Approximately 3270 soft tissue sarcomas are diagnosed in England and Wales each year[1]
- They are more common in patients aged over 50 years
- The lower limb is the most common site, followed by the upper limb and then the trunk; head, face, and neck sarcomas occur more often in the under 20s
- The incidence is increasing, possibly owing to improvements in diagnosis and data recording,[1] but possibly also because of an ageing population
- Any unexplained lump, in any site, presenting to a general practitioner, especially if one or more red flag signs are present, needs urgent investigation and/or referral.

Box 1: Red flags for possible sarcoma[10]

- An unexplained lump that is:
  -Bigger than 4.3 cm (size of a golf ball)*,
  -Increasing in size*,[11]
  -Deep to fascia (the mass becomes less obvious on muscle contraction), or
  -Painful
- Recurrence of a previously excised benign tumour

*The first two signs are the most reliable

How is soft tissue sarcoma managed?

Prompt referral of all suspicious lumps to specialist centres where soft tissue sarcomas are managed by multidisciplinary teams is desirable. At a sarcoma centre, next steps include a magnetic resonance imaging scan (fig 1⇓), followed by a core needle biopsy, usually under ultrasound guidance. If biopsy is diagnostic of a soft tissue sarcoma, staging computed tomography follows, to assess tumour load and presence of metastases. Where centres with multidisciplinary teams are not available in the United Kingdom, there is usually a sarcoma diagnostic centre linked to a treatment centre and multidisciplinary team.

Non-metastatic soft tissue sarcomas are typically managed by combination radiotherapy and surgery.[13] Radiotherapy may be induction (before surgical resection) or adjuvant (following surgical resection), with pros and cons for each. Surgery is usually wide excision, with limb salvage, but margins may sometimes be compromised to preserve function. Amputation is sometimes necessary. Chemotherapy is usually reserved for metastatic disease and some paediatric soft tissue sarcomas (usually rhabdomyosarcoma).

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Patient consent: The case is hypothetical, but an anonymised scan from a deceased sarcoma patient has been used to highlight worrying features.

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Figure

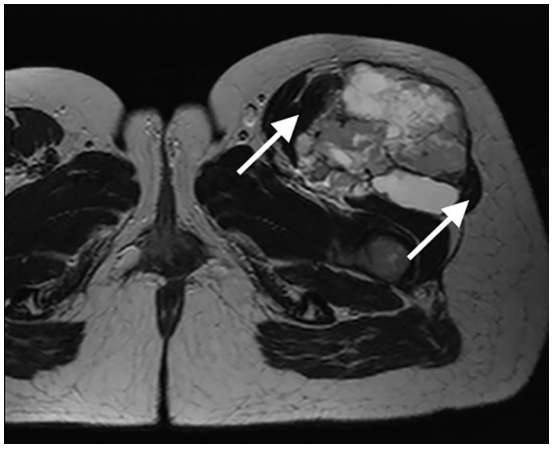

Figure 1 Axial T2 magnetic resonance imaging scan showing large mass in anterior proximal left thigh. Mass shows heterogeneous signal intensity with fluid levels and large areas of necrosis and haemorrhage. Mass lies predominantly in intermuscular plane between rectus femoris (left arrow) and tensor fascia lata (right arrow), with rectus femoris partly infiltrated by mass