Alveolar soft part sarcoma presenting with back pain

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DESCRIPTION

A 35-year-old man was referred to the Sarcoma service with a 2-year history of worsening back pain, left-sided radicular pain and a mass on CT scan. In the 6 months following the onset of the back pain, he developed progressive numbness and paraesthesia in the L2 dermatome. This was worse on standing. On clinical examination, power was unaffected but somewhat limited by pain. No 'red flag' symptoms were present (table 1). The CT scan demonstrated a large retroperitoneal mass on the left with invasion into the L2 vertebra (figure 1 and 2) and bilateral pulmonary metastases. USS-guided biopsy revealed high grade alveolar soft part sarcoma. He was started on palliative radiotherapy (30 Gy and 10 fractions) and given six cycles of doxorubicin and ifosfamide. Imaging at 8 months postdiagnosis has indicated stable disease.

Table 1 'Red flag' symptoms of back pain[4]

Sphincter and gait disturbance	Saddle anaesthesia	Severe or progressive motor loss
Widespread neurological deficit	Age <20 or >55 years	Previous malignancy
Systemic illness	Human inmmunodeficiency virus	Weight loss
Intravenous drug use	Steroid use	Structural deformity
Non-mechanical pain (no relief with bed rest)	Fever	Thoracic pain

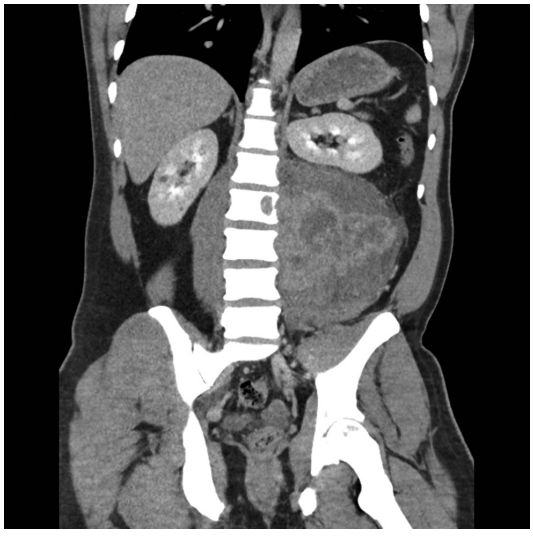

Figure 1 Tumour causing erosive destruction of the left lateral aspect of the L2 vertebrae.

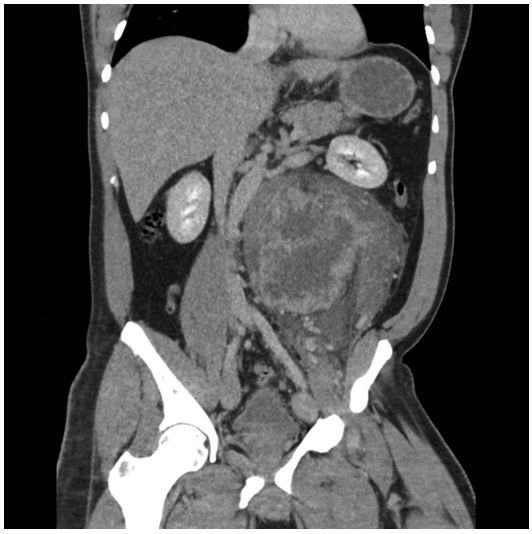

Figure 2 16.4×12.4×19.4 cm mass with central necrosis, closely related to the left psoas muscle.

Alveolar soft part sarcoma is a very rare condition, believed to account for <1% of all soft tissue tumours.[1] Diagnosis is made by a combination of histology and immunohistochemistry; looking for strong nuclear expression of transcription factor 3.[1] Patients are typically young and present with metastases.

Complete wide surgical excision by a sarcoma surgeon is advised in cases where the disease remains localised. Recurrence following complete excision is rare, but metastases may still occur years later.[2] In patients with localised disease survival rates of 60% at 5 years and 38% at 10 years have been documented.[2] Chemoradiotherapy for metastatic disease has been documented in the literature, with limited success.[3]

Learning points

- This unfortunate case serves to remind us that despite the absence of 'red flag' symptoms, clinical suspicion should be raised in the presence of progressively worsening symptoms over a short duration.

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