CASE REPORT

Recurrent solitary fibrous tumour of the thoracic spine. A case-report and literature review

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Summary  Solitary fibrous tumours (SFTs) are rare tumours originating in the soft tissues. SFT development in the spine is an exceedingly rare event about which little is known. We describe a case of SFT of the thoracic spine in a 56-year-old woman. She presented with neurological deficits that required emergency resection, which was incomplete. Two subsequent local recurrences prompted further surgical procedures. At last follow-up, 12 months after the last procedure, function was satisfactory and there was no evidence of tumour recurrence. The management of SFTs is not well standardised, and no proven adjuvant treatments are available to date. Complete excision is effective in controlling disease progression. Prolonged follow-up is mandatory.

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Introduction

Solitary fibrous tumours (SFTs) are rare soft tissue tumours with an annual incidence of 1.4/106 population [1]. The first case was reported in 1931 and originated in the pleura [2], which was subsequently shown to be the most common site of involvement. However, SFTs have been reported at many other sites over the last 20 years, including the mediastinum, lung [3], kidney, thyroid gland, orbit [4], and spine. Aggressive forms with metastases account for less than 20% of cases [5]. The prognosis of SFT depends chiefly on the degree of regional spread and risk of local recurrence.

Case-report

A 56-year-old woman presented in 2006 with rapid-onset pelvic pain radiating to the anterior aspect of both thighs. Her physical examination showed evidence of spinal cord compression with pain in the distal thoracic spine, right T-10 radicular pain, and proximal motor function impairment below the level of compression. Sensation was normal, but partial sphincter function impairment manifesting as urinary urgency was noted. Magnetic resonance imaging (MRI) (Fig. 1) disclosed an extradural tumour located within the
spinal canal at T10-T11 and measuring 30 mm by 52 mm. The tumour caused spinal cord compression with signs of ischaemia (high signal on T2 images and low signal on T1 images). Spinal cord decompression was achieved rapidly by performing T10-T11 laminectomy with partial excision of the tumour.

Histology (Fig. 2) showed a compact proliferation of small spindle-shaped tumour cells arranged in short, intertwined, plexiform bundles with an abundance of blood vessels. The cells contained small amounts of eosinophilic cytoplasm with ill-defined margins; the nuclei were round or oval and contained fine chromatin and small nucleoli. The number of mitoses was only two per ten high-power fields. Immunocytochemistry studies revealed strong labelling for CD34 antibody, as well as cytoplasmic and nuclear PS-100 labelling of about 20% of cells. Immunolabelling was negative with pankeratin markers (AE/AE3 and EMA), vascular markers (CD31 and FVIII), and muscle markers (smooth-muscle alpha-actin, H-caldesmon, and desmin). The final diagnosis was SFT of the thoracic spine.

The neurological manifestations resolved initially then recurred after 6 months. Imaging studies (Fig. 3) visualised a local recurrence. Preoperative embolisation was considered. However, the arteriography showed that the vascular supply originated from the Adamkiewicz artery, contraindicating embolisation (Fig. 4). Surgery was performed for tumour excision and posterior vertebral fusion from T8 to L1. The symptoms recurred 4 years later and were shown by MRI to reflect a large tumour recurrence. The treatment consisted in T10 hemivertebrectomy with additional anterior vertebral fusion via the extended posterolateral approach (Fig. 5). The histological examination showed the same tumour cells with a considerable increase in the number of mitoses to eight per high-power field. No adjunctive treatment was given. No complications occurred after any of the surgical procedures.

At last follow-up 12 months after the third procedure, the patient had no clinical or radiological evidence of recurrence. Function was satisfactory, with no residual neurological deficits.

**Discussion**

SFTs involving the spine were first described in 1996 [6]. We are aware of only 18 previously published cases. Munoz et al. [7] reported that mean age at diagnosis was 46.5 years and that the male-to-female ratio was 1.4/1. The thoracic spine is predominantly affected (56.3%), followed by the cervical spine (31.2%). Only two cases have been described at the lumbosacral level [6,8].

Spinal SFTs are usually intramedullary (58%) or extradural and extramedullary (24%). Only two previous cases of extradural SFTs within the spinal canal have been described. One was a benign tumour developed on the posterior aspect of the L1 vertebral body [8] and the other was located on the posterior aspect of T9 [9].
SFTs produce non-specific clinical manifestations related to slowly progressive compression of the spinal cord or of a nerve root. Imaging studies visualise a soft tissue tumour within the spinal canal. There are no specific radiological features.

The pathological features of SFTs were recently redefined in the new classification scheme for soft tissue tumours developed by the World Health Organisation [10]. The gross examination shows a greyish, well-circumscribed, oval tumour. In some cases, a thin capsule is visible. A patternless histological appearance is characteristic: the spindle-shaped cells are arranged in intersecting bundles associated with a sclerous stroma and blood vessels resembling those seen in haemangiopericytomas. The definitive diagnosis is provided by immunohistochemistry, which shows strong ubiquitous CD34 staining; staining is also positive for Bcl-2, vimentin, and CD99, but these markers are less specific. Staining is usually negative for cytokeratins, smooth-muscle alpha-actin, desmin, PS-100, EMA, and HMB-45.

About 10% to 15% of SFTs exhibit malignant behaviour [11], producing distant metastases or local recurrences. Factors of adverse prognostic significance are location in the abdomen, mediastinum, or retroperitoneal space; high cellularity; a high mitotic index with more than four mitoses per 10 × 400 fields; necrosis; tumour size greater than 5 cm; and negative CD34 staining. Nevertheless, malignant transformation has been reported in patients without any of these factors, even after several years of remission [12]. We are aware of a single reported case of metastases from an SFT involving the spine. The patient had a thoracic SFT with benign histological features [7]. The curative treatment of SFTs relies on surgery, which should be performed immediately with the goal of achieving complete excision of the tumour [13]. The rationale for this strategy lies in the poorer prognosis of recurrences compared to primary tumours. In addition, achieving tumour excision is often more challenging with recurrences than with primaries. In most cases, the treatment of recurrences requires increasingly extensive procedures, particularly regarding vertebral fusion. No adjunctive treatment has been proven effective to date, even in the classical pleural forms. Prolonged follow-up is
indispensable to ensure the early detection of local recurrences and metastases. With these precautions, long-term survival is 70% in patients with pleural SFTs [13].

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References