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CASE REPORT

Metastatic thymoma presenting as spontaneous epidural lumbar haematoma

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Abstract We report the case of a 44-year-old man who was found to have metastatic thymoma to his lumbar spine presenting as a spontaneous epidural haematoma. The man presented with back pain and cauda equina like symptoms in the absence of trauma, antiplatelet or anticoagulant agents. Following a laminectomy and excision of the epidural collection he made a full neurological recovery. Histopathology of the haematoma demonstrated metastatic thymoma. To the best of our knowledge, this is the first such case of metastatic thymoma to the lumbar spine presenting as a spontaneous epidural collection. We believe, in all patients with spontaneous spinal epidural haematoma and a background of malignancy, histopathological analysis should be sought.

Keywords Metastatic · Thymoma · Spontaneous · Epidural · Haematoma

Introduction

Spontaneous spinal epidural haematoma (SSEH) is an exceedingly rare condition with an incidence of 0.1 per 100,000 [1]. A number of potential causes for SSEH have been identified including anticoagulant and antiplatelet use, bleeding diatheses, vascular malformations and hypertension but there have only been four previously described cases where an underlying malignancy has been responsible [2–5]. Three of those cases were haematological malignancies, the fourth metastatic bronchial adenocarcinoma. To the best of our knowledge, this is the first case of an SSEH arising from metastatic thymoma.

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Case report

History and examination

This 44-year-old man presented with a 2-week history of midline lower back pain, gait disturbance, right leg sensory disturbance, bladder discomfort and bowel incontinence. He described claudicant lower limb symptoms for 6 weeks.

The man had a past history of pericardial effusion and locally advanced type B2 thymoma, which were diagnosed concurrently 5 years earlier. He had received 3 cycles of neoadjuvant chemotherapy with cisplatin, adriamycin and cyclophosphamide, after which only partial debulking surgery was possible as the tumour had encased the great vessels. Adjuvant radiotherapy was also administered. He had been disease free in 5 years of follow-up. He denied any previous spinal trauma or procedures. He also denied anticoagulant or antiplatelet use.

On examination there was a mixed distribution weakness of both lower limbs. His ankle power was most affected with absent ankle jerks.

Investigations

Full blood count, electrolytes, coagulation and renal function tests were normal. The patient was afebrile.

A pre- and post-contrast lumbar spine MRI demonstrated a posterior epidural lesion at L4–5 with irregular rim enhancement (Fig. 1). The lesion was 3.5 cm in diameter and causing significant compression of the cauda

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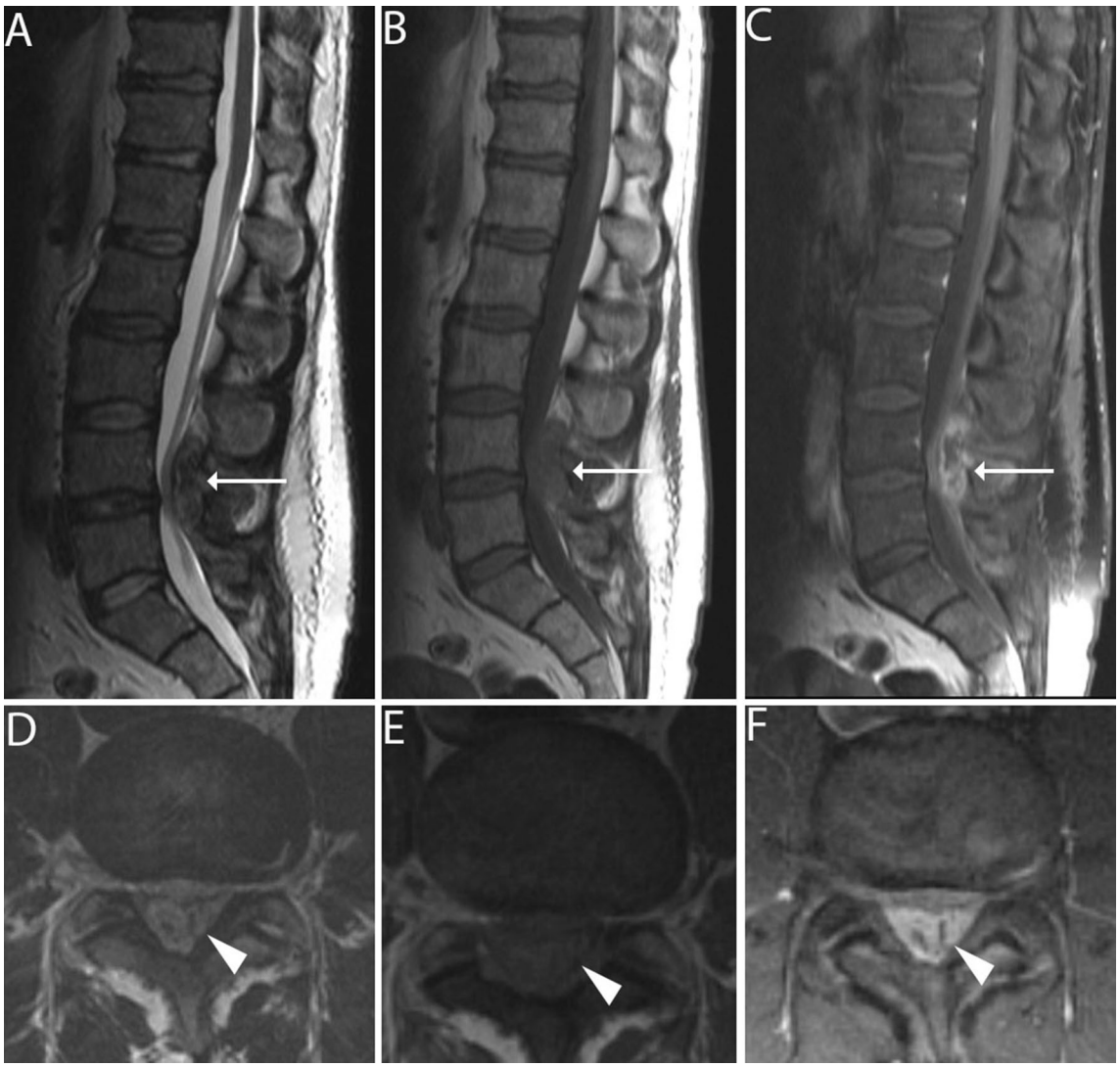

Fig. 1 **a**, **d** Sagittal and axial T2 weighted MRI demonstrating L4/5 epidural lesion with thecal sac compression; **b**, **e** sagittal and axial T1 pre-gadolinium MRI demonstrating L4/5 epidural lesion; **c**, **f** sagittal and axial T1 post-gadolinium MRI demonstrating L4/5 enhancing lesion. *Arrows* in panels **a–c** demonstrating the lesion; *arrowhead* in panels **d–f** demonstrating the lesion and thecal sac compression

equina. The differential diagnosis included epidural abscess, epidural haematoma and neoplasia.

Operation and postoperative course

The patient underwent emergency L3/4 laminectomy and excision of the epidural collection. Intraoperatively an epidural haematoma was visualised at L3/4 with some extension to L5. This was removed in its entirety and sent for histopathological analysis. The dura was opened and no intradural mass was found.

Postoperatively, the symptoms improved and he was able to mobilise with assistance. MRA and autoantibody testing postoperatively failed to identify a cause. The patient was discharged and had outpatient rehabilitation. Unfortunately he was lost to initial follow-up.

Histopathology from the haematoma demonstrated metastatic thymoma (Figs. 2, 3). There was some difficulty contacting the patient but eventually contact was made via his oncologist and the patient had an MRI 11 months postoperatively. This demonstrated metastatic deposits within L2 and L3. The patient remained asymptomatic. He received radiation to his lumbar spine. Subsequently a whole body SPECT/CT detected further metastasis in his left ileum which was also irradiated. He has had further surveillance imaging which demonstrated liver metastases. These were treated with selective internal radiation fields. The patient has remained remarkably well despite his

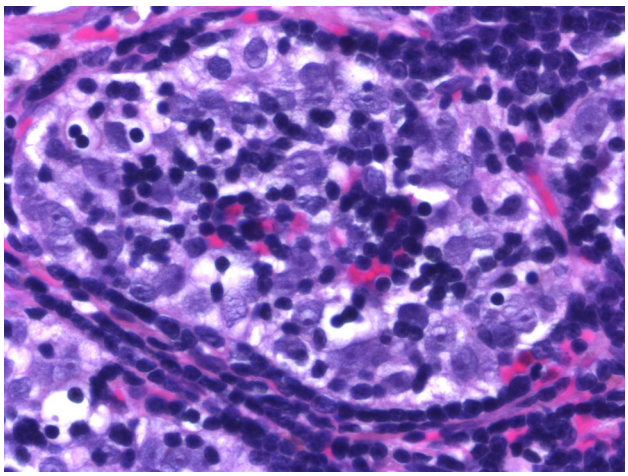

Fig. 2 H&E ×40 reactive lymphocytes and polygonal (cortical) epithelial cells with vesicular chromatin and prominent nucleoli consistent with thymoma

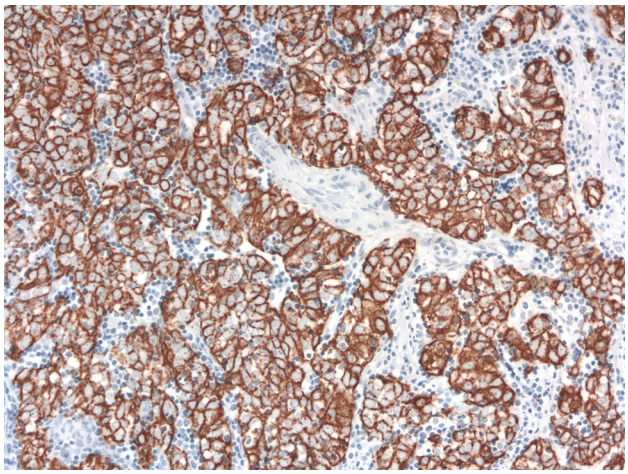

Fig. 3 MNF116 positive immunostaining of the neoplastic epithelial cells for pan-cytokeratin (MNF116) consistent with metastatic thymoma

metastatic disease. At 2 years follow-up he was asymptomatic and had a normal neurological examination.

Discussion

Spontaneous spinal epidural haematoma

First described by Blauby [6], spontaneous spinal epidural haematoma (SSEH) is a very rare cause of spinal cord compression with an estimated incidence of 0.1 per 100,000 people [1]. SSEH can affect all age groups but is most common after 50 years of age [7, 8]. Anatomical distribution of SSEH tends to be predominate in the cervical and thoracic cord with C5 to T2 contributing one-third of the reported segments [1, 8]. Patients present with localised pain and symptoms of spinal cord or nerve root compression. Time to surgery and degree of preoperative neurological deficit are the key prognostic factors [9, 10]; therefore, prompt diagnosis with MRI [11] and early decompressive surgery is the most effective management strategy [1, 9, 10].

A number of potential causes of SSEH have been described including anticoagulant and antiplatelet therapy, vascular malformations, pregnancy, and increases in vascular pressure [8]. Malignancy as a cause has only been described four times before [2–5]. Three of the patients had a haematological malignancy. Two had chronic myelogenous leukaemia with elevated immature white cell and platelet counts [3, 5] and the other had multiple myeloma in the spine with normal blood parameters but had received prophylactic low molecular weight heparin prior to onset of haematoma [2]. The authors suspected that the tumour may have caused epidural inflammation and epidural venous plexus fragility resulting in the haematoma. The possibility of a haemorrhagic microfracture was also considered but not demonstrated on autopsy [2]. The fourth case was a 60-year-old lady who was found to have bronchial adenocarcinoma with thoracic vertebral body metastases as well as an epidural haematoma [4].

Thymoma

Thymomas are relatively uncommon typically slow growing neoplasms of the thymic epithelial cells [12]. They are associated with autoimmune conditions including myasthaenia gravis and systemic lupus erythematosus amongst others, as well as blood dyscrasias, and other malignancies [12, 13]. The Masaoka staging system is the most widely used and defines the extent of tumour invasion into and beyond the thymic capsule and includes metastatic disease [14].

Metastatic thymoma is less common and extrathoracic metastasis even rarer with an incidence estimated between 1 and 6 % [15, 16]. The difficulty in studying extrathoracic metastatic thymoma lies in its rarity, and therefore, it is challenging to characterise. There are reviews examining extrathoracic metastatic thymoma which suggest that liver, bone and lymph node were the most common sites; however, brain, kidney and soft tissue metastasis has also been reported [16–18]. Metastasis of thymoma to the spine causing cord compression has been documented in a small number of cases [17, 19–21]. In some of these instances the spinal metastasis has been isolated, while in others it has been a component of disseminated metastatic disease. None of these cases has been associated with haemorrhage. In fact, metastatic thymoma in general is not associated with haemorrhage. There is only one case of cerebral

metastasis where a haemorrhagic lesion was seen intraoperatively [22]. In the previously mentioned SSEH attributed to malignancy, the mechanism was unclear, possibly due to more fragile epidural venous plexuses [2, 4]. Histopathologically, type B2 thymoma cells tend to palisade around vessels and cause dilation of perivascular spaces [23]. It is possible that in our patient there was vascular invasion resulting in haemorrhage. What is also unclear is the impact that histopathological subtype has on likelihood to metastasise, with Vladislav et al. finding in their series of 35 extrathoracic metastatic thymomas no predominance of a single tumour type [16]. Despite the uncertainty surrounding extrathoracic thymoma metastases there is a good 5-year progression-free survival rate of 41.7 % in Masaoka's group [14].

Management of SSEH and metastatic thymoma

Patients with SSEH typically present with localised pain and symptoms of cord compression. The two management options available are conservative and surgical. When there is complete preoperative sensorimotor deficit, delay in surgery less than 36 h has the best outcome, while in those with incomplete preoperative sensorimotor deficit, surgery within 48 h provided the best outcome [10]. As such once identified on MRI, the management of SSEH is with decompressive laminectomy [9]. This is even more important as surgery provides the opportunity for tissue sampling and the possibility of uncovering the underlying pathology as in our case.

Due to the small volume of cases of metastatic thymoma, a comprehensive management strategy remains elusive [15, 24]. In broad principles, the standard approach involves surgical resection where feasible with adjuvant radiation therapy [24]. In our patient, his tumour had been shown to be radiosensitive in the past, and as such he was given only radiotherapy. His treatment has had good effect from a neurological perspective with no neurological deficits or recurrence at 24 months follow-up.

Conclusion

Spontaneous spinal epidural haematoma is a very rare and rapid cause of spinal cord compression. In patients with a known malignancy, this should be excluded as the cause of the haematoma. Emergency decompressive laminectomy and postoperative rehabilitation are key aspects of management.

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