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Case Report

Primary cervical spinal epidural Extra-osseous Ewing's sarcoma

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Summary

Cases of primary spinal epidural Extra-osseous Ewing's sarcoma (EES) are rarely seen and a good prognosis for EES cannot be expected since a high incidence of local recurrence and metastasis frequently occur.

We present a case of cervical spinal epidural EES in a 7-year-old girl with long survival after tumour resection. She also received adjuvant treatment with peripheral blood stem cell transplantation (PBSCT). This management successfully improved her condition and she was in complete remission without neurological deficit 60 months after surgery. Such a good prognosis of EES in the cervical spine is very rare.

It is likely that magnetic resonance imaging (MRI) lead to a diagnosis of this rare type of EES at an early stage of the disease. PBSCT could be useful as an adjuvant to prolong the period of complete remission.

Keywords: Extra-osseous Ewing's sarcoma; epidural spinal tumour; laminoplasty; peripheral blood stem cell transplantation.

Introduction

Ewing's sarcoma of bone is mostly a childhood malignancy, first reported in 1921 [4]. It is the most frequent type of malignant bone tumour in children under 10 years of age, and its frequency is second in the second decade of life [12]. In 1969, five cases of paravertebral round cell tumours arising in soft tissue were reported in children [9]. On pathological examination, four of them were diagnosed as Ewing's sarcoma (ES) which was only known at that time to develop in bone. The epidural space may have been the site of origin in at least one case. Today, extra-osseous Ewing's sarcoma (EES) in different locations have been reported [8], but the incidence of primary spinal epidural EES is still very rare. Furthermore, diagnostic delay occurred in most cases, so that the prognosis was not as good as expected. Here we report that a patient with primary cervical spinal epidural EES who has a good prognosis treated by surgery and the adjuvant of PBSCT in addition to radio- and chemotherapy.

Case report

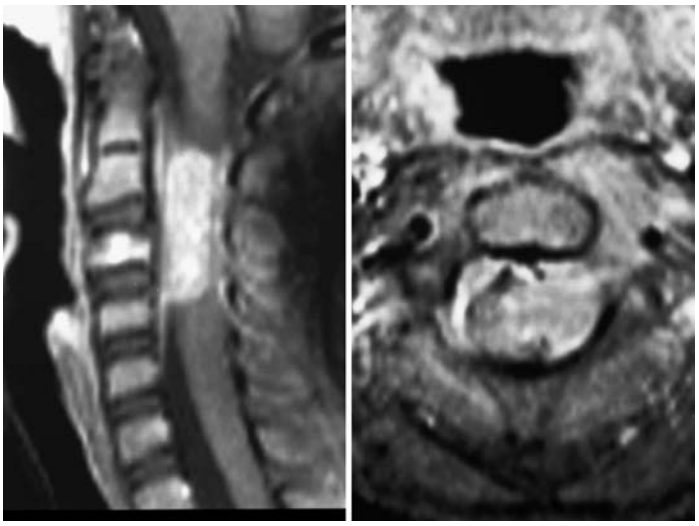
A 7-year-old girl complained of neck pain which had lasted for 10 days. She visited an orthopaedic surgeon. Atlanto-axial rotatory fixation was suspected, so that she was admitted for neck traction. Three weeks after the initial complaint, weakness of the left arm was noticed and the patient was refered to our institute. MRI revealed an extradural tumour at spinal C2-4 level, which displaced the spinal cord laterally (Fig. 1). Neither primary tumour nor metastasis elsewhere in the body were detected. Steroids and glycerol were injected, but weakness of the left arm progressed. On the next day after admission, bladder and bowel dysfunction and weakness of the right arm were observed.

Operation notes

Emergency open-door laminoplasty (C2-5) was performed. An extradural mass of 1.5×3 cm was seen on the left aspect of the spinal cord. The mass had no bone involvement and there was no relation to any exiting nerve root. Using microsurgical technique, it was subtotally resected since it was firmly adherent to the dura.

Histopathology

A photomicrograph of the biopsy specimen from the tumour demonstrated diffuse proliferation of sheets of cells with hyperchromatic nuclei showing little variation in shape and size and with scanty pale cytoplasm. Immunohistochemical staining for MIC-2 gene product showed a strong positive reaction in the tumour cells (Fig. 2). A diagnosis of cervical spinal epidural EES was made.



a b

Fig. 1. MRI reveals an extradural tumour at the spinal level C2-4, which displaces the spinal cord laterally

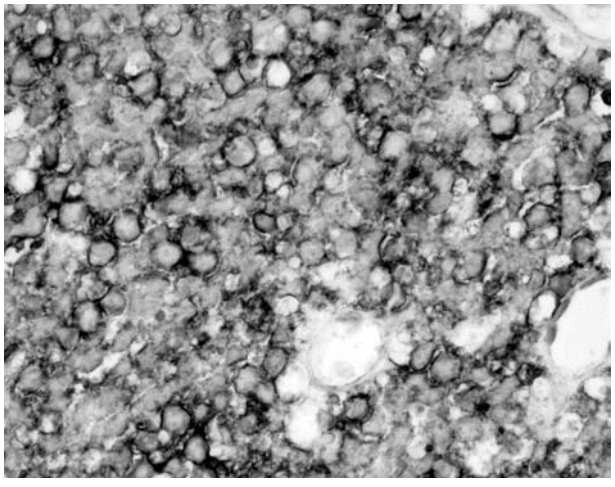


Fig. 2. Immunohistochemical staining for MIC-2 gene product showing strong positive reaction in the tumour cells. (×400)

Postoperative course

Radiation therapy (40 Gy) was given within 2 weeks after surgery and chemotherapy was continued for 5 months including peripheral blood stem cell transplantation (PBSCT) [11]. Postoperative bladder and bowel function recovered rapidly and arm weakness improved completely within 1 month. After 7 months of treatment the patient left the hospital without any motor weakness of the upper limbs. At 60 months after the surgery, the girl was in complete remission with no neurological deficit, although the X-ray showed a cervical S-shaped deformity.

Discussion

The striking morphological and ultrastructural similarities between Ewing's sarcoma of bone (ESB) and Extra-osseous Ewing's sarcoma (EES), and the same translocations involving band q12 of chromosome 22 found in both ESB and EES [2, 10], strongly suggest that ESB and EES are identical tumour types. The incidence of ESB is 1.7 to 2.1 per million people [12]. In one report, 11 cases of EES were found against 51 cases of ESB [7]. The absolute incidence of EES is not known, though it is very rare. Sixteen cases of EES arising primarily in the spinal epidural space are reported in the literature [6], but there are extremely few reports of a cervical location for primary spinal EES.

The mean diagnostic delay was 5.8 months because of the unspecific symptom at onset. Symptoms and signs include back and radicular pain, paresis of one or both legs, sensory disturbance, and bladder and bowel dysfunction caused by spinal cord compression. If the initial symptom is only neck pain, it is very difficult to make a diagnosis without MRI. To diagnose EES accurately, we have to always distinguish it from intervertebral disk herniation, other benign spinal tumours and malignancies [1, 3, 5]. From the histological appearances, lymphoma, rabdomyosarcoma should be excluded and the possibility of melanoma and neuroblastoma should be considered in the differential diagnosis. Immunohistochemical staining for MIC-2 gene product is helpful to diagnose EES from other malignancies.

Furthermore, a good prognosis for EES cannot be expected in view of the high incidence of local recurrence and metastasis. The most frequent sites of metastasis are bone and lung. Kaspers *et al*. reported that 10 of 16 patients (62.5%) died at 16 months after diagnosis of epidural EES on averages [6] and the average of no evidence of disease was 21.5 months. A adjuvant treatment, chemotherapy and radiation therapy are recommended

and have been given in most of the reported cases of EES. These adjuvant therapies are often harmful to the body, especially because haematogenic dysfunction occurs frequently. If a patient has haematogenic dysfunction under treatment, therapy should be suspended until the patient recovers. In our case PBSCT was performed so that we could improve the efficacy of adjuvant therapies. Because PBSCT is very useful in helping the patient recover hematogenous function quickly and easily, that is to say, PBSCT enables treatment to be given with high dose chemotherapy. Our patient was a young child and her physical strength declined after the operation; thus PBSCT was so helpful in allowing effective adjuvant treatment to be given.

In conclusion, if a patient complains of neck pain for a long time and a physical examination elicits neurological deficits, spinal epidural tumor including EES should be considered in the differential diagnosis. Furthermore it is suggested that PBSCT could improve the prognosis of EES treated by adjuvant therapies.

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Comment

The authors present an interesting case study of a rare tumour type, namely Extraosseous Ewing's sarcoma. They believe this to be the first report of such a lesion in the cervical spine. They describe the successful management of this tumour with subtotal surgical resection, radiotherapy and chemotherapy which included peripheral blood stem cell transplantation (PBSCT).

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