A malignant peripheral nerve-sheath tumour developed in the right S1 nerve root in a man aged 30 causing back pain and sciatica. CT and MRI revealed a destructive tumour of the sacrum invading the retroperitoneal space. The tumour was not resectable with an adequate margin. Chemotherapy, consisting of high-dose ifosfamide followed by a combination of vincristine, doxorubicin and cyclophosphamide, was given with success. Malignant peripheral nerve-sheath tumours are thought to respond weakly to chemotherapy, but the response in our patient was complete.

Received 20 November 2002; Accepted after revision 24 March 2003

Malignant peripheral nerve-sheath tumour is a high-grade malignant tumour1 comprising approximately 10% of soft-tissue sarcomas. It tends to recur and metastasise.1 The management is controversial. The main aim is to achieve surgical excision with a tumour-free margin and control systemic spread.1-5 Although aggressive surgery and systemic chemotherapy have been tried, the prognosis remains poor.2-4 Adjuvant chemotherapy is thought to be important but its effects are controversial.2,4 We describe a patient who showed a complete response to high-dose chemotherapy.

Case report

A 30-year-old man complained of increasing lumbar pain with sciatica. Prolapse of the disc affecting the right L5 nerve root was diagnosed and decompression at the L4/L5 level was carried out. However, the symptoms gradually increased and he was referred to our clinic. A plain radiograph of the lumbar spine showed fenestration of the lamina of L4. CT demonstrated destruction of the right S1 root and dural canal of the sacrum (Fig. 1). MRI revealed a destructive tumour of the sacrum which invaded the retroperitoneal space. The tumour had a low intensity of the T1- and a high intensity on the T2-weighted image, suggesting a myxoid tumour of neurogenic or chondroid origin (Fig. 2). An open biopsy was performed which showed a fascicular spindle-cell pattern indicating a malignant peripheral nerve-sheath tumour (Fig. 3). Immunohistochemical examination showed that the tumour spindle cells were positive for S100 protein but negative for cytokeratin, desmin, and α-smooth muscle actin. Resection with an adequate margin was thought to be impossible. We therefore administered systemic adjuvant chemotherapy consisting of alternating high-dose ifosfamide followed by a combination of vincristine, doxorubicin and cyclophosphamide.6 A total of 16 g/m² of ifosfamide was given intravenously and Mesna (uromitexan) was administered at 60% of the ifosfamide. While recovering from the adverse effects of this drug, an intravenous bolus of 1.5 mg/m² (maximum single dose, 2 mg) of vincristine was given, followed by cyclophosphamide at 1800 mg/m² and doxorubicin at 60 mg/m². Since the size of the tumour did not change after this, a further course of chemotherapy was given. After two courses, the tumour decreased in size and became circumscribed with a reactive bone shell (Fig. 4). We then performed a wide resection with excision of the right sacrum and right sciatic nerve which gave an adequate surgical margin. The resected specimens showed complete necrosis of the tumour cells and a tumour-free margin. The patient has been well without recurrence or metastasis for two years since surgery (Fig. 5).
Malignant peripheral nerve sheath tumour is rare and difficult to diagnose and treat. If such spread occurs and an adequate surgical margin cannot be obtained because of the size and site of the tumour, adjuvant chemotherapy is usually advocated. However, its effectiveness is unclear and it has been reported not to alter the survival of patients significantly.

Resection of malignant pelvic tumours is challenging, because of the anatomical complexity. If the tumour is not initially resectable, amputation is usually advocated. Adjuvant high-dose chemotherapy should be considered before surgery. If the response of the tumour to chemotherapy is good it may be resected with an adequate margin. It is well recognised that high-grade adult-type soft-tissue sarcomas have a poor prognosis, which can be improved by effective systemic adjuvant chemotherapy. Recently, high-dose ifosfamide therapy has been reported to be effective in synovial sarcoma.

MRI showing a destructive tumour of the sacrum which had invaded the retroperitoneal space. It had a low intensity on the T1- (a) and a high intensity on the T2-weighted image (b). This suggested a myxoid tumour of neurogenic or chondroid origin.

Photomicrograph showing that the tumour was composed of spindle cells with a fascicular pattern. The nuclei showed atypia and mitosis (haematoyxin and eosin x 25).

CT scan showing a decrease in the size of the tumour and surrounding reactive bone shell after two courses of chemotherapy.

Radiograph showing reconstruction of the right sacrum and absence of recurrence two years after surgery.
sarcoma. Yokoyama et al showed that the alternating use of high-dose ifosfamide, doxorubicin, vincristine, and cyclophosphamide induced a higher response rate than either ifosfamide or doxorubicin alone, and was effective in high-grade soft-tissue sarcomas. Our case shows that high-dose neoadjuvant chemotherapy should be considered in high-grade malignant peripheral nerve sheath tumours and may improve the prognosis.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References

TRAUMATIC SPONDYLOLISTHESIS OF THE LUMBOSACRAL SPINE WITH MULTIPLE FRACTURES OF THE POSTERIOR ELEMENTS

H. Miyamoto, M. Sumi, O. Kataoka, M. Doita, M. Kurosaka, S. Yoshiya
From the Kobe National Hospital, Japan

We describe a patient with a traumatic spondylolisthesis of L5 and multiple, bilateral pedicle fractures from L2 to L5. Conservative treatment was chosen, with eventual neurological recovery and bony union. We are not aware of previous reports of this pattern of injury.


H. Miyamoto, MD, Orthopaedic Surgeon
M. Doita, MD, Orthopaedic Surgeon
M. Kurosaka, MD, Orthopaedic Surgeon
S. Yoshiya, MD, Orthopaedic Surgeon
Department of Orthopaedic Surgery, Kobe University Graduate School of Medicine, 7-5-2 Kusunoki-cho, Chuo-ku, Kobe 650-0017, Japan.

M. Sumi, MD, Orthopaedic Surgeon
O. Kataoka, MD, Orthopaedic Surgeon
Department of Orthopaedic Surgery, Kobe National Hospital, 3-1-1 Nishiochial, Sama-ku, Kobe 654-0155, Japan.

Correspondence should be sent to Dr H. Miyamoto.