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Case Report

The Early Detection of Recurrence of Malignant Peripheral Nerve Sheath Tumor by Frequent Magnetic Resonance Imaging

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Surgery has a key role in the treatment of malignant peripheral nerve sheath tumors (MPNSTs), but the resectability of paraspinal MPNSTs is only 20%. Therefore, spinal MPNSTs show frequent recurrence and poor prognosis. Local recurrence is much more common than metastasis for MPNSTs, and surgery still has a key role in the treatment of local recurrence. Therefore, it is important that recurrence must be detected before resectability is lost. However, no evidence-based follow-up protocol has been established for MPNST. The authors performed gross total resection in a 34-year-old woman presented with thoracic MPNST. Adjuvant radiotherapy and chemotherapy were not administered since these adjuvant therapies generally do not improve survival in MPNST and may cause additional neurovascular damage. Instead, the authors monitored the primary site every 3 months using magnetic resonance imaging to detect local recurrence at the earliest opportunity. The tumor recurred locally on two occasions without overt symptoms at 21 and 24 months postoperatively. These recurrences were treated successfully by gross total removal.

KEY WORDS : Follow-up · Malignant peripheral nerve sheath tumor · Recurrence.

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are rare malignant tumors that originate from diverse cells in the peripheral nerve sheath. MPNST has an incidence of 0.001% in the general population, but the incidence is increased up to 4% among type I neurofibromatosis patients[2]. MPNSTs usually occur in the extremities and nerve plexuses; intraspinal occurrence is much rarer. Magnetic resonance imaging (MRI) alone cannot distinguish MPNSTs from benign schwannomas, and positron emission tomography (PET) may show an elevated metabolic rate in malignant tumors[7]. Surgery is the mainstay of treatment, but only 20% of paraspinal MPNSTs are resectable[3], and therefore, spinal MPNSTs show frequent recurrence and have a poor prognosis. Moreover, although radiotherapy provides local control, it has little effect on long-term survival, and furthermore, MPNSTs are chemotherapy insensitive[3,8]. Local recurrence is more common than metastasis in MPNST[1], and thus, surgery still plays a key role in the treatment of locally recurrent tumors. Accordingly, the early detection of recurrence, before resectability is lost, is very important. Currently, however, there are no evidence-based follow-up protocols available for MPNSTs. Here, the authors report a surgically treated case of MPNST, in which local recurrence was detected twice at an early stage by 3-monthly MRI follow-ups.

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CASE REPORT

A 34-year-old woman presented with paraparesis. A neurological examination revealed leg muscle power of grade II, and urinary and fecal incontinence. Thoracic spine MRI visualized two abutting masses (Fig. 1A, B) in the intradural extramedullary (IDEM) space and in the left side extradural space at the T8 level. There was no evidence of a malignant tumor, and no stigmata of von Recklinghausen's disease were noted.

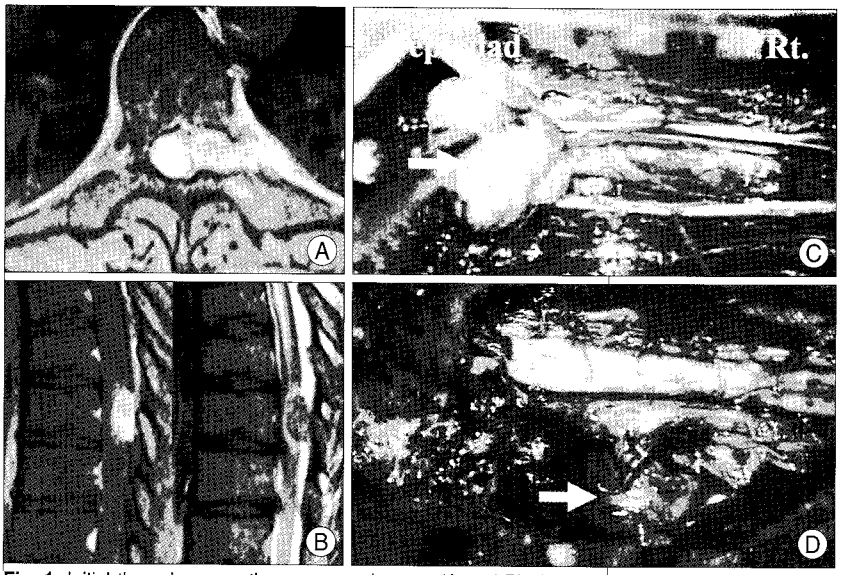

Fig. 1. Initial thoracic magnetic resonance images (A and B) showing a well enhanced intradural extramedullary mass and an additional abutting extradural mass. The intradural mass is located in left side of the spinal cord (C). There is also an extradural mass (D).

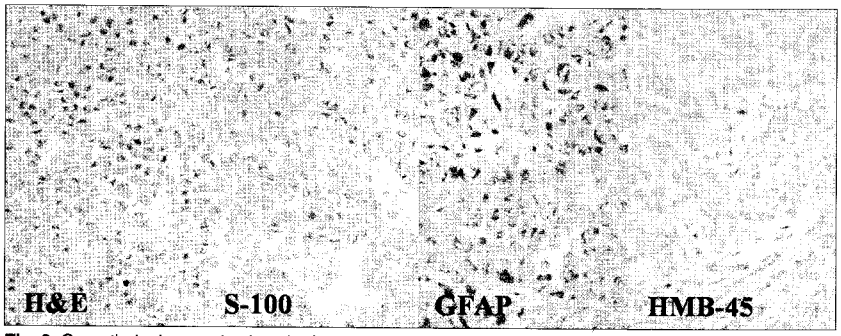

Fig. 2. On pathologic examination, the lesion is compatible with malignant peripheral nerve sheath tumor. Immunohistochemical staining shows positive reactivity for S-100 and GFAP, but negative reactivity for HMB-45.

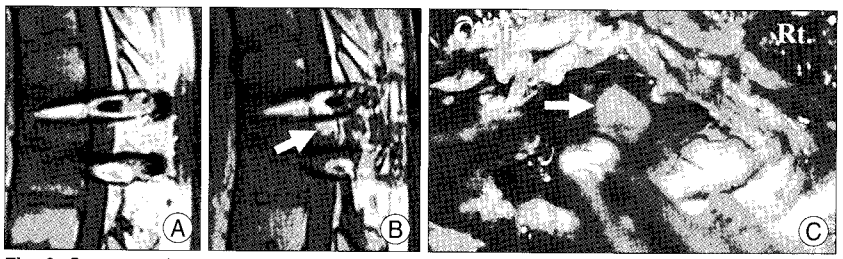

Fig. 3. Contrast enhanced T1 weighted magnetic resonance images: before (A) and after (B) first recurrence. Tumor is located in right side of the spinal cord (C).

The authors presumed that these lesions were schwannomas originating from the left T8 spinal nerve root. After laminectomy, gross total removal of the intradural mass with originating nerve root was performed (Fig. 1C). Intradural and extradural masses were tandem pathology along single nerve root. Frozen biopsy of the excised specimen indicated malignant potential, and thus, the authors removed the extradural mass completely. During the procedure, the left side T8 pedicle was also removed (Fig. 1D). After removal of extradural mass, transpedicular screw fixation was performed at the T8-9 level. Tumors had good surgical interface and showed focal intratumoral bleeding. A subsequent pathological examination concluded that both tumors were MPNSTs (Fig. 2). The patient's paraparesis was not improved postoperatively. Neither radiotherapy nor chemotherapy was ordered because they are known not to improve survival rate and may even cause much neurovascular damage. Instead, the authors performed MRI on a 3 monthly basis at the primary site to detect local recurrence at the earliest opportunity. Twenty-one months after first surgery, local recurrence was detected in the right side (opposite side of the primary site) IDEM space at the T8 level (Fig. 3). This recurrence was substantially asymptomatic; the patient only complained a mild pricking pain in her right flank for three weeks. Gross total removal was performed, and the gross finding was identical to that of first surgery. Three months after this first recurrence, a second recurrence was detected at the left 8th intercostal space along the intercostal nerve, just distal to the neural foramen (Fig. 4A). On this occasion, there were no subjective symptoms associated with the recurrence. The tumor was removed gross totally via thoracotomy (Fig. 4B). Frozen biopsy showed no evidence of malignant cells in the remnant stump, which was composed of intercostal nerve and vessels. The patient has been followed for 14 months with no evidence of recurrence.

DISCUSSION

Only surgery with negative surgical margin has survival benefit in MPNST[8]. Definitive surgery requires a clear tumor margin of at least 2 cm on all sides[3], and this method is usually attempted in lesions which are located in the distal extremities. Occasionally, this is achieved by limb amputa-

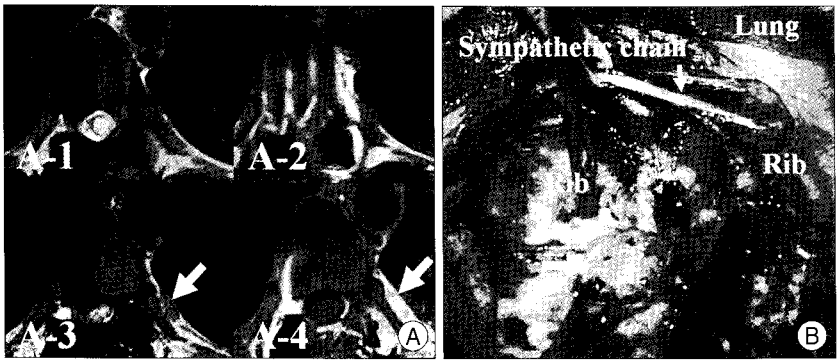

Fig. 4. Magnetic resonance images : before (A-1 and A-2) and after (A-3 and A-4) second recurrence. Tumor is located in the intercostals space.

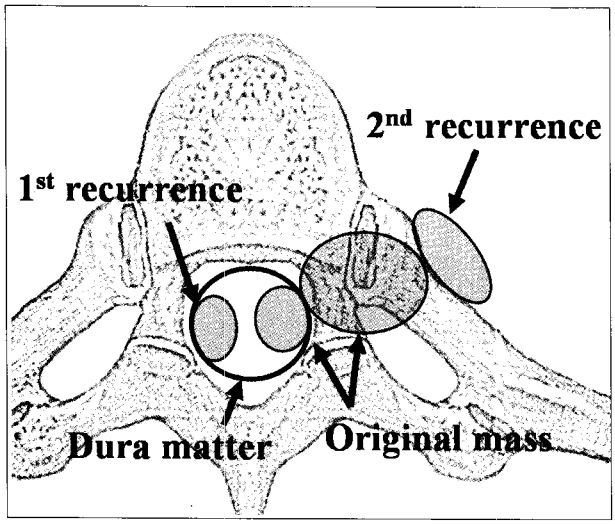

Fig. 5. Schema for tumor locations.

tion. However, in spinal cases, definitive surgery is impossible, and gross total resection often becomes the realistic goal. Fortunately, many spinal MPNSTs have good surgical margins like schwannomas, and gross total removal can be achieved at first surgery[4-6].

MRI has little value in terms of distinguishing MPNSTs from schwannomas, though PET may show an increased metabolic rate in malignant tumors[7], but no consensus has been reached concerning in inclusion of PET during preoperative work-up. Furthermore, even when a tumor is located at a distal extremity and PET reveals hot uptake, definitive surgery is not recommended before a pathological report has been issued, because definitive surgery means severe functional impairment and sometimes amputation. Moreover, definitive surgery is not possible for spinal MPNSTs. Attempting gross total resection is the same on both spinal MPNST and schwannomana, and thus, PET is not regarded as essential component of the preoperative work-up of spinal MPNSTs. When resecting extraforaminally protruding schwannomas, frozen biopsy is essential to elucidate the risk of malignancy. If frozen section findings reveal malignancy potential, the tumor must be removed totally. During first surgery in the present case, the authors extended exposure to achieve gross total removal after receiving a frozen report, which indicated that the tumor had malignant potential. During second revision surgery, the authors biopsied the remained stump to confirm no evidence of residual malignancy.

Although adjuvant radiotherapy increases the local control rate, it has little effect on long-term survival, and MPNSTs are also chemotherapy insensitive[3,8]. MPNSTs frequently recur and local recurrence is much more common than metastasis[1]. Surgery is still the favored modality for treating local recurrence. Therefore, it is important that recurrence must be detected before resectability is lost. In the absence of evidence-based follow-up protocols for MPNST, the authors decided to monitor the primary focus using MRI during follow-up. Recurrence occurred twice during follow-up (Fig. 5). On both occasions, no cardinal symptoms related to recurrence were evident. If the authors waited until radiculopathy had become overt, the tumor might have invaded surrounding tissues, and the opportunity for gross total resection would have been lost.

CONCLUSION

The favoring of local recurrence rather than metastasis by MPNST indicates that frequent MRI at the primary focus can be useful in early detection of recurrence. In the described case of thoracic spinal MPNST, local recurrences were detected twice at an early period by MRI at the primary focus without overt symptoms, and on both occasions, gross total resection was achieved.

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