

Malignant Peripheral Nerve Sheath Tumor with Horner's Syndrome: A Case Report

Masaki Hara, MD,¹ Yasunori Matsuzaki, MD,¹ Tetsuya Shimizu, MD,¹ Masaki Tomita, MD,¹ Takanori Ayabe, MD,¹ Yusuke Enomoto, MD,¹ Shunsuke Wada, MD,¹ Hiroyuki Tanaka, MD,² Hiroaki Kataoka, MD,² Kazuki Nabeshima, MD,³ and Toshio Onitsuka, MD¹

We report on a 42-year-old woman with malignant peripheral nerve sheath tumor (MPNST) arising from the cervical sympathetic nerve. A collar incision and partial sternotomy were performed at the second intercostal space. The mass was spindle shaped and connected to the sympathetic trunk on the cranial and caudal sides, and it compressed the left carotid sheath on the median side. After the patient's uneventful recovery from surgery, adjuvant radiotherapy was administered to the area of resection. The patient remains well 5 years after surgery with no evidence of recurrence. (Ann Thorac Cardiovasc Surg 2008; 14: 246–248)

Key words: mediastinal mass, Horner's syndrome, malignant peripheral nerve sheath tumor

Introduction

Malignant peripheral nerve sheath tumor (MPNST) is an uncommon soft-tissue malignancy. We report on a 42-year-old woman with MPNST arising from the cervical sympathetic nerve. The rare occurrence of MPNST in the sympathetic nerve is described together with a review of the literature.

Case Report

A 42-year-old woman was admitted to our hospital because of Horner's syndrome (left ptosis, miosis) of 6 months duration. She had undergone total gastrectomy as a result of early gastric cancer 4 months earlier. A chest X-ray revealed a left upper mediastinal mass. Computed tomography (CT) (Fig. 1A) and magnetic

resonance imaging (MRI) (Fig. 1B) disclosed cystic and solid components within the mass, which extended into the left thoracic space.

The patient underwent removal of the tumor under general anesthesia. A collar incision and partial sternotomy were performed at the second intercostal space. The mass was spindle shaped and connected to the sympathetic trunk on the cranial and caudal sides, and it compressed the left carotid sheath on the median side. The left vertebral artery was reconstructed with direct anastomosis because it was strongly adhered to the tumor.

On gross examination, the tumor measured 7 × 4 × 2.5 cm and the cut surface of the excised mass was mostly yellowish tan with focal hemorrhage (Fig. 2). On microscopic examination, the tumor consisted of a proliferation of spindle-shaped cells accompanied by hemorrhage, hemosiderin deposition, hyalinization, and cystic change (Fig. 3A). The tumor cells had elongated hyperchromatic nuclei and were arranged haphazardly, or they formed intersecting short fascicles or long wavy bundles. Cellular density was variable. Mitotic figures were occasionally seen [up to 3–4/10 high-power field (hpf)] (Fig. 3B), and the MIB-1-positive rate was 20% in the highest portion. Immunohistochemically, the cells showed variable positivity for S-100 protein and

From ¹Department of Cardiovascular, Thoracic, and General Surgery and ²Section of Oncopathology and Regenerative Biology, Faculty of Medicine, University of Miyazaki, Miyazaki; and ³Department of Pathology, Fukuoka University School of Medicine, Fukuoka, Japan

Received February 26, 2007; accepted for publication June 21, 2007
Address reprint requests to Masaki Hara, MD: Department of Cardiovascular, Thoracic, and General Surgery, Faculty of Medicine, University of Miyazaki, 5200 Kihara, Kiyotake, Miyazaki 889-1692, Japan.

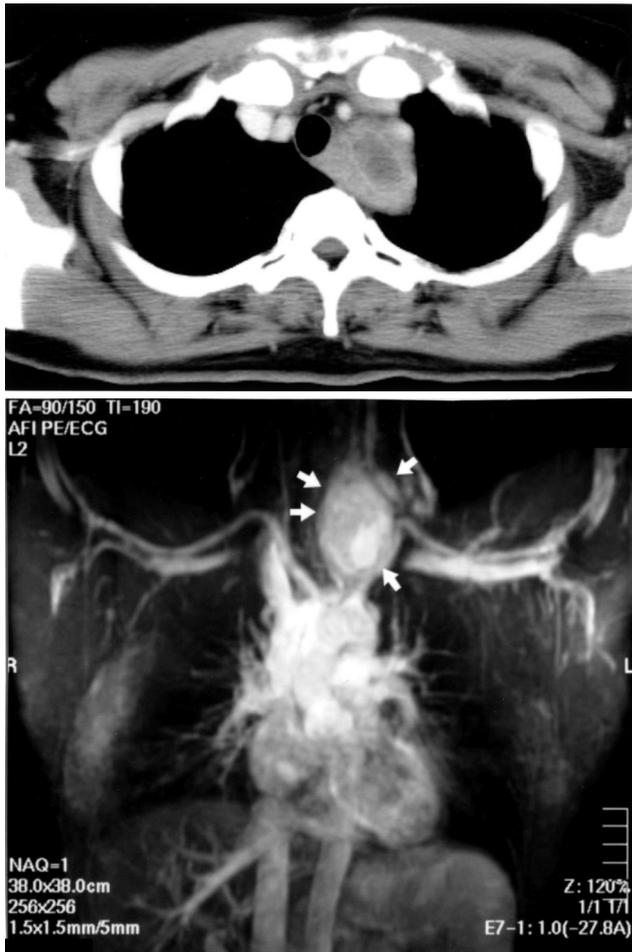


Fig. 1. Computed tomography (CT) (A) and magnetic resonance imaging (MRI) (B) of the chest revealing a left mediastinal mass with cystic and solid components extending into the left side of the chest.

negativity for desmin. Based on these operative and microscopic findings, the tumor was diagnosed as MPNST, low grade, that had probably arisen from the left cervical sympathetic nerve.

After the patient's uneventful recovery from surgery, adjuvant radiotherapy was administered to the area of resection. The patient remains well 5 years after surgery with no evidence of recurrence.

Comment

The MPNST is relatively rare and accounts for 5% of all malignant soft tissue tumors.¹⁾ It is a highly aggressive tumor capable of arising de novo from the peripheral nerve or from preexisting benign neurofibromas. Schwannoma of the cervical sympathetic chain is a rare

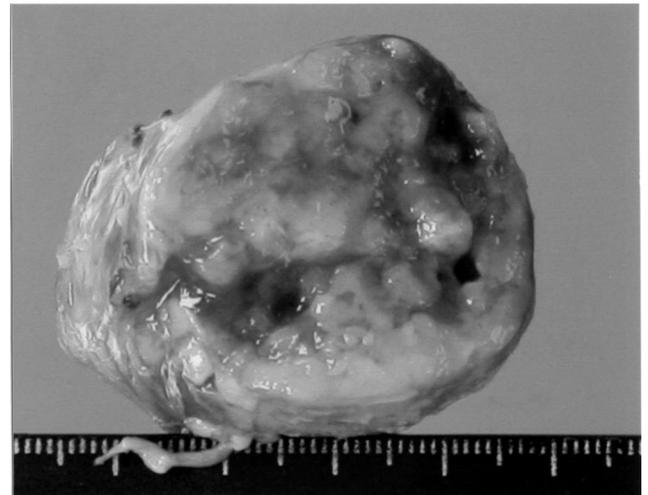


Fig. 2. On gross examination, the tumor measured 7 × 4 × 2.5 cm, and the cut surface of the excised mass was mostly yellowish tan with focal hemorrhage.

nerve tumor with fewer than 40 confirmed cases in the English literature.²⁾ MPNST of the sympathetic trunk is even rarer. MPNST of peripheral nerves is occasionally associated with neurofibromatosis type 1.³⁾ In our case, however, no features of neurofibromatosis type 1 were detected.

The imaging features of a malignant MPNST of the cervical sympathetic chain might be difficult to distinguish from those of a benign cervical sympathetic chain schwannoma. The diagnosis might be suggested on the basis of rapid growth or extensive nerve involvement. Most schwannomas arising from the cervical sympathetic chain present as an asymptomatic solitary neck mass.⁴⁾ Our case developed Horner's syndrome, suggesting the tumor's malignant potential. At histological analysis, MPNSTs are unencapsulated infiltrating tumors composed of spindle cells arranged in a whirling pattern with irregular nuclei, cyst formation, and nuclear palisading.⁵⁾ The diagnosis of a low-grade MPNST was made when atypical features in a plexiform neurofibroma, such as increased cellular density, nuclear atypia, and low levels of mitotic activity, were presented.⁶⁾

The 5-year survival for MPNST is reported to range from 40% to 66%.³⁾ Even with aggressive therapy, a local recurrence of tumor is seen in 50% of patients.⁷⁾ Although a lymphatic spread of MPNST is rare, hematogenous metastasis generally occurs as a late event with spreading to lungs or bone in <33% of patients with

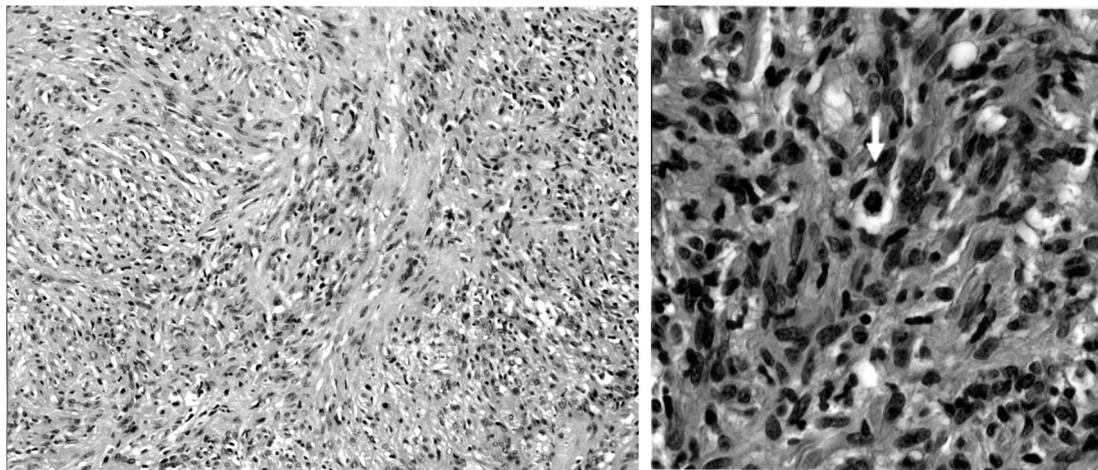


Fig. 3.

A: Histology of the tumor demonstrating a proliferation of spindle-shaped cells (hematoxylin and eosin stain: $\times 40$).

B: Mitotic figures were seen (white arrow, up to 3–4/10 hpf) (hematoxylin and eosin stain: $\times 400$).

A | B

MPNST of the head and neck.⁸⁾ The best outcome for patients with MPNST is expected after local resection combined with adjuvant radiotherapy and chemotherapy.³⁾

References

1. Das Gupta TK, Brasfield RD. Solitary malignant schwannoma. *Ann Surg* 1970; **171**: 419–28.
2. Souza JW, Williams JT, Dalton ML, Solis MM. Schwannoma of the cervical sympathetic chain: it's not a carotid body tumor. *Am Surg* 2000; **66**: 52–5.
3. al-Otieschan AA, Saleem M, Manohar MB, Larson S, Atallah A. Malignant schwannoma of the parapharyngeal space. *J Laryngol Otol* 1998; **112**: 883–7.
4. Hood RJ, Reibel JF, Jensen ME, Levine PA. Schwannoma of the cervical sympathetic chain. The Virginia experience. *Ann Otol Rhinol Laryngol* 2000; **109**: 48–51.
5. Hrehorovich PA, Franke HR, Maximin S, Caracta P. Malignant peripheral nerve sheath tumor. *Radiographics* 2003; **23**: 790–4.
6. Zhou H, Coffin CM, Perkins SL, Tripp SR, Liew M, et al. Malignant peripheral nerve sheath tumor: a comparison of grade, immunophenotype, and cell cycle/growth activation marker expression in sporadic and neurofibromatosis 1-related lesions. *Am J Surg Pathol* 2003; **27**: 1337–45.
7. Murphey MD, Smith WS, Smith SE, Kransdorf MJ, Temple HT. From the archives of the AFIP. Imaging of musculoskeletal neurogenic tumors: radiologic-pathologic correlation. *Radiographics* 1999; **19**: 1253–80.
8. Stone JA, Cooper H, Castillo M, Mukherji SK. Malignant schwannoma of the trigeminal nerve. *Am J Neuroradiol* 2001; **22**: 505–7.