Answer

Malignant Peripheral Nerve Sheath Tumor

During surgery, the tumor appeared to originate from the pelvic bottom with bowel invasion because it was densely adhered there. The postoperative course of the patient was uneventful. Unfortunately, he died 2 years after surgery due to peritoneal and pleural dissemination.

Immunohistochemical examination of the tumor revealed positive expressions of S-100 protein, neuron-specific enolase, and vimentin, while there was a lack of immune profiles of actin, α–smooth muscle actin, desmin, CD34, c-kit (CD117), epithelial membrane antigen, and keratin. Epithelial membrane antigen and keratin are markers derived from epithelial tissues, whereas vimentin is a mesenchymal molecule. These staining results of epithelial membrane antigen (negative), keratin (negative), and vimentin (positive) in this tumor indicated that it was of a nonepithelial origin. On the other hand, S-100 protein and neuron-specific enolase are both neurogenic markers, whereas actin, α–smooth muscle actin, and desmin are myogenic ones. Both CD34 and CD117 are molecules well known to exclude a gastrointestinal stromal tumor. Those results gave definitive immune profiles showing that the tumor was of a neurogenic origin rather than a myogenic one or a gastrointestinal stromal tumor.

The pathological spectrum of peripheral nerve sheath tumors is diverse; they arise from Schwann cells, frequently in association with an existing neurofibroma.[1] Although schwanommas generally have benign behaviors leading to displacement rather than invasion of the adjoining structures, a rare variant termed *ancient schwannoma* mimics malignant features such as microscopic degenerative and heterogeneous changes as well as macroscopic gross appearances.[2] Despite such findings, ancient schwannoma rarely undergoes malignant transformation because its malignant form, malignant peripheral nerve sheath tumor (MPNST), usually develops de novo.[1] Malignant peripheral nerve sheath tumors have the same biological features as undifferentiated sarcomas with locally aggressive behaviors as well as distant metastatic diffusion. They compose 3% to 10% of all soft-tissue sarcomas.[3] Malignant triton tumor is a rare histological variant of MPNST characterized by rhabdomyoblastic differentiation.[4] Both MPNSTs and malignant triton tumors most commonly manifest in the extremities, trunk, or head and neck and have a poor prognosis.[5]

Sometimes MPNSTs reach large proportions before producing symptoms due to mass effects.[5] Preoperative radiologic workup hardly allows discrimination of this entity from other soft-tissue tumors as well as benign forms from malignant diseases unless tumor invasion or metastasis is found.[5] An accurate preoperative diagnosis is essential; however, even benign huge schwannomas including ancient schwannomas may be overdiagnosed as MPNSTs. The occasional presence of cytological atypia as well as hyperchromatic cells may lead to diagnostic difficulty.[6] Fine-needle aspiration rarely produces an adequate sample.[5,6] Even in excisional biopsy it may have results that are not confirmatory, as in our case. This difficulty is also due to the unavailability of definite pathological criteria as well as immunohistochemical markers specific for MPNSTs.[6]

On the basis of our findings, this case was finally diagnosed as abdominal MPNST with invasion of the distal ileum and sigmoid colon rather than malignant triton tumor or ancient schwannoma. Despite evidence of cytological atypia as well as a macroscopic gross and invasive appearance, the mitotic activity was low (1 mitosis/10 high-power fields) and the Ki-67 index was less than 5%. Radical en bloc resection is the mainstay of treatment for MPNSTs because they are usually chemoradioresistant. Survival appears to depend on complete tumor resection. Recent articles have documented that the tumor size of MPNSTs is the most reliable, independent prognostic factor; larger tumors were associated with worse outcomes.[1,7]

In conclusion, abdominal MPNST is a diagnostic and therapeutic challenge. Radical en bloc resection is the treatment of choice for this orphan disease.

Accepted for Publication: February 24, 2012.
Correspondence: Kensuke Adachi, MD, PhD, Department of Surgery, Tokyo Metropolitan Tama Medical Center, 2-8-29 Musashidai, Fuchu, Tokyo, 183-8524, Japan (kensuke_adachi@tmhp.jp).
Author Contributions: *Study concept and design:* Adachi. *Acquisition of data:* Adachi and Takuma. *Analysis and interpretation of data:* Adachi, Enatsu, and Ishizawa. *Drafting of the manuscript:* All authors. *Critical revision of the manuscript for important intellectual content:* Adachi. *Administrative, technical, and material support:* All authors. *Study supervision:* Adachi.
Conflict of Interest Disclosures: None reported.

REFERENCES

1. Pisters PW, Leung DH, Woodruff J, Shi W, Brennan MF. Analysis of prognostic factors in 1041 patients with localized soft tissue sarcomas of the extremities. *J Clin Oncol.* 1996;14(5):1679-1689.
2. Strauss DC, Qureshi YA, Hayes AJ, Thomas JM. Management of benign retroperitoneal schwannomas: a single-center experience. *Am J Surg.* 2011;202(2):194-198.
3. Anghileri M, Miceli R, Fiore M, et al. Malignant peripheral nerve sheath tumors: prognostic factors and survival in a series of patients treated at a single institution. *Cancer.* 2006;107(5):1065-1074.
4. Aldlyami E, Dramis A, Grimer RJ, Abudu A, Carter SR, Tillman RM. Malignant triton tumour of the thigh: a retrospective analysis of nine cases. *Eur J Surg Oncol.* 2006;32(7):808-810.
5. Miguchi M, Takakura Y, Egi H, et al. Malignant peripheral nerve sheath tumor arising from the greater omentum: case report. *World J Surg Oncol.* 2011;9:33.
6. Tortorelli AP, Papa V, Rosa F, Pacelli F, Doglietto GB. Image of the month: retroperitoneal schwannoma. *Arch Surg.* 2006;141(12):1259-1261.
7. Zou C, Smith KD, Liu J, et al. Clinical, pathological, and molecular variables predictive of malignant peripheral nerve sheath tumor outcome. *Ann Surg.* 2009;249(6):1014-1022.