Retroperitoneal schwannoma mimicking lymph node metastasis of seminoma

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RETROPERITONEAL SCHWANNOMA MIMICKING LYMPH NODE METASTASIS OF SEMINOMA

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We report a case of retroperitoneal schwannoma mimicking lymph node metastasis. A 32-year-old man presented with an intrascrotal mass, and underwent radical orchiectomy. Histological examination revealed a typical seminoma of the testis. Systemic work-up for staging demonstrated a retroperitoneal mass, which was located in the suprahilar interaorto-caval region. Under the diagnosis of retroperitoneal metastasis of the seminoma, he underwent three courses of cisplatin, vinblastine, and bleomycin (PVB) therapy. Persistence of the tumor after the chemotherapy led to retroperitoneal lymphadenectomy, histological examination then revealing a benign schwannoma. The location of the tumor and its unresponsiveness to chemotherapy might be useful indicators for differentiating lymph node metastasis of seminoma from primary tumors of other origin.

Key words: Schwannoma, Retroperitoneum, Seminoma, Lymph node metastasis

INTRODUCTION

Schwannoma or neurilemoma is a common tumor of the peripheral nerves, benign schwannomas accounting for 0.3% of retroperitoneal tumors. In most patients with a retroperitoneal schwannoma, symptoms only become manifest when the tumors became large. Only a few cases have been incidentally found during the course of examinations conducted for other diseases. We report a case of retroperitoneal schwannoma, which appeared to be lymph node metastasis at presentation, in a patient with seminoma.

CASE REPORT

A 32-year-old man presented with an intrascrotal mass. Physical examination revealed a solid mass of the right testis, 4x3.5x3 cm. He had no pigmentation as found in von Recklinghausen's disease. Urinalysis, complete blood count, and blood chemistry were normal. He underwent right radical orchiectomy. Histological examination revealed a typical seminoma of the testis. Exploration for metastases by ultrasonography and computerized tomography (CT) demonstrated a retroperitoneal mass, located between the abdominal aorta and the inferior vena cava, cephalically at the level of the renal arteries (Fig. 1). Under the diagnosis of retroperitoneal lymph node metastasis of seminoma, the patient underwent three courses of cisplatin, vinblastine and bleomycin (PVB) therapy. Persistence of the tumor after the chemotherapy led to retroperitoneal lymphadenectomy. Histological examination of the specimen revealed a benign schwannoma of the retroperitoneum. The patient remains well 38 months after the operation.

Pathological findings. The resected retroperitoneal tumor had been located in the suprahilar interaorto-caval region. The tumor was encapsulated, measuring 3.6x2.6x2 cm. Its cut surface was gray-white with some central necrosis and hemorrhage. Histologically, spindle cells with twisted nuclei were observed proliferating in the fibrous stroma (Fig. 2). No nuclear pali-sading was apparent; mitotic figures were rarely present. An immunohistochemical study, employing the indirect peroxidase method previously described, demonstrated the spindle tumor cells to be positive for vimentin (Fig. 3A) and the 8-subunit of S100 protein (S100-8, Fig. 3B). In contrast, the tumor cells showed no staining
Fig. 1. (A) Ultrasonography and (B) computerized tomography demonstrating presence of a tumor (arrowheads) between the abdominal aorta (a) and the inferior vena cava (v).

Fig. 2. Spindle cells proliferating in fibrous stroma (HE x 55)

for the α-subunit of S100 protein (Fig. 3C), desmin, muscle-specific actin, epithelial membrane antigen, or γ-enolase. The histological and immunohistochemical findings thus corresponded to those of benign schwannoma.

DISCUSSION

Schwannoma is a common benign tumor of the peripheral nerves, 45% of benign solitary schwannomas being found in the head and neck, 33% in the extremities, 9% in the soft tissues of the trunk and 14% in various other unusual sites. About 0.7% solitary schwannomas occur in the retroperitoneum with benign schwannoma lesions accounting for 0.3% of retroperitoneal tumors. In the approximately one hundred cases of retroperitoneal schwannoma reported in Japan the sex distribution was equal and the patients were usually between 30 and 60 years of age. Of particular interest was the finding in one report that 16% of the patients with benign solitary schwannoma also had a malignant tumor apparently unrelated to peripheral nerves, although no possible explanation was attempted.

Retroperitoneal schwannomas usually present as large masses with abdominal pain, or with nonspecific gastrointestinal complaints. Tumors are symptomatic only by virtue of their large size. Although ultrasonography and CT incidentally demonstrate a retroperitoneal tumor as in the present case, the examinations do not provide enough information to enable a specific preoperative diagnosis.

About 25% of the patients with seminomas have been found to demonstrate metastasis at presentation. Although semi-
nomas first metastasize to retroperitoneal lymph nodes, suprahilar lymph nodes are rarely involved at an early stage\(^\text{10}\). The location of the presently described tumor and its unresponsiveness of chemotherapy may therefore be useful indicators for differentiating lymph node metastasis of seminomas from primary tumors of other origin.

Schwannomas can usually be easily recognized under the light microscopy. They consist of two components: a highly ordered cellular component (Antoni A area) and a loose, myxoid component (Antoni B type)\(^\text{4}\). Those arising in unusual locations may be confused with other benign and spindle-cell neoplasms, including malignant fibrous histiocytoma and smooth muscle tumors\(^\text{11}\). Immunohistochemical exploration with S100-\(\beta\) is useful for confirming the specific diagnosis\(^\text{12}\), as was shown in the present case.

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和文抄録

セミノーマのリンパ節転移を疑わせた後腹膜神経節腫の1例

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セミノーマ患者にみられ、後腹膜リンパ節転移を疑わせた後腹膜神経節腫の1例を経験したので報告する。セミノーマに対して高位精巣摘除術を施行後、転移検査のために超音波検査と CT を行ったところ、後腹膜腫瘍が認められた。セミノーマの後腹膜転移と診断し化学療法を施行したが、化学療法後も腫瘍の縮少が認められなかったため、後腹膜リンパ節郭清を施行した。病理組織学的検索により後腹膜神経節腫と診断した。精巣腫瘍患者にみられる後腹膜腫瘍についてはその局在部位と化学療法に対する反応性に注意する必要がある。

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