Case Report

A Case of Primary Mediastinal Ependymoma

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A 50-year-old female complained of back pain. Computed tomography showed a well-defined spindle-shaped mass 3.4 × 1.0 cm in size in the left paravertebral posterior mediastinum at the Th 3–4 level. The tumor was completely resected via thoracoscopic surgery, which showed no invasion into the surrounding tissue. It consisted of a cyst with a tiny mural solid element. By means of histological and immunohistochemical examinations, we diagnosed the tumor as ependymoma, and the patient remains alive 59 months after resection. As far as we know, 8 reported cases with primary mediastinal ependymoma have been reported. They had the following specific characteristics: (1) All patients were adult females; (2) The tumors were usually located at the paravertebral upper mediastinum; (3) Most of the tumors consisted of cystic and solid elements; (4) The tumors usually made no invasion into the surrounding tissues, though 2 cases had lymph-node metastases. (Ann Thorac Cardiovasc Surg 2009; 15: 332–335)

Key words: mediastinal ependymoma, immunohistochemical examination, thoracoscopic surgery

Introduction

Ependymomas are well-known tumors occurring in the brain and spinal cord, and they represent 5% and 16% of primary intracranial and spinal cord tumors, respectively.[1)] However, ependymomas located outside the central nervous system (CNS) are rare. Here we present a patient with primary ependymoma located in the posterior mediastinum.

Case Report

A 50-year-old female had complained of back pain since March 2002. In July 2003, computed tomography (CT) showed a well-defined spindle-shaped mass 3.4 × 1.0 cm in size at the left paravertebral posterior mediastinum. Magnetic resonance imaging showed low-signal intensity on unenhanced T1-weighted images (T1WIs), high-signal intensity on T2-weighted images (T2WIs), and no enhancement following contrast media administration (Fig. 1). In August 2003, the tumor was resected via thoracoscopic surgery. Although the tumor bordered on descending aorta, the esophagus, and vertebra, there was no invasion into these organs.

The resected tumor consisted of a cyst, including serous fluid with a tiny mural solid element (Fig. 2). Microscopically, the mural solid element demonstrated trabecular architecture. The cuboidal tumor cells with

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Fig. 1. Magnetic resonance images showed high-signal intensity on T2-weighted images (T2WIs) **(A)**, and no enhancement following contrast media administration on T1-weighted images (T1WIs) **(B)**.
Arrows indicate the tumor.

round/oval nuclei had a cilialike terminal bar at the tumoral side (Fig. 3). In an immunohistochemical examination, the tumor was positive for glial fibrillary acidic protein (GFAP) (Fig. 4), CD 56, vimentin, cytokeratin (CK) 7, epithelial membrane antigen (EMA), estrogen receptor (ER) (Fig. 5), and progesterone receptor (PR). It was negative for chromogranin A, synaptophysin, S-100, and CK 20. From these histological and immunohistochemical findings, the tumor was diagnosed as ependymoma.

Postoperative examination with CT and MRI showed no other tumors in the brain, sacrum, or ovarium. The patient remains alive without tumor recurrence 59 months after surgery.

Discussion

Ependymomas located outside the CNS are rare. Although most are found in the subcutaneous tissue posterior to the sacrum or in the presacral space,[2,3] they have also been reported in ovarian and paraovarian regions.[4,5] Seven cases with mediastinal ependymoma have been reported, and they are summarized in Table 1.[6–10] Including the present case, the clinical characteristics of the 8 cases were as follows: (1) All 8 were females; (2) Mean age was 48 ± 13 years (range: 36–71); (3) All were located at the paravertebral posterior mediastinum; (4) Of the 6 cases in which the levels were described, all tumors but one were located above the upper side of vertebra Th 5; (5) Of the 7 cases in which the cut surface was described, all were a mixture of cyst and solid elements; (6) Lymph node metastasis was seen in 2 cases and metastasis at the

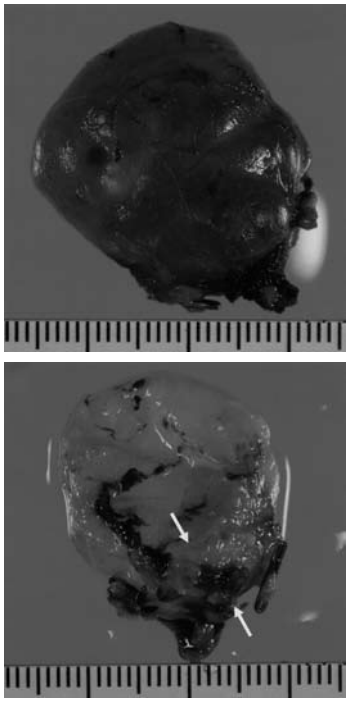

Fig. 2. Gross appearance of the resected cyst, which had a smooth surface and a thin wall **(A)**.
Arrows indicate the solid element in the cyst **(B)**.

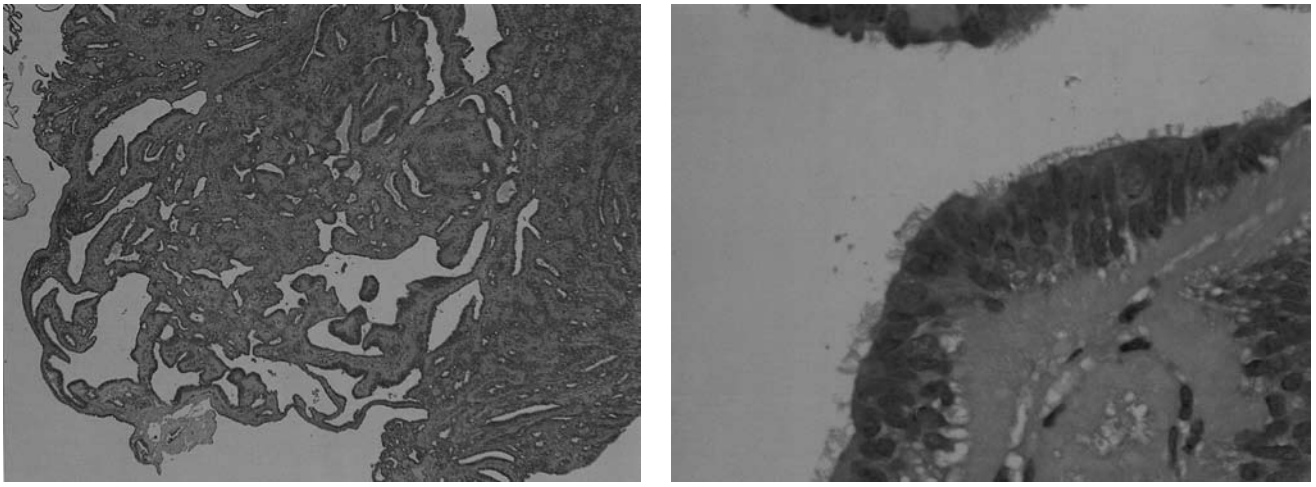

Fig. 3.
A: Histological appearance of the solid element of the tumor with ependymal rosettes and ependymal canals.
B: Cuboidal tumor cells with round/oval nuclei had peculiar cilia at the luminal side.

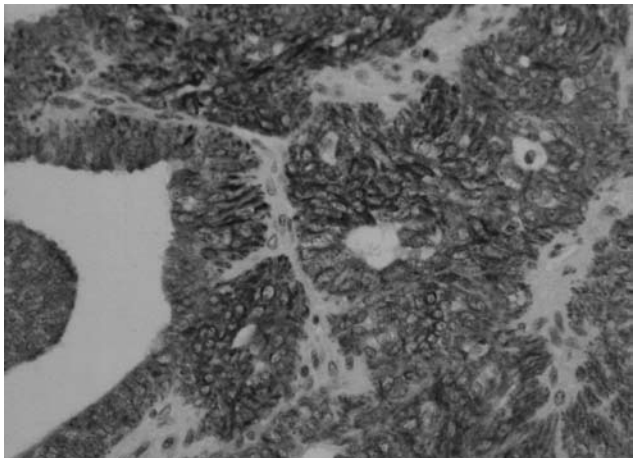

Fig. 4. Glial fibrillary acidic protein (GFAP) immunoreactivity was predominantly observed around tumor vessels.

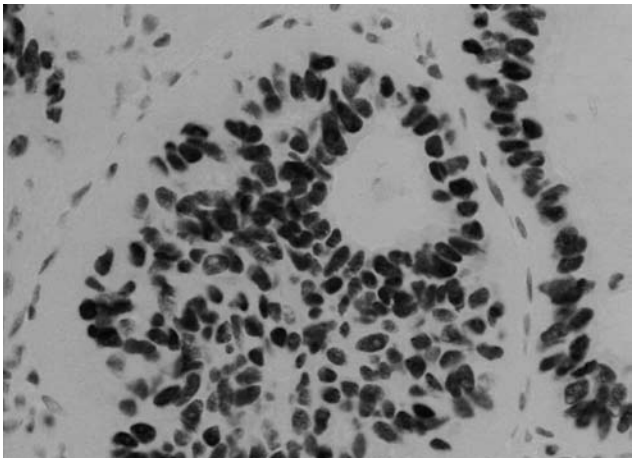

Fig. 5. Estrogen receptor (ER) immunoreactivity was observed in the nuclei in the tumor cells.

Table 1. Clinical characteristics of mediastinal ependymomas

Authors (year)	Age/Sex	Location	Maximum size (cm)	Cut surface	Invasion	Metastasis	Prognosis (months)
1. Doglioni et al. (1988)[6]	51/F	Posterior, upper	4.2	Cyst with solid	–	Lymph node Visceral pleura	4, death by complication
2. Nobles et al. (1991)[7]	36/F	Posterior, upper	9.0	Cyst with solid	–	–	ND
3. Wilson et al. (1998)[8]	36/F	Posterior, upper	7.0	ND	–	–	108, alive
4. Wilson et al. (1998)[8]	71/F	Posterior, ND	5.0	ND	–	Lymph node	12, alive
5. Wilson et al. (1998)[8]	42/F	Posterior, ND	6.0	ND	–	–	72, alive
6. Neumann et al. (1998)[9]	59/F	Posterior, upper	4.2	Cyst with solid	–	–	ND
7. Estrozi et al. (2006)[10]	39/F	Posterior, lower	8.0	Solid	–	–	29, alive
8. Present case	50/F	Posterior, upper	2.8	Cyst with solid	–	–	59, alive

F, female; ND, not described.

pleura of the lung in one; (7) None showed a direct invasion of the surrounding tissues; (8) Of the 6 cases in which prognosis was described, all patients but one were alive without recurrence from 12 to 108 months after resection.

In immunohistochemical examinations, GFAP expression of the tumor cells around the vessels was useful for confirming histological impressions of ependymoma. GFAP was strongly positive in the cytoplasm of the tumor cells in the present case, which contributed to the diagnosis. Recently, Idowu and his colleagues reported differences of immunophenotypes between extra-axial and CNS ependymoma. The percentages of antigen-positive tumors in extra-axial and CNS ependymomas were 80% and 10% in CK 7, 100% and 10% in ER, and 80% and 20% in PR.[11] In the present case, CK 7, ER, and PR were positive, which conformed to the immunohistochemical features in extra-axial ependymoma.

Sacrococcygeal ependymomas are considered to arise from the coccygeal medullary vestige, a remnant of the primitive neural tube,[12] or from heterotopic ependymal cell crests, which are occasionally found in the sacrococcygeal tissue.[13] Because mediastinal ependymomas are usually located in the paravertebral region, they are also considered to arise from misplaced paravertebral ependymal rests.[8]

Although ependymomas of CNS sometimes metastasize, most of them outside CNS are slow-growing low-grade malignancies.[14,15] In mediastinal ependymomas, case Nos. 1 and 4 in Table 1 had each been observed as an asymptomatic posterior mediastinal mass for 16 and 9 years, respectively, which exhibited regional lymph node metastasis at the time of surgery. However, no patients showed invasion into the surrounding tissues. Therefore a complete resection could cure most mediastinal ependymomas.

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