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A Recurrent Cellular Schwannoma

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Cellular schwannoma is an uncommon variant of schwannomas that can occur in a peripheral nerve. Although cellular schwannomas typically do not differ in prognosis from regular schwannomas, they are known to cause local recurrence when not completely resected. Here, we report the case of a patient with cellular schwannoma of the posterior mediastinum, which recurred after 13 years.

Key words: 1. Mediastinal neoplasms
2. Local neoplasm recurrence
3. Neurilemmoma

CASE REPORT

A 49-year-old female was diagnosed with a 7×6×4 cm posterior mediastinal mass on the right side of the 8th, 9th, and 10th thoracic vertebrae. She was asymptomatic. The mass was found incidentally by a routine check-up chest X-ray image (Fig. 1A). Tumor excision was performed through a right posterolateral thoracotomy, and no evaluation of the resection margin was conducted. Histological examination indicated increased cellularity, mild cellular atypism, and low mitotic activity, which was consistent with a benign peripheral nerve sheath tumor. On the basis of these findings, the tumor was diagnosed as a schwannoma. The possibility of recurrence was not considered, and no long-term follow-up surveillance was performed.

Thirteen years later, she was referred to our hospital with an asymptomatic right posterior mediastinal mass. Similar to the previous occurrence, a cellular schwannoma was detected by a screening computed tomogram. The images revealed a 5.6×4.6×2.5 cm lobulated mass at the level of the 9th, 10th, and 11th thoracic vertebrae with the possibility of involvement in the 11th right neural foramen. To determine the correlation between the mass and the spinal nerve, we performed magnetic resonance imaging. The soft-tissue mass originated from the 11th intercostal nerve, extending to the right neural foramen of the 11th and 12th thoracic vertebrae. Due to its heterogeneous signal intensity, the tumor was thought to be a benign neurogenic tumor (Fig. 1B).

Through the previous incision, a right posterolateral thoracotomy was conducted. We found two solid round masses at the paravertebral area of the 10th, 11th, and 12th thoracic spinal bodies. With the exception of the apex area, pleural adhesion was minimal. A neurosurgeon examined the lesion and confirmed that there was no severe extension of the tumor through the vertebral foramen. The two connected masses along with the 11th intercostal nerve were resected with

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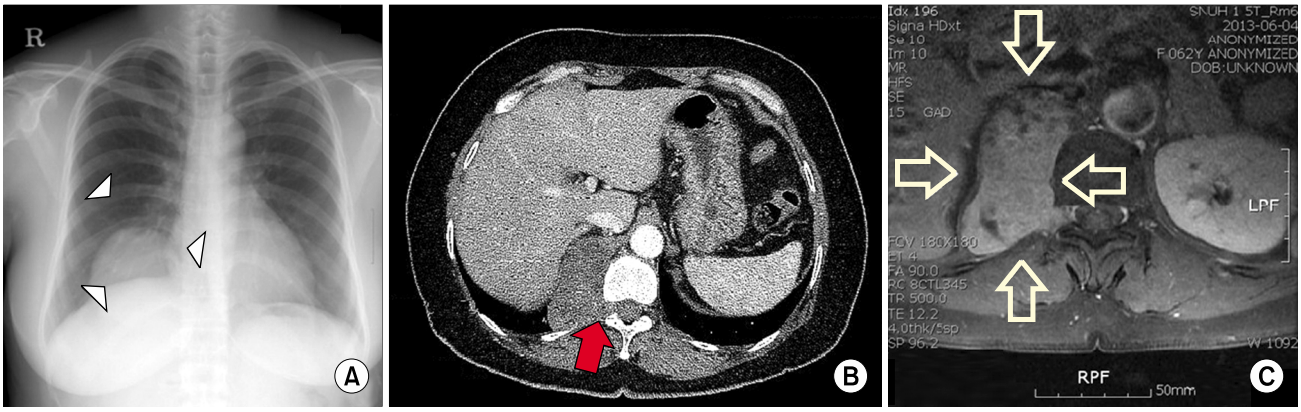

Fig. 1. Image studies from the first and second surgeries. (A) Chest radiogram prior to the first surgery that shows a mediastinal mass (white arrow head). (B) Thirteen years later, a computed tomography scan detected a posterior mediastinal mass (red arrow). (C) A magnetic resonance image reveals a solid mass that originated from the 11th intercostal nerve (white empty arrow).

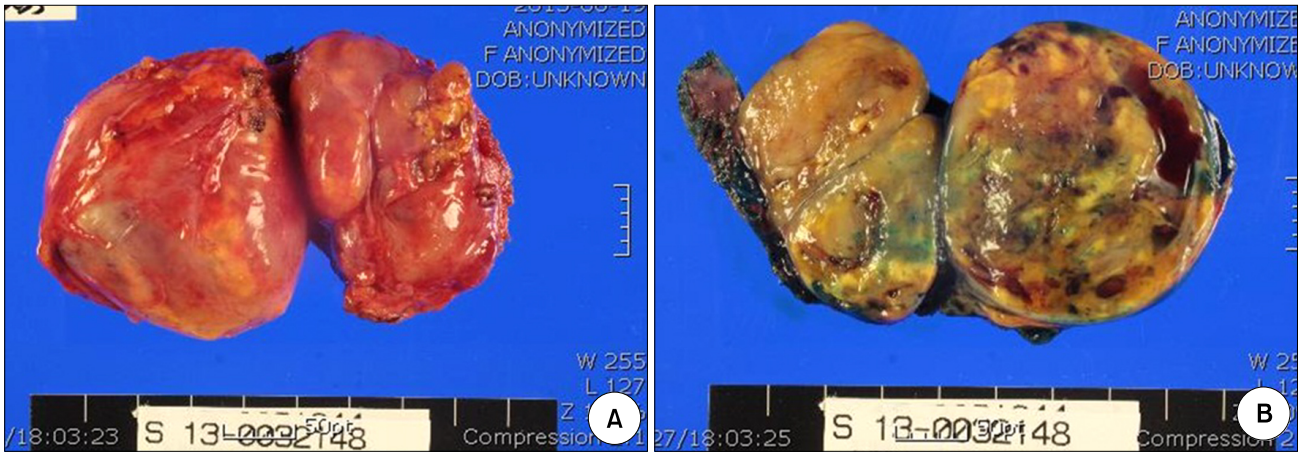

Fig. 2. Macroscopic images of the cellular schwannoma. (A) Well-encapsulated masses connected with the fibrous capsule. (B) Cross section of the tumor that shows partial cystic change.

sufficient margins. An intraoperative frozen biopsy confirmed no involvement of the tumor at the resection margin. Three days later, the patient was discharged without complications.

A gross examination of the specimen showed two well-encapsulated, round masses measuring 4×5 cm each. A cross-section showed thin-walled, firm, slightly yellow solid tumors with a cystic change. The two tumors were connected by a fibrous capsule (Fig. 2). Microscopic examination revealed a markedly increased cellularity and fascicles of spindle cells occasionally associated with a herringbone or storiform pattern. Compact and hypercellular fascicles that recapitulated the Antoni A areas were identified. An immunohistochemical study demonstrated diffuse and strong positivity for the S-100 protein (Fig. 3). On the basis of these findings, the tumors were diagnosed as cellular schwannoma.

Because we could not find the remaining pathologic specimen or microscopic images from the earlier tumor occurrence, a direct comparison between the two tumors could not be performed. However, the pathologic description of the first tumor was consistent with cellular schwannoma. Further, the slight difference between the two lesions may be attributed to the larger size and upward extension of the first tumor. Therefore, we made a clinical diagnosis that the tumors were most likely recurred cellular schwannomas caused by an in-

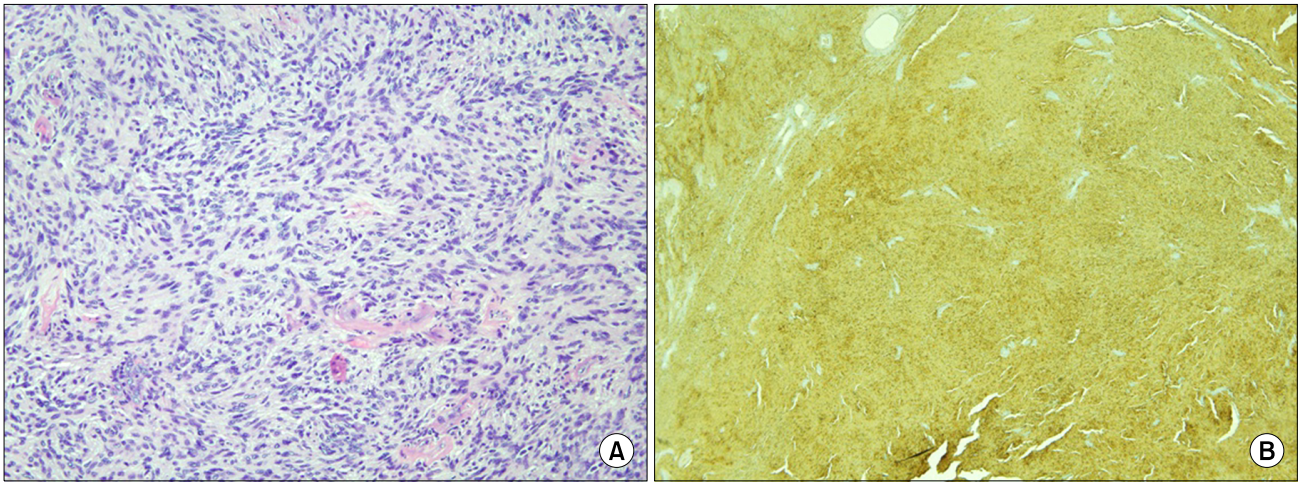

Fig. 3. Microscopic images of the cellular schwannoma. (A) Markedly increased in cellularity with compact and hypercellular fascicles (H&E, ×100). (B) Immunohistochemical study demonstrated strong positivity for S-100 protein (×40).

complete resection performed 13 years earlier.

DISCUSSION

Cellular schwannoma is an uncommon variant of schwannomas, which accounts for approximately 4.6% of the benign peripheral nerve sheath tumors and predominately affects middle-aged adults. It was first described by Woodruff et al. [1] in 1981 as a highly cellular nerve sheath tumor that arises in spinal nerves.

Most cases present as a slowly growing tumor in the paravertebral region of the mediastinum and retroperitoneum [2,3]. Therefore, similar to our case, most cases of schwannomas have no symptoms and are diagnosed incidentally. However, sometimes, when the tumor grows large, a patient may show symptoms related to its mass effect. Chen et al. [4] reported a case of a patient who presented with progressive dyspnea caused by a large cellular schwannoma in the left thoracic cavity that compressed the lung.

Although cellular schwannoma is a well-recognized variant of schwannomas with more than 200 reported cases, it remains difficult to diagnose and can be misdiagnosed as other tumors, such as the well-differentiated malignant peripheral nerve sheath tumor, leiomyosarcoma, fibrosarcoma, melanotic schwannoma, or solitary fibrous tumor [2,3]. In this case, we hypothesize that an incorrect diagnosis was made at the time of the first surgery.

Histologically, cellular schwannoma is a well-circumscribed or encapsulated mass. Signs of hemorrhage are common, but cystic degeneration is uncommon. Cystic change was grossly evident in only 5% to 6% of the patients. Cellular schwannomas differ from ordinary schwannomas because of their prominent hypercellularity composed nearly entirely of Antoni A [2]. The presence of thick fibrous capsules, hyaline thick-walled vessels, and S-100 positive cells in cellular schwannoma enable the distinction from a low-grade malignant peripheral nerve sheath tumor or a leiomyosarcoma [5].

While there have been no reports of metastasis or tumor-related deaths, cellular schwannoma differs from classical schwannoma in its ability to cause local recurrences. The variable mitotic activity appears to be related to the risk of recurrence [6]. In our case, a cellular schwannoma locally recurred and slowly grew until it was identified after 13 years. Its slow growth rate and lack of invasiveness suggest benign features.

Although the intraoperative diagnosis of cellular schwannoma is possible by frozen biopsy [5], histologic confirmation is typically made by permanent biopsy, which occurs long after the surgery. Therefore, it is important to bear in mind the possibility of cellular schwannoma and to ensure a complete resection with sufficient margins. Microscopic confirmation of the resection margin via frozen examination is mandatory in

the case of a close or suspicious macroscopic resection margin. Patients should be informed regarding the possibility of local recurrence in the case of cellular schwannoma.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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