Recurrent intracranial solitary fibrous tumor with cerebrospinal fluid dissemination

Case report

KATSUYOSHI MIYASHITA, M.D., YUTAKA HAYASHI, M.D., HIRONORI FUJISAWA, M.D., MITSUHIRO HASEGAWA, M.D., AND JUNKOH YAMASHITA, M.D.

Department of Neurosurgery, Kanazawa University Hospital, Kanazawa, Japan

Solitary fibrous tumor (SFT) is a benign and rare neoplasm. To date, only 37 patients with intracranial SFTs have been reported. Although a number of the tumors were recurrent and some later underwent malignant transformation, none of these lesions progressed to cerebrospinal fluid (CSF) dissemination. In this paper the authors report a case of SFT in which the lesion recurred several times and ultimately was disseminated by the CSF. The patient was a 63-year-old woman with multiple intracranial and spinal tumors. Fifteen years before this presentation, at the age of 48 she had been hospitalized for resection of a falcotentorial tumor. During the ensuing 15 years she underwent multiple surgeries and sessions of radiation therapy for recurrent lesions. The exclusive location of her tumors in the subarachnoid space at the end of this 15-year period indicates CSF dissemination of the tumor.

The tumor that was resected when the patient was 48 years old and the latest resected lesion were analyzed by performing immunohistological CD34, epithelial membrane antigen, vimentin, S100 protein, and reticulin staining, and determining the MIB-1 labeling index (LI). Most of the results were identical, and both tumors were diagnosed as SFT according to a staining pattern that showed a strong and diffuse positive reaction for CD34. Nevertheless, the authors noted that the MIB-1 LI increased from less than 1% in the original tumor to 13% in the latest tumor. The increased proliferation of MIB-1 indicates that the malignant transformation could have occurred during tumor recurrence with CSF dissemination.

KEY WORDS • solitary fibrous tumor • cerebrospinal fluid dissemination • CD34 • MIB-1

In 1931, Klemperer and Rubin16 became the first physicians to describe an SFT based on their encounter with a spindle cell tumor in pleura originating from mesenchymal cells. In 1996, Carneiro and colleagues6 first described the meningeal SFT in a report on seven cases. To date, 37 cases of meningeal SFT4,6,7,9,17,19,22–24,26–30 and 13 cases of spinal SFT2,5,6,8,11,14,15,17,18 have been reported in the literature. Solitary fibrous tumor is a benign lesion that rarely recurs, and no previous cases of CSF dissemination have been reported. We describe the first case in which a meningeal SFT recurred several times and ultimately progressed to CSF dissemination.

Case Report

History. This 63-year-old woman presented at our hospital with left-sided chest pain and gait oscillation. Fifteen years earlier, at the age of 48 years she had undergone resection of a brain tumor that bordered the tentorium cerebelli. The histopathological diagnosis of the tumor made at that time was fibrous meningioma. Seven years after the first operation, the tumor recurred in the same region. The tumor was again resected and the patient received radiotherapy in which 60 Gy was locally applied. Three years after the second surgery, tumors recurred in multiple regions, including the original site and the foramen magnum. These tumors were resected in a third operation. Three years later the lesions in the foramen magnum grew back. A fourth operation was performed and the patient underwent craniospinal radiotherapy during which 50 Gy was applied from the posterior fossa to the C-5 level. This time, MR imaging revealed dozens of tumors in multiple regions, including the posterior fossa and the spinal cord. The main tumors were resected and stereotactic radiosurgery was performed for residual tumors in the posterior fossa.

Current Examination. An MR image of the brain revealed homogeneously enhancing multiple masses in the posterior fossa and the foramen magnum (Fig. 1). Magnetic resonance images obtained in the cervical and thoracic regions demonstrated homogeneously enhancing masses at T-5, T-4, T-3, and C-7. The tumor located at T-5 severely compressed the cervical cord (Fig. 1c).

Operations. We gave surgical priority to those lesions suspected to be causing the woman’s symptoms and most
closely linked to the prognosis of her disease. At the first operation the tumor at T-5 was located in the subarachnoid space and totally removed. One month later, the patient underwent suboccipital craniotomy and multiple tumor resections of the posterior fossa. All these tumors were located in the subarachnoid space and did not adhere to surrounding tissues. The tumor at the front of the pontomedullary junction encased the inferior cranial nerves. Although this tumor was not adherent to the nerves, the portion of the tumor situated beyond the nerves was left untouched to avoid nerve injury. After the second surgery, stereotactic radiosurgery was performed to eliminate the residual portion of that lesion.

**Histopathological Findings.** The lesions were primarily...
Disseminated intracranial solitary fibrous tumor

The specimen was strongly and diffusely positive for CD34, positive for vimentin, and nondiagnostic for both EMA and S100 protein. Areas surrounding tumor clusters demonstrated reticulin staining. Based on these findings, we diagnosed the lesions as SFTs. The MIB-1 LI was 13%. We immunohistochemically analyzed the tumor specimens that had been obtained at the first operation 15 years earlier. Nearly identical immunohistochemical staining patterns were found, and the previous tumor was also diagnosed as an SFT. The MIB-1 LI of the tumor obtained at the first operation, however, was less than 1% (Fig. 3).

Postoperative Course. Sequential MR images revealed gradual growth of residual tumors in the cervical and thoracic spine. The patient refused additional surgery and instead underwent radiotherapy during which 40 Gy was applied to spine lesions from C-6 to T-8. Several months have passed since she underwent radiotherapy and her symptoms have not progressed.

Discussion

Solitary fibrous tumor is a benign lesion. Metastatic SFT is extremely rare; only one previous case has been reported. Furthermore, neither multiple intracranial nor spinal CSF dissemination of the SFT has been described until now. Our patient is the first case of a documented SFT with CSF dissemination. In this patient 8 years intervened before the first recurrence, but intervening periods to later recurrences gradually shortened. During the patient's latest hospitalization, her symptoms progressed and her tumors grew rapidly. Ultimately, the tumor acquired the ability to multiply and progress to malignancy in the process of recurrence. The increased proliferative activity along with the malignant transformation of this disease was supported by the change in the MIB-1 LI. The MIB-1 LI rose from less than 1% at the first operation to 13% at the latest operation. Although malignant transformation due to irradiation has been described in various cases of intracranial tumors, it has never been reported in cases of SFT. Irradiation may induce tumor cells to become malignant with an increased MIB-1 LI; however, in this case there was no direct evidence to support such a hypothesis. Previously, five cases of local recurrence and two cases of malignant SFT have been reported. The documented histopathological features of malignant SFT have included a thick chromatin concentration, nuclear pleomorphism, and a high mitotic rate. Our case also exhibited hypercellularity and an extremely high MIB-1 LI. Typical cellular pleomorphism and nuclear atypism, however, were not present.

The SFT is usually a benign spindle cell tumor and thus a differential diagnosis should include fibrous meningioma, hemangiopericytoma, schwannoma, fibrosarcoma, myofibroblastoma, and others. Hemangiopericytoma and meningioma are especially important possible choices for the differential diagnosis. 1) Microscopically, an SFT is composed of fascicles of spindle cells and rich collagen fibers existing between individual cells. 2) The whorl formations and psammoma bodies often seen in meningioma are absent. 3) The SFT displays a negative reaction for EMA, whereas the reaction usually is positive in meningiomas. 4) The SFT exhibits a negative reaction to S100 protein immunostaining, which usually induces a positive reaction in schwannomas. 5) In hemangiopericytoma and schwannoma, the reticulin staining demonstrates a dense pattern that indicates staining of areas surrounding individual tumor cells. In an SFT, the staining can be observed around tumor clusters, but not around the cells themselves. 6) The SFT is strongly and diffusely positive for CD34 staining. In hemangiopericytoma and fibrous meningioma there is a tendency for the reaction to be only partially positive and the staining weak. The CD34 staining pattern is very important for the differential diagnosis.
both EMA and S100 protein. The surroundings of the tumor clusters were stained by reticulin staining. Based on these findings, we concluded that the diagnosis of SFT was reasonable. A molecular biological technique such as comparative genomic hybridization would have allowed us to differentiate the SFT from a hemangiopericytoma. Unfortunately, we were unable to perform this analysis, because all the specimens had been embedded and fixed in paraffin and the DNA that was extracted exhibited a high level of degradation. Nevertheless, an advanced diagnostic tool such as genomic hybridization may be effective for cases in which the differential diagnosis is difficult.

Conclusions

We have described the first documented case of an extremely rare meningeal SFT that progressed to malignant transformation and CSF dissemination over a course of repeated recurrences. Because SFT is an unfamiliar entity among meningeal tumors, we suspect that some SFTs may have been misdiagnosed as meningiomas or hemangiopericytomas in the past. If such misdiagnoses have occurred, some tumors could have been recurrent and disseminate, such as the one we describe. Meticulous clinical and pathological observation is required.

References


Manuscript received October 21, 2003. Accepted in final form July 15, 2004.

Address reprint requests to: Yutaka Hayashi, M.D, Department of Neurosurgery, Kanazawa University Hospital, 13-1 Takaramachi, Kanazawa, Japan. email: yuh@ns.m.kanazawa-u.ac.jp