Metastatic solitary fibrous tumor of the meninges

Case report


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Solitary fibrous tumor (SFT) is a unique tumor composed of interstitial dendritic cells that was first described in the thorax and subsequently reported in diverse organs. Extrathoracic SFTs are predominantly benign but rare malignant cases have been documented. In the nervous system, SFT has been described as a meningeal lesion although all 14 previously reported cases were benign. The authors report the first case of a meningeal SFT occurring in a 55-year-old woman. The tumor first presented as a meningeal lesion that after three recurrences over a 10-year period metastasized to the soft tissues and lungs. The potentially malignant nature of cranial SFTs, especially those with atypical histological features and high mitotic counts, should be recognized.

KEY WORDS • meningeal tumor • metastasis

Solitary fibrous tumor was first described in the pleura by Klemperer and Rabin in 1931. Solitary fibrous tumor is characterized histologically by fascicles of spindle cells intermingled with bands of collagen matrix. The histogenesis of this tumor is debatable, although it is thought that it may be related to CD34-positive interstitial dendritic cells. In the past decade, SFT has increasingly been recognized in many other sites outside the thoracic cavity, especially in the soft tissue. Atypical or malignant SFTs are often encountered in the thorax but extrathoracic malignant SFTs are much more rare. In 1996 SFT of the meninges was first reported as a lesion distinct from fibrous meningioma. To our knowledge, 14 SFTs of the meninges have been described in the literature. We describe the 15th case, which unfortunately was malignant and metastasized to the lungs and soft tissues; it represents the first reported malignant SFT of the meninges.

Case Report

History. This 55-year-old woman had a history of right cerebellar meningioma, which was excised in 1989, followed by fractionated radiotherapy. The tumor recurred within 2 years and was treated by repeated excision. Three years later, the tumor recurred again and was treated this time with gamma knife surgery. The patient subsequently presented to our hospital in April 1997 with increasing clumsiness of the right hand.

Abbreviations used in this paper: CT = computerized tomography; EMA = epithelial membrane antigen; HPC = hemangiopericytoma; hpf = high-power field; MR = magnetic resonance; SFT = solitary fibrous tumor.

Examination and Operation. Admission MR imaging demonstrated recurrence of a 4.5-cm meningioma at the junction of the right occipital bone and the petrous bone (Fig. 1). Small cystic areas were noted in the tumor. A total excision of the lesion was performed, and the tumor was found to be attached to the tentorium with a good plane of cleavage from adjacent brain. The patient made a good postoperative recovery. The results of pathological examination indicated a fibroblastic meningioma. Clinical and pathological materials obtained in the previous operations were not available for review.

Reoperation and Adjuvant Therapy. The patient presented again in December 1998 with a rapidly enlarging mass of the right posterior neck. A CT scan demonstrated a 5-cm contrast-enhancing right paraspinal mass located underneath the right trapezius muscle from C2–4 (Fig. 2). Repeated CT and MR studies did not demonstrate recurrent intracranial tumor. A preoperative chest x-ray film revealed multiple cannon ball lesions (Fig. 3). Excision of the mass was performed and CT-guided fine-needle aspiration of the lung nodules was accomplished. On review of the histological findings for the neck tumor, the results of fine-needle aspiration of the lung lesions, and the intracranial tumor tissue resected in 1997, the diagnosis was revised to SFT of the meninges with metastases to lungs and neck. Chemotherapy with 5-fluorouracil and interferon-α-2b was initiated. After two cycles, the patient refused further chemotherapy treatments because of side effects.

Postoperative Course. Five months after excision of the neck mass, the patient noticed a small subcutaneous lump in her right chest wall. Fine-needle aspiration did not yield sufficient materials for diagnosis. Despite the clinical suspicion of further metastatic disease, the patient refused ad-
ditional treatment and examinations. She was last seen at a follow-up examination 11 months after excision of the neck mass and was relatively asymptomatic.

**Histopathological Findings.** Pathological specimens obtained during the excision of the posterior fossa tumor in 1997 consisted of a discrete, globular mass 4 cm in diameter attached to a thin piece of dura. Histologically, the tumor showed mostly sclerotic tissues with slender fascicles of spindle cells haphazardly disposed and intimately intermingled with bundles of dense hyalinized keloidlike collagen. Focally, some staghorn and arborizing vessels were seen (Fig. 4 upper and center). In one area, the tumor became hypercellular with spindle cells. Cellular pleomorphism was moderate but mitoses in this area were between three and four per 10 hpf. The soft-tissue mass in the neck that was subsequently excised was well circumscribed and showed no invasion into adjacent muscles. A uniform sarcomatous pattern similar to the hypercellular region of the intracranial tumor with long cellular fascicles of spindle cells was seen (Fig. 4 lower).

At the time of diagnosis of the first tumor, no special staining was performed. Subsequently, both the cranial and neck tumors were examined with special stains; MIB-1 labeling was 3% in the brain tumor and 5% in the neck tumor. As seen in Fig. 5, both tumors showed cytoplasmic positivity to CD34 (dilution 1/30; Novacastra, Newcastle-upon-Tyne, UK). Tumor cells were negative for EMA, S-100 protein, desmin, cytokeratin, and factor VIII–related antigen. The positive controls were as follows: CD34 and factor VIII–related antigen, internal controls with vasculature; MIB-1, a glioblastoma multiforme; EMA and cytokeratin, a colorectal carcinoma; S-100 protein, a schwannoma; desmin, normal skeletal muscle. For negative controls, primary antibodies were replaced with 5% normal rabbit serum, the other steps being identical to the test staining. Reticulin staining showed a slight increase in fibers but not the pericellular deposition characteristic of HPC. Fine-needle aspiration of the lung nodules showed clusters of moderately pleomorphic spindle cells similar to the cranial and soft-tissue tumors. Electron microsco-

**Fig. 1.** Axial T1-weighted MR image demonstrating a contrast-enhancing tumor at the junction of the right petrous and occipital bones. Small cystic areas were observed in the tumor.

**Fig. 2.** Axial CT scan demonstrating a 5-cm contrast-enhancing tumor in the right paraspinal region.

**Fig. 3.** Chest x-ray film obtained in 1998 revealing cannon ball lesions in both lung fields.
sampling. The tumor cells were positive for CD34, and MIB-1 labeling was 3%.

Discussion

Solitary fibrous tumors were first described in the pleura but have now been documented in virtually every organ. Close to the central nervous system, occurrence in paracranial sites, such as orbit and paranasal sinuses, has also been described. The histological features of SFTs form a spectrum but the essential characteristics are clear circumscripton; alternating hypercellular foci and hypocellular sclerotic foci; short spindle or ovoid cells in a haphazard, storiform, or fascicular arrangement; and intimate intertwining of thin or thick collagen fibrils with spindle cells. Immunoreactivity to CD34 is usually a prominent feature. CD34 antigen is a 110-kD transmembrane cell surface glycoprotein originally described as a marker for human hematopoietic stem cells and is now widely used as a vascular marker. Normally, a subpopulation of stromal cells or fibroblasts labeled as dendritic interstitial cells stains for CD34 and it is likely that SFT represents the neoplastic counterparts of these CD34-positive dendritic interstitial cells. However, CD34 staining can be lost in high-grade foci in SFTs.

Within the thorax, approximately 13 to 23% of pleural SFTs are malignant, manifesting as local invasion, local recurrence, intrathoracic spread, or distant metastasis. However, the vast majority of extrathoracic SFTs seem to follow a benign course and only rare recurrence or metastasis has been documented. We propose histological criteria for malignancy as follows: more than four mitoses per 10 hpf, nuclear atypia, hypercellularity, and necrosis. In these atypical and/or malignant cases MIB-1 labeling has been highly variable. The malignant tumors also tend to be big, often in excess of 10 cm. At least in thoracic SFTs, lesions that are infiltrative, especially those with atypical features and mitoses, are more likely to behave in an aggressive fashion. However, the clinical behavior of extrathoracic SFTs is unpredictable. Some reported cases were histologically atypical or malignant but clinically there was no recurrence or metastasis. Also, recurrent tumors do not always show histological atypia. Even in the atypical and/or malignant examples of extrathoracic SFTs in which recurrences and metastases were documented, only one patient has died and then only of other causes and recurrence took many years to appear in some cases. In the case reported here, although the meningeal tumor was predominantly sclerotic and hypocellular, the hypercellular sarcomatous area met the criteria of malignancy among extracranial tumors described by other authors: hypercellularity, cellular atypia, and mitotic frequencies. The indolent nature of the present case is also consistent with the long interval for the recurrence and metastasis to take place described in some of the extrathoracic SFTs. Also, despite the fact that the patient received radiation therapy after the tumor was initially resected, we do not believe the histological changes were radiation induced because the original tumor also exhibited atypical features.
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There have been a few reports of SFTs that occur in the brain, mostly masquerading as a meningioma, as in this case.1–3,15–18 To our knowledge, 14 such tumors have been reported and all were benign except in one case in which the tumor recurred after subtotal resection. One case each showed invasion of brain and spinal nerve root but no metastasis was recorded.2 None of the reported cases of nervous system SFTs exhibited features of atypical or malignancy except one case described in the series published by Carneiro, et al.,2 which had five mitotic figures per 10 hpf. However, there was no recurrence or metastasis in this case after 10 years of follow up. Otherwise, our case has histological and other staining characteristics similar to previously reported intracranial cases.

In the brain, the main differential diagnosis is meningioma: which SFTs may resemble because of the focal presence of arborizing and staghorn vessels. Fortunately, SFTs are usually strongly positive for CD34, whereas for HPC, negative staining or at most weak and patchy staining is obtained.16,22 Also, HPC has a characteristic pericellular reticulin-staining pattern that is absent in SFT, as in this case. Moreover, the hypocellular areas of SFTs possess thick bands of keloidlike collagen, which are as a rule absent in HPC.4,5 Distinguishing SFTs from HPCs is important because HPCs are low-grade malignant tumors with a significant rate of recurrence and metastasis over the years.9 The other immunohistochemical properties, for example, positivity to vimentin, are similar.16 Distinguishing these tumors from fibrous meningiomas can be achieved by immunohistochemical staining for EMA because even very fibroblastic meningiomas are EMA positive.13 Also, under an electron microscope, meningiomas should show interdigitating processes with desmosomes.

Conclusions

We report what appears to be the first case of a meningeval SFT that has metastasized outside the nervous system. This case illustrates the importance of distinguishing SFT from a fibrous meningioma, because very rarely, the former behaves in an aggressive fashion similar to its soft-tissue or thoracic counterparts. This is particularly important in tumors in which hypercellularity, high mitotic counts, and cellular atypia are present. These patients may need close clinical and radiological follow up.

References


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