

June 2001: 61 year old woman with confusion and obtundation

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Clinical History

A 61 year-old female was found in her home moaning and unresponsive. She was taken to hospital where she was uncooperative and too obtunded to provide a history. According to her daughter, there was a 3-week history of progressive confusion, lethargy, headaches, and reduction in her spontaneous speech. On neurological examination she was disoriented and had a mild expressive aphasia. She was profoundly demented, unable to recognize her own daughter. Papilledema was noted as well as a right pronator drift.

Investigations included imaging of the head by CT and MRI (Figure 1), as well as cerebral angiography. A 5 cm. medial left frontal enhancing mass was identified. It distorted the frontal horn of the left lateral ventricle and inferiorly impinged on the left optic nerve. In addition, a large (2 cm) calcified partially thrombosed right carotid-ophthalmic artery aneurysm was noted. The patient underwent a left frontotemporal craniotomy and near-complete resection of the tumor, apart from a small portion adherent to the left anterior cerebral artery.

Gross

Intraoperatively the left frontal mass was bluish gray, firm and well demarcated from the adjacent brain.

Microscopic

Microscopically the left frontal mass showed a heterogeneous architecture with areas that were densely cellular alternating with areas that were hypocellular and myxoid (Figure 2a). The cellular regions were composed of highly pleomorphic, plump, spindle cells

arranged in fascicles, bands, and storiform patterns (Figure 2b). Elsewhere, pleomorphic and multinucleated cells were more widely dispersed on a myxoid background (Figure 3). The vascular bed was composed of variably sized capillary and sinusoidal vessels. Mitotic figures were easily found throughout the tumor with an average of 4 mitotic figures/10 HPF. Individual cells possessed a light reticulin investment. Immunohistochemical staining for actin, desmin, EMA, cytokeratin, S100, GFAP, NSE, synaptophysin, and KP1 was negative. The neoplastic cells were positive for CD34 (Figure 4), vimentin, and alpha-1-antichymotrypsin. Proliferative markers (Ki67) revealed variable staining density with up to 25 % of cells labeling focally (11% average in 10 high power fields). Electron microscopy failed to reveal features of specific lineage for the neoplastic cells.

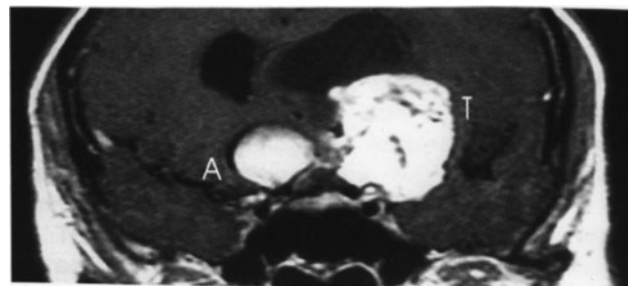


Figure 1.

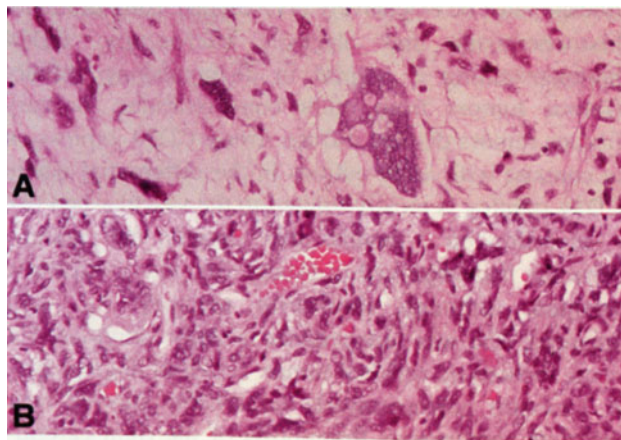


Figure 2.

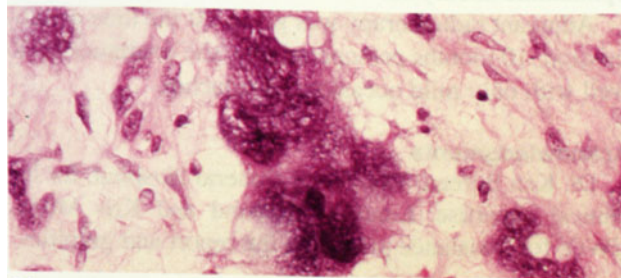


Figure 3.

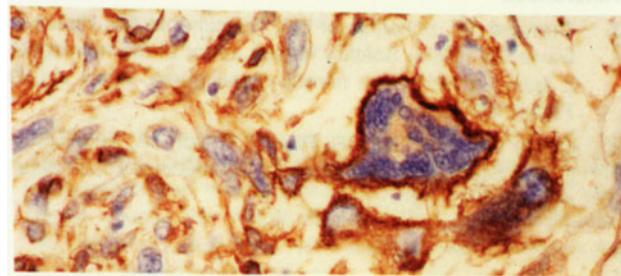


Figure 4.

Diagnosis

Malignant solitary fibrous tumor

Discussion

The solitary fibrous tumor was first described as a pleural lesion but has subsequently been reported in numerous extrathoracic sites. Meningeal examples have been reported but are rare (1,2,4,5). Extrathoracic malignant solitary fibrous tumors are extremely rare and have not been reported previously intracranially. Anaplastic features including areas of increased cellularity, necrosis, nuclear atypia, and greater than 4 mitoses/10 high power fields are felt to be associated with aggressive behavior (6) although the collective experience is limited.

The differential diagnosis includes hemangiopericytoma, anaplastic meningioma and malignant fibrous histiocytoma. As with any poorly differentiated tumor, cell lineage must be extensively investigated using immunohistochemistry and electron microscopy. CD34 immunostaining is helpful in distinguishing solitary fibrous tumors from other considerations. Solitary fibrous tumors display diffuse strong CD34 expression in contrast with other entities in the differential where CD34 staining is mostly limited to the vascular bed (3).

In follow-up 2 months after surgery, the patient's dementia had improved markedly. She was completely oriented, although short-term memory was still impaired. She was independent in daily activities. CT scans showed no residual tumor. Cerebral angiography at 3 months postoperatively remarkably showed near complete spontaneous thrombosis of the right carotid-ophthalmic aneurysm.

Adjuvant radiotherapy was given. At the time of submission, 10 months postoperatively, she continues to do well with only mild occasional headaches.

Acknowledgments

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References

1. Carneiro SS, Scheithauer BW, Nascimento AG, Hirose T, Davis DH (1996) Solitary fibrous tumor of the meninges; a lesion distinct from fibrous meningioma. *Amer J Clin Pathol* 106: 217-224
2. Challa VR, Kilpatrick SR, Ricci P, Wilson JA, Kelly DL Jr (1998) Solitary fibrous tumor of the meninges. *Clin Neuropath* 17: 73-78
3. Perry A, Scheithauer BW, Nascimento AG (1997) The immunophenotypic spectrum of meningeal Hemangiopericytoma: a comparison with fibrous meningioma and solitary fibrous tumor of meninges. *Amer J Surg Pathol* 21: 1354-1360
4. Prayson RA, McMahon JT, Barnett GH (1997) Solitary fibrous tumor of the meninges. *J Neurosurg* 86: 1049-1052
5. Slavik T, Bentley RC, Gray L, Fuchs HE, McLendon RE (1998) Solitary fibrous tumor of the meninges occurring after irradiation of a mixed germ cell tumor of the pineal gland. *Clin Neuropathol* 17: 55-60
6. Vallat-Decouvelaere AV, Dry SM, Fletcher CD (1998) Atypical and malignant solitary fibrous tumors in extrathoracic locations. *Amer J Surg Pathol* 122: 1502-1511

Abstract

Cases of the Month, April-June, 2001

The April Case of the Month (COM). The contributors report a case of a 70 year-old woman with recurrent meningiomas, one of which showed rhabdoid and lipomatous differentiation. Histopathological study of the first and second previous resections showed only typical meningothelial meningioma. On the third craniotomy, a new tumor specimen showed an admixture of classic meningothelial meningioma with lipomatous and rhabdoid foci. Immunohistochemical studies showed diffuse reactivity for epithelial membrane antigen and vimentin, as well as focal positivity for desmin and smooth muscle actin in the areas with rhabdoid features and S100 protein in the lipomatous foci. The presence of these three different and concomitant histological patterns only in the third surgical resection might support a metaplastic origin and, also, corroborates the concept that rhabdoid features are suggestive of an aggressive behavior.

The May COM. A 32-year-old woman with a history of previous mastoid surgery presented with bilateral extremity weakness and ambulatory instability. MRI revealed a dural-based mass completely encircling the upper cervical spinal cord. Workup was significant for an abnormally elevated c-ANCA, positive at a dilution of 1:128. A portion of the lesion was removed by a posterior surgical approach to decompress the cervical cord. Histologic examination of the dura showed a dense granulomatous infiltrate with vasculitis and giant cells; coupled with the positive c-ANCA, the process was felt to be most consistent with Wegener's granulomatosis. Wegener's granulomatosis infrequently involves the dura or meninges and has not previously been reported to affect dura of the cervical cord. Symptomatic improvement followed surgical decompression and high-dose corticosteroid therapy, with resultant resolution of an elevated c-ANCA titer.

The June COM. A 61 year old female presents with a three week history of increasing confusion, lethargy and headache. A neurological exam revealed disorientation, mild expressive aphasia, bilateral papilledema, and a right pronator drift. She had a craniotomy and resection of tumor. The tumor histologically was consistent with a solitary fibrous tumor displaying malignant features of hypercellularity, marked nuclear atypia, high mitotic activity, and a high proliferation index.

This case is unique as the first malignant variant of solitary fibrous tumor to be reported intracranially.

For a more complete discussion of these cases, additional micrographs and information regarding submission of cases, please access the WWW at: <http://www.brainpathology.com>. We welcome comments about these or similar cases our readers may have encountered.