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Rhabdoid meningioma occurring in an unrelated resection cavity with leptomeningeal carcinomatosis

Case report

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✓ Rhabdoid meningioma (RM) is a recently described, aggressive variant of meningioma. The authors report a case of RM occurring in the resection cavity of an unrelated neurosurgical procedure, temporal lobectomy for intractable seizures. The patient presented with intractable headache 10 years after the temporal lobectomy. Imaging revealed a dura-based, uniformly enhancing lesion within the resection cavity. She underwent gross-total resection and the findings of the surgical pathological report were consistent with an RM, with a dramatically elevated MIB-1 index of approximately 50%. The patient's clinical course was complicated by severe pain and communicating hydrocephalus secondary to rapid dissemination of malignant cells throughout the CSF pathways. Despite aggressive measures, including tumor resection, ventriculoperitoneal shunt placement, and the initiation of conventional radiation therapy, the ensuing leptomeningeal carcinomatosis proved to be rapidly fatal.

KEY WORDS • rhabdoid meningioma • leptomeningeal carcinomatosis • resection cavity

RHABDOID morphological characteristics have been identified in particulary malignant meningiomas in several recent reports.[5,8,9,11,15,21,22] Rhabdoid meningioma is a meningothelial tumor composed of sheets of loosely cohesive cells with eccentric nuclei, frequent mitotic figures, prominent nucleoli, and eosinophilic cytoplasmic inclusions. The inclusions are composed of whorls of intermediate filaments, most commonly vimentin.[5,8,15] Immunohistochemical staining typically is strongly positive for vimentin and endothelial membrane antigen, but negative for S100 protein, actin, HMB-45, glial fibrillary acidic protein, and AE1/3.[5,8,15] The term RM has been used to describe both tumors with clear histological transitions from typical meningothelial cells and those that clinically and radiologically resemble typical meningiomas but contain only the rhabdoid structure.[8,15]

Rhabdoid meningioma appears to have a particularly poor prognosis and is classified as a Grade III neoplasm in the 2002 World Health Organization classification.[10] In the largest case series[15] 13 of 15 patients had at least one recurrence after gross-total resection of the tumor, and two had documented extracranial metastases. The median time to death was 3.1 years after the first appearance of the rhabdoid structure. In one previous case report[23] the authors described a patient with CSF dissemination of malignant cells that, although not termed as such, bore morphological and immunohistochemical similarities to an RM. The CSF cytopathological findings in the present case have been reported in a separate publication focusing on CSF cytomorphology.[14]

Risk factors for rhabdoid transformation of meningiomas are currently unknown. Numerous case-control and observational studies[1,6,7,16–20] have led investigators to suggest a general association between head trauma and meningioma formation. This association appears particularly strong between 10 and 19 years after head trauma.[16] Whether meningiomas occur more frequently in patients who have undergone neurosurgical procedures has not been studied. We report the case of an RM that developed in a resection cavity and was associated with aggressive leptomeningeal dissemination.

Case Report

History. This 26-year-old woman with a history of intractable complex partial epilepsy had undergone a left inferior temporal lobectomy, amygdalectomy, and hippocampectomy in 1992 and placement of a vagal nerve stimulator in 2001. The initial procedure involved resection of 7 cm of the inferior temporal lobe with preservation of a small portion of the deep anterior and medial tissue. No epileptogenic lesion had been identified preoperatively. Tissue removed at surgery demonstrated both contemporaneously and retrospectively only normal brain, without evidence of meningioma, mesial temporal sclerosis, or cortical dysplasia. Postoperatively, the patient's seizures remained partially controlled by a regimen of two medications.

Examination. In July 2002, the patient presented to the

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computerized tomography; MR = magnetic resonance; RM = rhabdoid meningioma.

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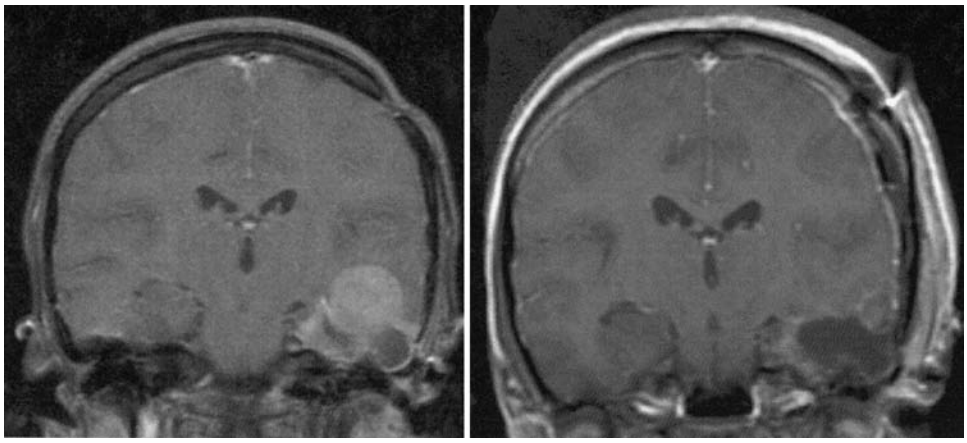

FIG. 1. Coronal Gd-enhanced MR images. *Left:* Preoperative image revealing a dura-based enhancing lesion originating from the resection cavity in the left temporal lobe. *Right:* Postoperative image demonstrating resection of the mass with some residual enhancing tissue in the resection cavity.

neurology clinic with complaints of severe headache, sacral pain, episodic lethargy, and vomiting of 3 weeks duration. She presented 5 weeks postpartum, following an uncomplicated pregnancy and delivery. The results of the neurological examination were within normal limits. Magnetic resonance imaging of the brain revealed a uniformly enhancing 3-cm-wide dura-based mass within the resection cavity in the left temporal lobe; moderate vasogenic edema of the underlying brain was also present (Fig. 1 *left*). No meningeal enhancement was noted and the ventricles were normal in size.

Operation. The patient was hospitalized and underwent gross-total resection of the tumor 6 days later (Fig. 1 *right*). The tumor was adherent to the overlying dura mater, requiring resection of a portion of the dura and closure aided by placement of Duraguard. All obvious tumor was resected, although it appeared to be highly infiltrative and postoperative MR imaging revealed enhancement of the surgical bed, which was suggestive of residual tumor. The pathological findings (Fig. 2) were consistent with an RM.

Postoperative Course. The patient's postoperative course was complicated by persistent CSF leakage and wound infection, requiring additional hospitalization 1 month later. The patient was treated with broad-spectrum antibiotics and underwent surgical wash-out and duraplasty.

Second Hospitalization. On Day 7 of the second hospital-

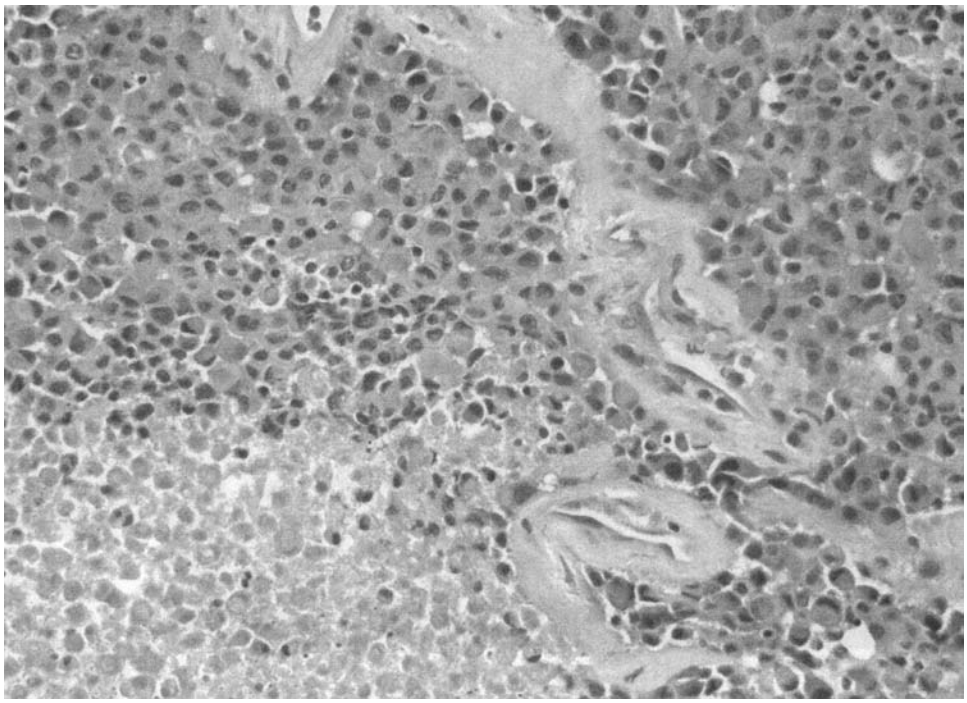

FIG. 2. Photograph of a brain specimen obtained at surgery showing the tumor with areas of necrosis and cells with the rhabdoid structure. H & E, original magnification × 200.

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ization, the patient experienced acute neurological deterioration and a dilated, unresponsive right pupil. Immediate medical reversal of cerebral herniation was undertaken. An emergency CT scan of the head demonstrated ventriculomegaly consistent with communicating hydrocephalus, prompting immediate placement of an intraventricular catheter. Although her level of consciousness improved, the patient manifested progressive weakness of the right leg and arm and experienced excruciating sacral pain, requiring very high doses of intravenous narcotics. On Day 13, contrast-enhanced MR imaging of the entire spine revealed enhancement of the conus medullaris (Fig. 3). At that time, CSF obtained by lumbar puncture was found to contain 1 mg/dl of glucose, 654 mg/dl of protein, no red blood cells/mm^3, and one white blood cell/mm^3. A CSF cytopathological analysis confirmed the presence of rhabdoid cells identical to the original tumor.

On Day 15, the patient underwent a second episode of decreased consciousness with dilated pupils. An emergency CT scan of her head demonstrated progressive ventriculomegaly, requiring revision of the intraventricular catheter for CSF drainage. It was evident at this stage that the woman would require long-term CSF drainage and we considered placement of a ventriculoperitoneal shunt. Despite the risk of intraperitoneal tumor seeding, the patient's dependence on external CSF drainage prompted us to perform ventriculoperitoneal shunt placement on Day 21.

Colleagues on the radiation oncology service recommended craniospinal external-beam radiation therapy beginning with the sacrum. On Day 24, following the first session of radiation therapy, the patient again displayed a rapidly declining mental status. A CT scan of the head was obtained and revealed diffuse cerebral edema without significant hydrocephalus, which resulted in cerebellar tonsillar herniation. Despite aggressive resuscitative efforts, including administration of mannitol and hypertonic saline, hyperventilation therapy, and aspiration of CSF through the ventriculostomy bulb, her neurological condition deteriorated further. Brain death was confirmed by clinical parameters and life-sustaining therapies were withdrawn. Permission for an autopsy was granted by the patient's family.

Pathological Findings. An extraaxial tumor was excised from the left temporal lobe via craniotomy on July 23, 2002. The tumor consisted of sheets of cells with eccentric nuclei and abundant eosinophilic cytoplasm and cytoplasmic inclusions consistent with a rhabdoid structure (Fig. 2). There was a moderate amount of pleomorphism and necrosis. The MIB-1 index was dramatically elevated to approximately 50%. The tumor cells stained focally but strongly for endothelial membrane antigen and vimentin, but did not stain for glial fibrillary acidic protein, AE1/3, HMB-45, or S100 protein. Estrogen and progesterone receptors were absent. No regions of conventional meningioma were present.

The CSF cytopathological analysis revealed malignant cells with markedly enlarged, eccentrically placed nuclei, which were morphologically similar to those seen in the specimen obtained during the original excision. Further details on the findings are provided in a separate publication.[14]

The autopsy showed extensive residual tumor in the left temporal lobe resection cavity and a diffuse leptomeningeal spread of tumor along the spinal cord and nerve roots (Fig. 4). Microscopically, significant perineural and vascular invasion was identified within the meninges with direct extension of tumor cells into the brain parenchyma. Diffuse cerebral edema was noted with cerebellar tonsillar herniation. The systemic examination was unremarkable, and no tumor foci were found outside the CNS and the meninges.

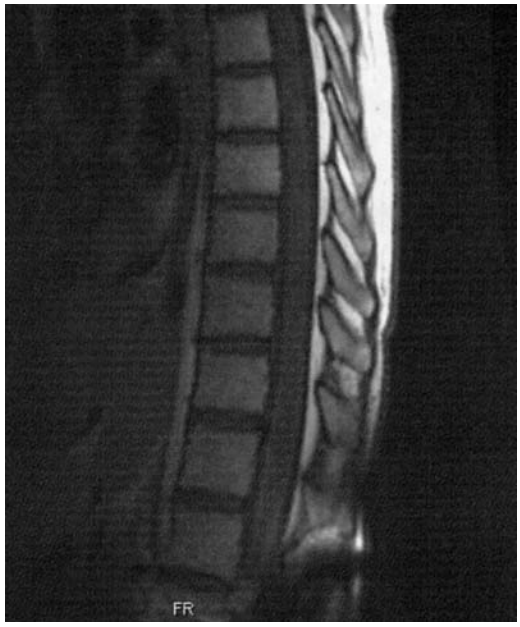

FIG. 3. Midsagittal Gd-enhanced MR image of the lumbosacral spinal cord demonstrating linear enhancement of the meninges surrounding the conus medullaris.

Discussion

We report the first case of an RM that occurred in the resection cavity after a neurosurgical procedure for an unrelated, benign disease—temporal lobectomy for amelioration of complex partial seizures. One prior case report contains details on the occurrence of an RM in the resection cavity of an incompletely excised ganglioglioma.[2] The appearance of an RM at the resection site may be related to the observation that remote head trauma is associated with later development of meningioma.[1,6,7,16–20] In a study of 200 patients with meningiomas and 400 healthy volunteers, Phillips, et al.,[16] reported the strongest association (odds ratio 4.33; 95% confidence interval 2.06–9.10) with head trauma occurring between 10 and 19 years before the diagnosis of the meningioma. In the case reported here, the tumor was diagnosed 10 years after the patient underwent temporal lobectomy. The presumed mechanism involves posttraumatic inflammation, healing, and release of growth factors leading to neoplastic transformation. Although similar processes are likely to occur postoperatively, the potential association between remote neurosurgery and subsequent meningioma formation has not been explored. With the increasing popularity of epilepsy surgery in young patients, it may be possible to study this question in humans.

Malignant transformation after surgical trauma has been studied in several extraneural organ systems including the skin, bone, and liver. Most researchers have invoked the

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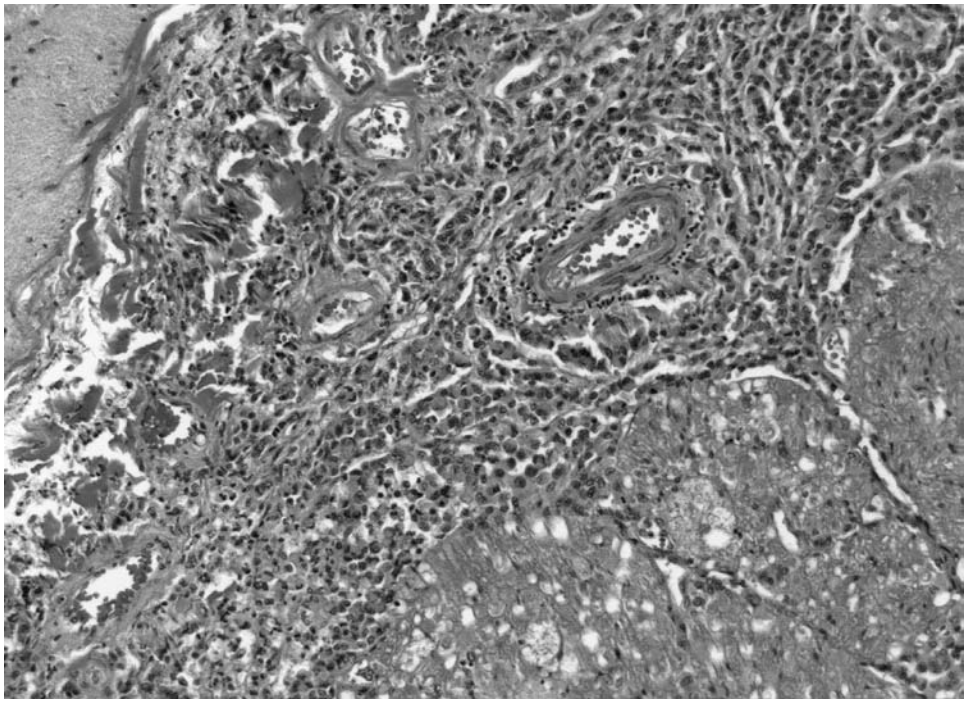

FIG. 4. Photomicrograph of a specimen of the posterior thoracic spinal cord obtained at autopsy. Malignant cells encase the spinal cord and the posterior spinal nerve roots. H & E, original magnification × 100.

"two insult" model of malignant transformation to speculate that tumor growth at operative sites requires a genetic or environmental predilection for malignancy that is exacerbated by growth factors at the site of surgical trauma. This theory was tested in an experimental rat model in which neonatal rats exposed to intravenously administered high-dose nitrogen-based carcinogens underwent surgical trauma to the brain. Compared with a control group, which was also exposed to carcinogens, twice as many neoplasms, including gliomas and vestibular schwannomas, developed at the site of injury in the surgical trauma group.[13] This study did not address meningiomas. The need for an underlying predilection for malignancy might explain the rarity of postoperative tumor growth.

The management of malignant meningioma has included maximal surgical debulking followed by external-beam radiation therapy with or without systemic chemotherapy. Conventional radiation therapy and gamma knife surgery have been used in other cases of RM with variable results.[5,8,15,23] Conventional radiotherapy appears to be efficacious in controlling recurrent typical and malignant meningiomas.[3,4,12] Whether this therapeutic modality has efficacy in the rhabdoid variant requires further study. This patient died after a single session of sacral radiation therapy.

Rhabdoid meningioma is an aggressive variant that carries a very poor prognosis. In this case, it occurred in the resection cavity of an unrelated neurosurgical procedure and metastasized throughout the CSF pathways. Despite gross-total resection, placement of a ventriculoperitoneal shunt, and the initiation of external beam radiation, the tumor was rapidly fatal. The patient died 38 days after tumor resection and 11 days after identification of malignant cells within the CSF.

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