Extra-Axial Cystic Meningioma without Dural Attachment in an Adult: Case Report and Review of Literature

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Introduction

Meningiomas are commonly derived from dural-based arachnoid cells and hence are usually attached to the dura.1 Rarely, meningiomas may present without any dural attachment.2,3 Meningiomas without dural attachment most commonly occur in the ventricles and rarely from the parenchyma.4,5 Intraparenchymal meningiomas are extremely rare in “meningiomas without dural attachment” as classified by Cushing and Eisenhardt.6 They are usually solid tumors.7 Cystic meningiomas are rare tumors accounting for 1.7 to 7% of meningiomas.8,9 Children have higher incidences of cystic meningiomas, intraparenchymal meningiomas, and meningiomas without dural attachment.10 Cystic meningioma without dural attachment in an adult is very rare. We report the case of this very rare entity and review the literature.

Materials and Methods

A 47-year-old female presented with an altered sensorium. She had a history of progressive bifrontal headache for 2 months. On general examination, her vital signs were within normal limits. On neurological examination, she was confused, not following verbal commands, and was partially opening her eyes on painful stimuli. Her pupils were 3 mm in size bilaterally and normally reacted to light. Planters were extensor on both sides. Fundoscopy revealed papilledema. Computed tomography (CT) scan of the brain showed an 8 cm × 6 cm cystic lesion with a solid component in the left frontoparietal region with a midline shift. The solid part of the lesion was enhancing on contrast but the cyst rim was not. Intraoperatively, the cyst was filled with amber-colored fluid, which was drained, and the solid component was completely excised. Histopathological examination of the solid tumor component confirmed cystic meningioma. At 2 years of follow-up, she has no evidence of recurrence. We report the fifth case of this very rare entity and review the literature.
A region with a solid component (►Fig. 1A). The solid part of the lesion was enhancing on contrast, but the cystic was not enhancing (►Fig. 1B). The ipsilateral frontal horn was significantly compressed with a midline shift of 1.5 cm. Intraoperative photomicrograph showing an extra-axial solid component lying over the brain parenchyma following cyst decompression and was weakly adherent to the arachnoid trabeculae without dural attachment (C). It was brownish-yellow, soft, fleshy, minimally vascular solid mass (D).

Histopathological examination of the solid tumor component showed nests of monotonous cells separated by loose fibrovascular tissue. The tumor cells showed dispersed chromatin and indistinct cytoplasm. Occasional nuclear holes and nucleomegaly were noted along with whorling. The fibrovascular tissue showed capillary proliferation and cystic changes (►Fig. 2). Focal calcification was also noted. Nuclear atypia and mitosis were not seen. Immunohistochemistry revealed diffusing positive for vimentin, and MIB-1 was less than 1%.

Postoperatively, the patient became conscious and made a slow and progressive recovery in the first 72 hours. On the
fourth postoperative day, she again became unconscious. An urgent CT scan brain was performed that revealed bilateral extradural hematomas (►Fig. 3A). Bifrontal craniotomy was performed and extradural hematomas removed. Postoperative CT scan after evacuation of clot revealed no evidence of lesion with reversal of midline shift (►Fig. 3B). She made a significant clinical recovery without any neurological deficit. At 2 years of follow-up, she has no clinical signs of recurrence.

Discussion

Meningiomas without dural attachment can be classified into five types: intraventricular meningioma, pineal region meningioma, deep sylvian fissure meningioma, intraparenchymal or subcortical meningioma, and others.11 They probably originate from ectopic meningothelial cells, which are not directly adjacent to the dura mater.12 They are more common in males. Meningiomas without dural attachment most commonly occur in the ventricles. However, they may also occur in the sylvian fissure, pineal region, and even in the brain parenchyma.11,13–15 Intraparenchymal meningiomas are rare intracranial tumors and only 27 cases of intraparenchymal meningiomas have been reported.4,5 They arise from the arachnoid cells of the pia mater, which are supplied by the perforating blood vessels on entry to the brain sulci.16 Intraparenchymal meningiomas are most commonly present with seizures. The most common location is the frontal lobe. Fibroblastic meningioma is the most common histologic subtype of intraparenchymal meningiomas. Most of the intraparenchymal meningiomas are World Health Organization grade I meningioma.9,17 Intraparenchymal meningioma with no connection to the dura is very rare in adults.4,7 Cystic meningiomas are rare tumors accounting for 1.7 to 7% of meningiomas.8 The cysts in meningioma may be intra- or extratumoral. Cystic meningiomas are commonly located at the cerebral convexity and usually are of fibrous histology.7 Cysts most widely develop in fibroblastic and meningothelial meningiomas.5 Cystic meningiomas are classified into four types—(1) centrally located intratumoral cyst, (2) peripherally located intratumoral cyst, (3) peritumoral cyst with walls consisting of both adjacent parenchyma and the tumor, and (4) peritumoral cyst with walls formed by the arachnoid, while the cyst remains separated from the tumor by a distinct capsule.18 Various theories have been proposed for cyst formation in a meningioma. Intratumoral cysts may be the outcome of microcystic degeneration, ischemic necrosis, intratumoral hemorrhage, or intratumoral metastasis, and peritumoral cysts originate from peritumoral edema, widening of the subarachnoid space, demyelination, or peritumoral hemorrhage.9,19

The frequency of atypical meningioma among cystic meningioma is 12%, higher than that among all meningiomas, and suggests that atypical meningioma tends to form a cyst compared with other subtypes.20 Review of the histological classification of 69 cystic meningiomas showed meningotheliomatous (42.0%) and fibroblastic (27.5%) lesions were the most common types. Lesions were malignant in 7.2% of cases.21 Kolluri et al suggested that sarcomatous and angioblastic meningiomas are more frequently associated with cyst formation.22

Meningioma may be extremely difficult to distinguish at the preoperative diagnosis from glioma, metastatic brain.
tumor, lymphoma, subdural sarcomatous mass, or other intra-axial lesions, especially if located far from the para- or intraventricular regions or the sylvian fissure. Cystic meningioma is difficult to distinguish from a glioma with cystic changes or a metastatic neoplasm. One should consider intraparenchymal meningioma in the differential diagnosis of an intra-axial lesion in a child. The presence of cyst formation and heterogeneous enhancement mimic other neoplasms, such as gliomas, hemangioblastomas, and metastatic tumors.

CT and magnetic resonance imaging (MRI) are characteristics of typical meningioma and include a well-circumscribed, homogeneously enhancing extra-axial mass. The presence of an associated cyst and peritumoral edema may make it difficult to distinguish the tumor from a primary intra-axial glial neoplasm. In intraparenchymal meningioma, CT and MRI demonstrate no typical features, so preoperative diagnosis is difficult. Enhancement of the cyst wall strongly suggests the presence of tumor cells, but the absence of enhancement of the cyst wall does not exclude the presence of tumor cells in the cyst wall. Enhancement on CT or MRI indicates the presence of tumor cells in the cyst wall, and the enhancement depends on the presence and number of tumor cells.

For meningiomas, the primary treatment is surgery. Complete tumor resection is the most important factor in the prevention of tumor recurrence. Resection of the entire cyst wall is optimum, even if neuroimaging does not show enhancement of the cyst wall. However, the cyst wall often adheres tightly to the surrounding brain tissue and may be difficult to remove without damaging the adjacent brain tissue. After total resection of tumor and cyst wall, the cure rate is not significantly different between cystic and solid meningioma. Radiation therapy is beneficial in the treatment of residual meningiomas. Radiotherapy for atypical or malignant meningioma is recommended immediately after the initial surgery. The recurrence of a meningioma depends on the extent of removal, location of the tumor, and histological subtype. The extent of resection seems to be the most critical factor influencing the prognosis of a meningioma.

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References