A 37-Year-Old Woman With Dural-Based Intracranial Masses

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A 37-year-old woman presented with visual changes, headaches, numbness of the right side of the face, and difficulty swallowing due to numbness of the throat. The patient was 8 months pregnant and had a history of gestational diabetes. Following an uneventful delivery, imaging studies of the brain (computed tomographic scan and magnetic resonance imaging) demonstrated 2 masses. The first was a focally enhancing extra-axial mass, measuring 3.3 × 3.2 × 1.5 cm, which was found overlying the convexities of the right posterior frontoparietal regions. The second was a focal extra-axial mass located posteriorly in the midline, measuring 4.6 × 4.1 × 2.5 cm; this mass compressed both occipital lobes. These findings were interpreted as being compatible with meningiomas. An abnormal signal also appeared in the calvarium adjacent to the second mass, consistent with hyperostosis of the adjacent bone. The patient's preoperative neurological motor examination demonstrated bilateral papilledema and slightly decreased sensation to touch on the right side of her face.

The patient underwent an image-guided occipital craniotomy for resection of the occipital region mass with duraplasty and cranioplasty. The tissue received in pathology consisted of multiple irregular fragments of tan-yellow soft tissue, some of which showed attachment to the dura, and a segment of occipital bone with fragments of mass firmly attached to its convexity. The thickness of the calvarium measured 2.0 cm maximally. Histologically, the tumor showed marked microcystic changes throughout and was composed of fibrillary stellate cells, which gave the tissue a delicate, lacelike pattern (Figures 1 and 2). The cytoplasmic processes of the tumor cells surrounded round to oval microcysts. Other areas demonstrated a myxoid appearance, owing to aggregates of small extracellular cystic spaces filled with lightly staining fluid material. The nuclei were small, round to oval, and had stippled chromatin. Rare intranuclear pseudoinclusions and psammoma bodies were present. Small, focal areas of more recognizable meningothelial nests were also present. Mitoses and necrosis were not identified. A Ki-67 immunostain was performed, and a labeling index of less than 1% was noted.

What is your diagnosis?

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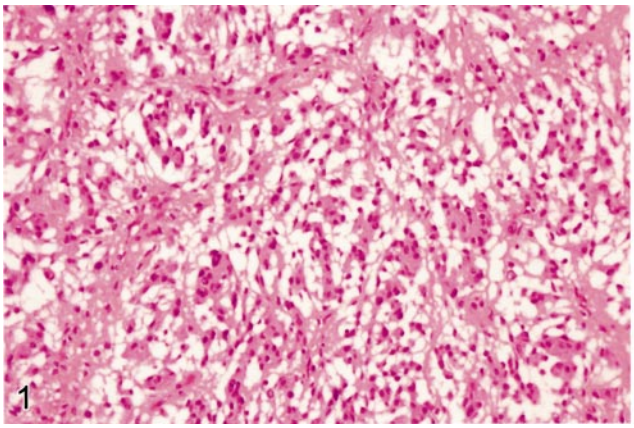
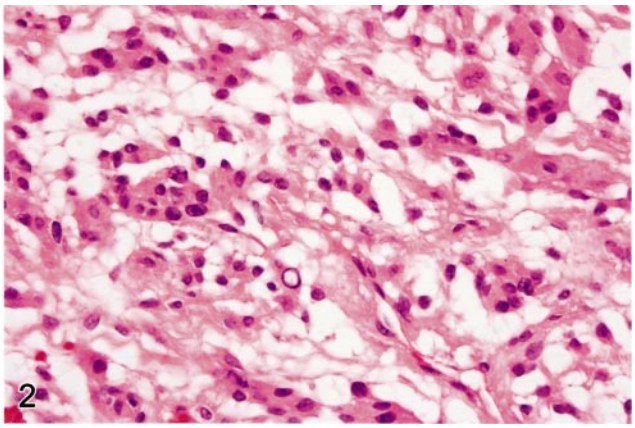




Pathologic Diagnosis: Microcystic Meningioma

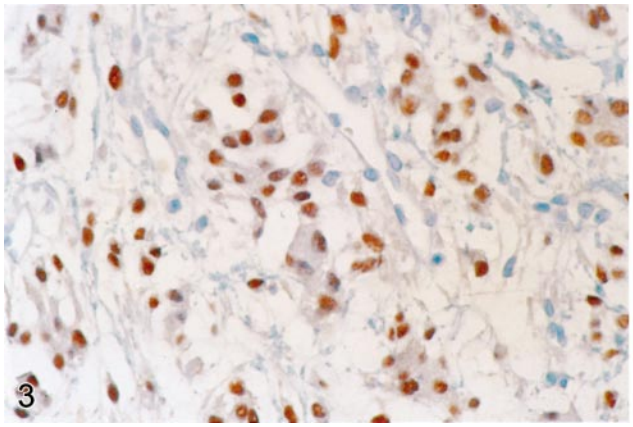
The large number of recognized histologic variants of meningioma underscores the phenotypic diversity of this tumor group. Focal microcystic changes have occasionally been identified in typical meningiomas.[1] However, tumors with extensive microcyst formation are rare. Such tumors were originally described and reported under a variety of designations, including humid, humid and myxomatous, vacuolated, and microcystic.[1–4] They have subsequently been classified as a distinct subgroup of meningiomas in the World Health Organization (WHO) classification of central nervous system tumors.[5] Despite their morphologic differences, microcystic meningiomas are considered to have a clinical course similar to typical meningiomas and are considered WHO grade I neoplasms.

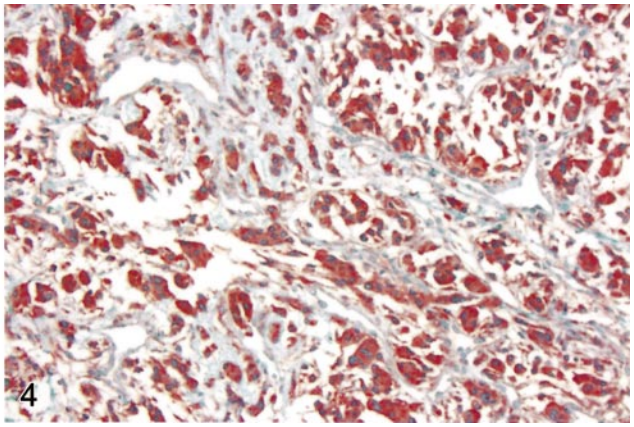
Histologically, microcystic meningioma is characterized by cells with elongated processes and by a loose myxoid background, giving the appearance of many microcysts. Pleomorphic cells may be focally evident. The tumor will usually show foci of typical meningothelial whorls. Electron microscopy has demonstrated the extracellular location of the microcysts.[6] Immunostains for estrogen and progesterone receptors were also performed in this case; the tumor showed diffuse positive staining for progesterone receptor (Figure 3), but did not stain for estrogen receptor. Immunostaining for epithelial membrane antigen (EMA) was strongly positive (Figure 4).

Meningiomas are generally considered to be of mesenchymal origin, arising from the arachnoidal cap cells; electron microscopy has shown the tumors to have interdigitating cell processes and desmosomal intercellular junctions. Microcystic meningiomas and classic meningiomas share the same immunohistochemical staining patterns. Their dual mesenchymal and epithelial properties are reflected by their staining for both vimentin and EMA. Microcystic meningiomas described in the literature have also shown positive staining for vimentin and EMA, with negative staining for cytokeratin and glial fibrillary acidic protein.[2,7,8] Classic meningiomas may also express estrogen and progesterone receptors; however, it is still uncertain what role the expression of these receptors plays in the formation and growth of these tumors and whether there is any utility in using hormone antagonists in treating tumors. Progesterone receptor expression is more common (approximately two thirds of meningiomas) in female patients.[5] There have also been reports of meningiomas manifesting and exhibiting rapid growth during pregnancy, as in our case.[9] Cell proliferation marker labeling indices in grade I tumors usually are low, as in this case; high rates of Ki-67 labeling in meningiomas are associated with an increased likelihood of local recurrence.[10]

Asymptomatic meningiomas can be managed by observation, since tumor growth is often indolent. Symptomatic meningiomas and those that are expanding, infiltrative, or that have considerable surrounding edema should be surgically resected. Occasionally, meningiomas will invade through the dura into the skull, where they may induce a hyperostosis, indicative of skull invasion. Overt attachment to or invasion of the adjacent brain is rare. Progression-free survival is higher in patients with gross total resection than in those with subtotal resection. External beam radiation therapy can also be used in patients in whom gross total resection is not possible or in managing higher grade tumors.

The importance of recognizing this entity lies in differentiating this tumor from other central nervous system tumors with a myxomatous appearance. Chordoid meningioma is characterized by trabeculae of eosinophilic, epithelioid cells in a myxoid background. A lymphoplasmacytic infiltrate may be prominent, and a few patients have associated hematologic conditions, such as Castleman disease.[5] Microcystic meningiomas can show considerable overlap with chordoid meningiomas, owing to myxoid stromal changes and cytoplasmic vacuolation. However, microcystic meningioma will not demonstrate cords or trabeculae of eosinophilic cells and will lack an inflammatory cell infiltrate. This distinction is important, since chordoid meningiomas are WHO grade II neoplasms and exhibit a high rate of recurrence following subtotal resection.[5] Myxomatous schwannomas will be positive for S100, negative for EMA, and would be unusual in this location. Microcystic gliomas stain with glial fibrillary acidic protein and are negative for EMA; they also would be unusual in an extra-axial location. Hemangioblastomas are characterized by large, lipid-containing, vacuolated stromal cells, which at times can demonstrate considerable nuclear pleomorphism and can mimic the microcysts of microcystic meningioma. However, hemangioblastomas are negative for EMA. In cases with prominent nuclear pleomorphism, metastatic carcinoma may be a consideration. The presence of meningothelial whorls and absence of mitotic figures can help make the distinction.





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