Multifocal Meningioangiomatosis: A Report of Two Cases

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Summary: We report the CT and MR findings in two patients with multifocal meningioangiomatosis, neither of whom had a family history or stigmata of neurofibromatosis. All lesions were located in the cortical and subcortical areas and had round dense calcifications with eccentric cysts. The masses were associated with surrounding edema and gliosis.

Meningioangiomatosis is a rare, benign hamartomatous lesion found in the cerebral cortex and leptomeninges (1). Grossly and microscopically, it is characterized by cortical meningovascular proliferation and leptomeningeal calcification (2). Its pathogenesis has not been established; however, it is known to be associated with neurofibromatosis (NF) in nearly 50% of reported cases (3). In most reported cases, the mass lesion is solitary, although multiple lesions have occasionally been described. To our knowledge, though, multiple lesions with accompanying eccentric cysts have not been described on CT or MR studies. Accurate diagnosis of meningioangiomatosis is important because it is a benign, surgically correctable cause of seizures.

We report two cases of multifocal meningioangiomatosis.

Case Reports

Case 1

A 47-year-old woman had a 5-year history of headaches and generalized seizures. Precontrast CT scans showed multiple round calcified masses and eccentric cysts with edema in the left frontoparietal and right parietal lobes (Fig 1A and B). On T1-weighted MR images, the cyst was nearly isointense with CSF (Fig 1C). Proton density— and T2-weighted images revealed areas of dark signal intensity surrounded by areas of high signal intensity (Fig 1D and E). The central areas of dark signal intensity corresponded to the calcifications seen on CT scans. No significant mass effect was demonstrated. The patient underwent a left parietal craniotomy, and a calcified mass with cyst was totally removed; however, a small calcified mass in the right parietal lobe was not removed. There was no invasion of the overlying dura. The calcified mass was present in the cerebral cortex, and a cystic cavity was found in the subarachnoid space. Pathologic examination of the resected specimen showed multiple fragments of firm, whitish, calcified tissue, measuring 2.0 × 1.0 × 1.0 cm and 0.2 × 0.1 × 0.1 cm, respectively. Microscopically, the lesions consisted of extensive fibrillary calcification and proliferating vessels surrounded by meningothelial cells (Fig 1F). The patient has been free of seizures and headaches for the 15 months since surgery.

Case 2

A 53-year-old man had a 2-year history of generalized seizures and headaches. Precontrast CT scans revealed dense round calcifications with eccentric cysts surrounded by edema in the left frontal and parietal lobes (Fig 2A). MR images of the brain showed heterogeneous areas of hypointense and isointense signal in the left frontal and parietal lobes on T1-weighted sequences (Fig 2B). The lesions showed irregular enhancement on contrast-enhanced T1-weighted images (Fig 2C and D). T2-weighted images revealed marked hypointensity corresponding to calcification and also revealed high signal intensity corresponding to cyst and edema (Fig 2E). The patient underwent left frontal and parietal craniotomies, and the calcified masses with cysts were totally removed. There was no involvement of the overlying dura. The calcified mass was located in the cerebral cortex and the cyst was in the subarachnoid space. The tumors were well demarcated, firm, whitish, and calcified. Microscopically, the lesions consisted of extensive fibrillary calcification and proliferating vessels surrounded by meningothelial cells. The patient has remained free of seizures and headaches during the 7 months since surgery.

Discussion

Meningioangiomatosis is a rare benign disorder characterized by the hallmarks of meningioma and angiomia (4). It is a very slow-growing tumor (5) whose features have previously been reported (1–25). Meningioangiomatosis was first described by Bassoe and Nuzum in 1915 (6) as an incidental autopsy finding in a 15-year-old boy; it was named by Worster-Drought et al in 1937 (1). Meningioangiomatosis primarily affects children and young adults, and seizures and/or headaches are the most common symptoms (2). It is associated with NF in nearly 50% of reported cases (3). A review of the literature suggests that meningioangiomatosis is strongly associated with NF2 rather than NF1 (7, 8).

Though its pathogenesis is unknown, three possible theories have been suggested (9): first, these lesions could represent a hamartoma; second, they could result from direct invasion of the brain tissue by a leptomeningeal meningioma; and third, they
could represent a vascular malformation. Characteristics of meningioangiomatosis are leptomeningeal calcification and meningovascular proliferation interwoven with bands of fibrous connective tissue (10). The pathologic criteria for meningioangiomatosis may be summarized as follows: leptomeningeal proliferations of nodules, whorls, or bands of meningothelial cells (ie, arachnoidal cap cells exhibiting marked degenerative reactions, such as calcification, fibrocartilage, or bone formation) in association with sharply demarcated intracortical plaques of proliferating small vessels and perivascular cuffs of spindle-shaped fibroblast-like cells (4). The calcification patterns vary from faint psammomatous calcification to dense osteoid (5). Meningioangiomatosis affects the cerebral cortex in 90% of cases and usually occurs in the frontal or temporal lobes, but is rarely found in the third ventricle, thalamus, cerebral peduncles (2), or brain stem (11). An association with meningioma or oligodendroglioma has occasionally been reported (12, 13).

Although multifocal cerebral lesions have been reported, meningioangiomatosis usually consists of well-demarcated solitary lesions (2, 14). Multifocal lesions or masses associated with cysts have not been described on either CT or MR studies. Our two patients with multifocal meningioangiomatosis were older than those in most reported cases and had no stigmata or family history of NF. On CT and MR examinations, the masses were composed mainly of dense round calcification and the cysts were eccentric to the mass. At surgery, the lesions were well demarcated and located superficially. Two lesions showed multiple calcifications with eccentric tumoral cysts. Cysts were also present in the subarachnoid spaces. It could be suggested that the mechanism of extratumoral cyst formation is
the same as that of cystic meningioma, in which the cyst may form as the result of a ball-valve mechanism with the gradual accumulation of CSF in several sulci between the tumor and brain parenchyma (15).

Meningioangiomatosis is a benign lesion that does not become malignant (5). Its accurate diagnosis is important because total surgical removal is the treatment of choice, and the prognosis after surgery is excellent in most cases. The differential diagnosis of radiologic images (10, 25) includes meningioma, oligodendroglioma, granulomatous meningitis, and parasitic diseases.

**Conclusion**

Meningioangiomatosis is a benign cerebral lesion characterized by leptomeningeal calcification and meningovascular proliferation. It exhibits peripheral edema or gliosis on CT and MR imaging studies. It usually manifests as a solitary lesion; however, as we report, multifocal lesions or masses associated with cyst formation may be present.

**References**

1. Worster-Drought C, Dickson WEC, McMenemy WH. Multiple meningeal and perineural tumors with analogous changes in the glia and ependyma. Brain 1937;60:85–117