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). A review of the literature suggests that meningioangiomatosis is strongly 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).

Discussion

Meningioangiomatosis is a rare benign disorder characterized by the hallmarks of meningioma and angiomatosis (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
Fig 2. Case 2: 53-year-old man.

A, Noncontrast CT scan reveals multiple round calcifications (closed arrows) with eccentric cysts (open arrow) and moderate edema.
B, On T1-weighted image (420/14/2), the lesions show inhomogeneous hypo- and intermediate signal intensity in the left frontal and parietal lobes.
C, On T2-weighted image (2600/90/2), areas of heterogeneous signal intensity were noted in the left frontal and parietal lobes, caused by calcification (closed arrows), cyst (open arrow), and edema.
D and E, On contrast-enhanced T1-weighted image (420/14/2), the lesions show irregular enhancement (arrows).

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