CT and MR Findings of Meningioangiomatosis

Man Soo Park, M.D., Dae Chul Suh, M.D., Woo Suk Choi, M.D., Sang Youl Lee, M.D., Haingsub R. Chung, M.D., Sang Jin Bae, M.D., Nam Hyeon Kim, M.D., Seung Mun Jung, M.D., Dae Sik Ryu, M.D.

Purpose: To characterize the CT and MR findings in patients with meningioangiomatosis (MA).

Materials and Methods: Four patients (18 to 53 years old, two females and two males) with MA were retrospectively reviewed. CT was used in four cases and MR in three. Pathologic specimens were obtained from all four.

Results: All lesions were located in the cortical and subcortical areas and showed spotty (n = 1), popcornlike (n = 2), or gyral (n = 1) calcification. The masses were associated with surrounding edema and gliosis. In two patients, lesions were multiple and were accompanied by eccentric cysts.

Conclusion: MA is a surgically correctable benign disease. Its radiologic characteristics are cortical or subcortical mass with various calcifications, associated peripheral edema and gliosis.

Index words: Brain, CT, Brain, MR, Meninges, neoplasms

May is a rare benign hamartomatous lesion of the cerebral cortex and leptomeninges. Grossly and microscopically it is characterized by cortical meningovascular proliferation and leptomeningeal calcification, though its pathogenesis is unknown. In almost 50% of reported cases it was associated with neurofibromatosis (NF). The reported CT findings of MA are various types of calcification with little or no contrast enhancement, or a hypodense round mass in the cortical or leptomeningeal area. In several cases, MR findings have been reported; abnormalities are confined to the cortex and consists of isointensity or hypointensity on T1 weighted images and T2 weighted images show a heterogeneous cortical mass surround by an area of increased intensity due to edema or gliosis.

Materials & Methods

We retrospectively reviewed six lesions in four patients with MA. Two were men and two were women, and their ages ranged from 18 to 53 years. Two had a history of seizures, and two had suffered seizures and headaches, but none had a family history or stigmata of NF.

Using a Simens Somatom Plus 4B scanner (Erlangen, Germany), all patients underwent unenhanced and contrast CT; three underwent MR on a Simens 1.0T Magnetom Expert, and on a 0.5T scanner at an outside institution. In three cases, T1(420/14/2, repetition time/echo time/excitations), proton density(2600/22/2) and T2(2600/90/2) weighted spin-echo images were obtained, and in two, gadolinium enhanced T1 weighted images were acquired. All lesions were evaluated for pattern of calcifications, associated cysts with mass, edema, location and extent of enhancement. One patient underwent angiography at an outside institution and all underwent surgery for tumor removal.
Results

The clinical data and radiologic findings of six lesions in four patients are summarized in Table 1.

On CT scans, all lesions showed calcifications which were popcornlike (n = 2), spotty (n = 1), or gyral (n = 1), and in all, there was edema. In two of the four, eccentric cysts were associated with the calcified mass (Figs 1 & 2). Lesions were located in the cortical and subcortical areas of the frontal (n = 1), frontoparietal (n = 1), parietal (n = 2), temporal (n = 1) and temporoparietal (n = 1) lobes. In two of the three patients who underwent MRI, T2-weighted images showed that the lesions were predominantly high signal intensity with central areas of low signal intensity (Fig. 1D, Fig. 2C); the latter corresponded to the calcifications seen on CT. On T2-weighted images, eccentric cysts, which were of isosignal intensity, with cerebrospinal fluid (CSF), were not clearly identified. On T1-weighted images, the lesions showed heterogeneous signal intensity and cysts were slightly hyperintense relative to cerebrospinal fluid (Fig. 2B). In the third patient, the lesion was intermediate on T1-weighted images, and on T2-weighted image.

![Images of CT scans and histopathologic specimen showing popcornlike calcifications and eccentric cysts with moderate edema, high signal intensity with central low signal intensity, proliferaated blood vessels surrounded by meningothelial cells and fibrillary calcification.](image-url)
calcification (+) -- multiple, popcornlike high SI with central low SI
cyst (+) -- eccentric edema (+), CE (-)
calcification (+) -- multiple, popcornlike high SI with central low SI
cyst (+) -- eccentric edema (+), CE (-)
calcification (+) -- amorphous
calcification (+) -- spotty high SI (T2WI)
cyst (-), edema (+)
CE (+) -- nodular
syst (-), edema (+), CE -- not performed

**Table 1. Clinical Features & Image Findings of Meningiogliomatosis Cases.**

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age / sex</th>
<th>Clinical Presentation</th>
<th>Location</th>
<th>CT findings</th>
<th>MR findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>47/F</td>
<td>Seizure headache</td>
<td>Lt. fronto-parietal</td>
<td>calcification (+) -- multiple, popcornlike cyst (+) -- eccentric edema (+), CE (-)</td>
<td>high + SI with central low SI (* T2WI) CE (-)</td>
</tr>
<tr>
<td>2</td>
<td>53/M</td>
<td>Seizure headache</td>
<td>Lt frontal Lt parietal</td>
<td>calcification (+) -- multiple, popcornlike cyst (+) -- eccentric edema (+), CE (-)</td>
<td>high SI with central low SI T2WI CE (+) -- amorphous</td>
</tr>
<tr>
<td>3</td>
<td>21/F</td>
<td>Seizure</td>
<td>Lt. temporal</td>
<td>calcification (+) -- spotty cyst (-), edema (+) CE (+) -- nodular</td>
<td>high SI (T2WI) CE (+) -- nodular &amp; leptomeningeal</td>
</tr>
<tr>
<td>4</td>
<td>18/M</td>
<td>Seizure</td>
<td>Rt. temporo-parietal</td>
<td>calcification (+) -- cortical gyral syst (-), edema (+), CE -- not performed</td>
<td>Not performed</td>
</tr>
</tbody>
</table>

* T2WI -- T2 weighted image, * CE -- contrast enhancement, * SI -- signal intensity, * Lt = left, Rt = right

In post-operative follow-up 15 (case 1), seven (case 2), 24 (case 3), six (case 4) months, seizures were free in all patients.

**Fig. 2.** patient 2

A. Precontrast CT scan reveals multiple popcornlike calcifications with eccentric cyst (open arrowheads) and moderate edema.

B. On sagittal T1 weighted image (420/14/2), lesions show inhomogeneous hypo-(arrow) and intermediate signal (open arrowhead) intensities in left frontal & left parietal lobes.

C. On axial T2 weighted image (2600/90/2), heterogenous signal intensities were noted in left frontal & parietal lobes due to calcifications (arrows), cyst (open arrowheads) and edema.

D. On coronal Gd-enhanced T1 weighted image (420/14/2), lesion (posterior mass) shows amorphous enhancement (arrows).
homogeneous high signal intensity. On Gd-DTPA-enhanced T1-weighted images, enhancement was observed in two of three cases. One lesion showed amorphous enhancement (Fig. 2D), and in the other, enhancement was cortical and leptomeningeal (Fig. 3D). In one patient, angiography was performed and the finding was normal.

Histopathologically the lesions showed extensive fibrillar calcification and a proliferation of vessels surrounded by meningothelial cells (Fig. 1E).

**Discussion**

MA is a rare benign disorder characterized by hallmarks of meningioma and angiomatosis (6). The tumor grows very slowly (7) and has rarely been reported in the literature in English (1-18). It mostly affects children and young adults and was first described by Bassae and Nuzum in 1915 as an incidental autopsy finding in a 15-year boy; it was first named by Worster-Drought et al. in 1937 (1, 6). Seizures and headaches are the most common symptoms (2), and in nearly 50% of reported cases it is associated with NF; a review of the literature suggests that its association with NF2 is stronger than with NF1 (9, 10).

Though its pathogenesis is unknown, three possible theories have been suggested (11). The lesions could be hemorrhage, they could be secondary to direct invasion of brain tissue from a leptomeningeal meningioma, or could represent a vascular malformation.

The characteristics of MA are leptomeningeal calcification and meningovascular proliferation interwoven with fibrous connective tissue bands (12). Its pathologic criteria may be summarized as follows: leptomeningeal proliferation of nodules, whorls or bands of meningothelial (i.e. arachnoidal cap) cells exhibiting fibrillar calcification and a proliferation of vessels surrounded by meningothelial cells.
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(6). Calcification patterns vary from faint psammomatous to dense osteoid(7). MA affects the cerebral cortex in 90% of cases and usually occurs in the frontal or temporal lobes; in the third ventricle, thalamus, cerebral peduncles(2), and brain stem(13) it is rare. In our cases, lesions were present in all lobes of the brain, and frequency did not vary accordingly to location. An association with meningioma or oligodendroglioma has occasionally been reported(14, 15).

Although multifocal lesions have been described, MA usually involves well-demarcated solitary lesions (2, 14). Multiple lesions and associated cysts have not, though, been described on CT scan and/or MR images. We experienced six lesions in four patients with MA. In two of the four, the lesions were multifocal and associated with an eccentric cyst. At craniotomies, all lesions were well demarcated and superficially located. The masses were composed of popcorn-like, spotty, gyriform calcification, and in two cases, eccentric cysts were noted in the subarachnoid space. We think that the mechanism of extratumoral cyst formation is same as that of cystic meningioma. Wasenko JJ et al(17) suggest the cyst may form as the result of a ball valve mechanism with the gradual accumulation of cerebrospinal fluid in several sulci between the tumor and brain parenchyma. Our patients were older than most reported cases, and had no stigmata or family history of NF. We thought that our MA may be sporadic rather than associated with NF-2.

MA is always benign(5), and because total surgical removal is the treatment of choice and the prognosis after surgery is most cases excellent, accurate diagnosis is important. Differential diagnosis of radiologic images include meningioma, oligodendroglioma, granulomatous meningitis and parasitic diseases. However, radiologic characteristics are non-specific and differential diagnosis is not easy(10, 18).

In conclusion, MA is a benign cerebral lesion and may be seen in patients with or without NF. Its CT and MR characteristics are a cortical or subcortical mass with various calcifications and peripheral edema or gliosis, and it is usually a solitary lesion. Multifocal lesions and associated cyst formation may, however, be seen.

References

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

목 적: 수막혈관종(meningioangiomatosis 이하 MA로 약함)의 CT 및 MR소견을 알아보고자 하였다.

대상 및 방법: 4명(18-53세, 남자 1명, 여자 3명)의 MA환자를 대상으로 후향적으로 CT(n=4), MR(n=3)소견을 분석하였고, 모든 환자에서 병리조직 소견을 얻었다.

결 과: 모든 병변은 피질 및 피질하부에 위치하였고, 다양한 석회화[spotty(n=1), popcornlike(n=2), gyral(n=1)]와 병변과 동반되어 부종과 신경교증을 보여주었다. 두명의 환자에서 병변은 다발성으로 나타났고 센터낭증(eccentric cyst)이 동반되었다.

결 론: MA는 수술로서 치료가 가능한 양성질환으로 방사선학적 소견은 병변이 피질과 피질하부위에 위치하며 다양한 석회화와 동반된 부종과 신경교증이 특징이었다.