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Case Report

A Case of Infantile Meningioangiomatosis with a Separate Cyst

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Meningioangiomatosis (MA) is a rare congenital tumor that occurs mostly in 5-15 year old children. There have been only 5 cases previously reported that described the cystic nature within these tumors. We present a case of a MA accompanied by a separate macrocyst. A normally developed 2 year-old female patient presented with partial and generalized seizures. The brain computerized tomogram and magnetic resonance imaging revealed the presence of a calcified mass accompanied by a cyst in the right parietal area, surrounded by low density and high attenuation edema and hemorrhage. Upon right parietal craniotomy, a 1.6 cm × 1.2 cm × 0.5 cm sized plate-like, gray-white, slightly hard mass was seen and it was completely excised. Approximately 1 cm from the mass in the anterior lateral direction, a cyst was found and subsequent biopsy of the cyst wall revealed no tumor tissue, and therefore the cyst was not removed. Pathologic report demonstrated the meningioangiomatosis. Follow up examination 2 years later showed no recurrence of the tumor, and there was no evidence of neurological deficits. Authors suggest that cysts that arise in the surrounding tissues of tumors may not be tumor cysts, and do not require surgical removal.

KEY WORDS : Brain tumor · Cyst · Epilepsy · Infant · Neurofibromatosis.

INTRODUCTION

Approximately 100 cases of meningioangiomatosis (MA) have been reported in the literature, among which 16 patients were accompanied by neurofibromatosis type 2 (NF2)[16]. The age range of patients with NF2 is diverse from 11 months to 70 years. While sporadic MA usually occurs as a solitary lesion mostly in children and young adults and presents as seizures or persistent headaches, NF2-associated MA has been reported to occur as multiple lesions and usually are asymptomatic, and therefore are found incidentally or during autopsy[7,16,25].

The mechanisms involved in the development of MA tumors have not yet been elucidated, but it has been suggested that as tumors do not grow or display malignant features in the absence of NF2, this condition has been considered in the same context with hamartomas or vascular anomalies. However, there are some opinions that MA may be related to NF2 gene mutation. There have been only 5 reported cases of MAs accompanied by the presence of cysts, and no reports that described the separate location of the cyst[11,12,17,23]. We present a patient in whom no tumor cells were observed in the macrocyst wall that was separately surrounded by a MA, and which therefore supports the theory that MAs are developmental anomalies related to lesions such as hamartomas.

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CASE REPORT

A 2-year-old female patient presented with partial complex seizure attacks followed by generalized seizures at our emergency facility. After the seizures abated, she fully recovered her mental status. Since birth, development was normal and there was no significant neurological deficit, except for decreased visual acuity of the right eye and right ptosis. Brain computed tomography (CT) demonstrated a high-density mass accompanied by a cyst in the right parietal area with low density and high attenuation in the surrounding tissues, suggesting a calcified mass with surrounding edema and hemorrhage (Fig. 1A, E). The magnetic reso-

nance imaging (MRI) showed that there was a discrete cystic lesion in the right peritrigonal portion (Fig. 1B-D). The mass in the T2-weighted image showed a central low signal intensity and peripheral iso-signal in the lesion, suggesting central calcification with peripheral subacute stage hemorrhage, and a low intensity signal on T1-weighted images and high intensity signal on T2-weighted images, indicating moderate peritumoral edema (Fig. 1F, G). After enhancement there was no evidence of cyst enhancement, but the mass adjacent to the cyst demonstrated dense enhancement (Fig. 1G, H). Therefore, the patient was initially diagnosed with a low-grade cystic glioma or vascular anomaly with cystic change from old hemorrhage.

A right parietal craniotomy was performed with a dural incision. The exposed brain was seen as normally colored, and subsequently localization was accomplished employing a neuronavigator (Fig. 2A). A cortical incision followed by dissection to a depth of 1.5 cm revealed a slight gray discoloration and a longish appearing 1.6 cm × 1.2 cm × 0.5 cm sized, plate-like, gray-white, slightly hard mass which was completely resected (Fig. 2B-H). After removal of the mass, a 1.8 cm × 1.5 cm × 1.5 cm sized, separate cyst was found about 1 cm anterolateral from the mass lesion (Fig. 2I). As multiple frozen biopsies of the cyst wall failed to detect any tumor tissues, and the cyst was close to the motor cortex and fibers, the cyst was not removed. The dural incision was tightly closed, the skull bone flap fixed, and the scalp was finally closed. The course of the procedure was uneventful and the final pathology report was a MA (Fig. 3). The patient was discharged 1 week after the procedure without any neurological complications. Follow up examination 2 year later demonstrated no recurrence of the tumor, and no

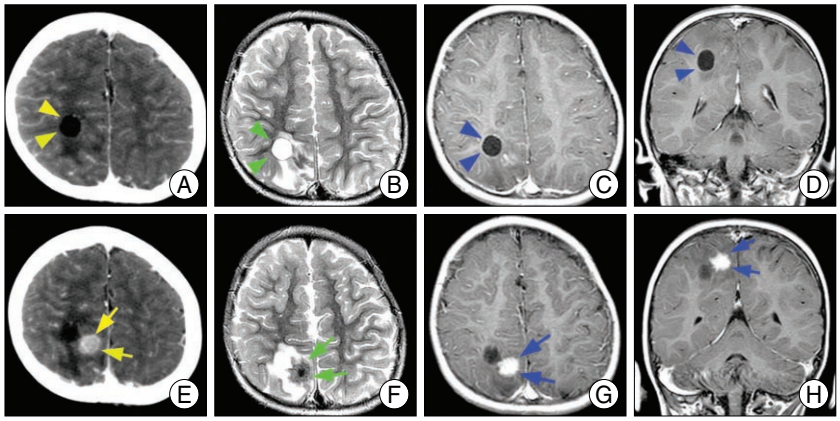

Fig. 1. Brain computed tomography and magnetic resonence imaging findings. Non-enhanced studies show a low-density cyst (yellow arrowheads) in the right parietal area with adjacent low-density area in the posterior medial direction in the computed tomography (A) that is seen as high signal intensity (green arrowheads) in the T2-weighted axial image of magnetic resonance imaging (B) and low signal intensity (blue arrowheads) in the T1-weighted axial and coronal images (C, D). Enhanced computed tomography demonstrate enhanced mass lesion (yellow arrows) (E) and enhnaced magnetic resonence imagings show a mass of low signal intensity (green arrows) and surrounding high signal intensity in the T2-weighted axial image (F) and high signal intensity (blue arrows) in the T1-weighted axial and coronal images (G, H) suggesting a calcified hemorrhagic mass lesion with surrounding edema.

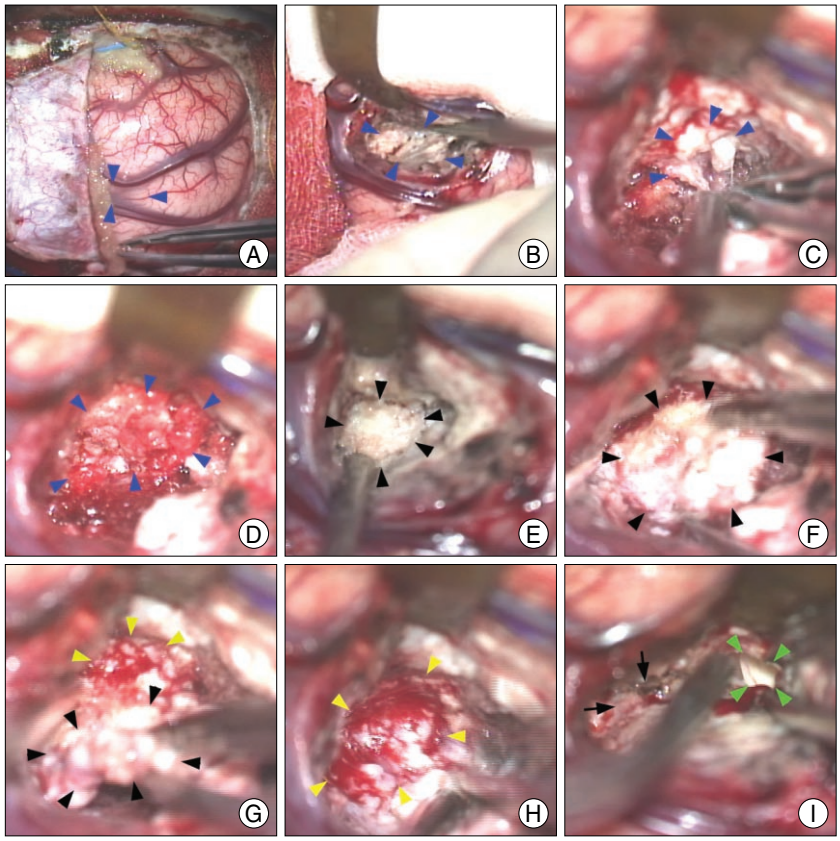

Fig. 2. Intraoperative findings. A : Neuronavigator system localized subcortical mass lesion area (blue arrowheads) between two large draining veins in the right parietal lobe. B : A cortical incision is followed by dissection to a depth of 1.5 cm revealed a slight gray discoloration (blue arrowheads). C : Anterior margin of mass lesion (blue arrowheads) has been dissected and resected. D : After removal of cortical layer, superficial lesion is exposed (blue arrowheads). E and F : After removal of superficial layer lesion, a longish appearing 1.6 cm × 1.2 cm × 0.5 cm, plate-like, gray-white, slightly hard calcified mass lesion (black arrowheads) is seen and dissected from the periphery. G and H : After calcified plate-like lesion (black arrowheads) has been removed as a one block, nomal subcortical layer is exposed (yellow arrowheads). I : After removal of the mass, a 1.8 cm × 1.5 cm × 1.5 cm, separated cyst opening (green arrowheads) is found about 1 cm anterio-lateral from the area (black arrows) of the mass lesion, that is not being removed.

other problems or neurological deficits were observed.

DISCUSSION

Since the first description of MA in 1915, there have been approximately 100 cases reported to date worldwide. Sixteen cases have been reported as NF2-associated MA, and these patients were characterized by predominant incidence among adults and no reports of a case in children less than 5 years of age[16]. To date, 84 cases of sporadic MA have been reported mainly in children and young adults, and among them 9 cases have been reported in children under 5 years of age[16]. Therefore, sporadic MA is very rare in children under 3 years of age, and to date only 5 cases have been described in the literature, thus making our case the sixth (Table 1)[1,2,15,22]. Sporadic MA has been reported mainly in the temporal lobe (33% of 84 cases), followed by the frontal lobe (25%), and the parietal and occipital lobes, in decreasing frequency, but MA accompanied by NF has been shown to occur mostly in the frontal lobe[16]. Multiple MA lesions accompanied by NF occur in 35% of patients, but the incidence of sporadic MA is only 13%.

The most common symptoms are seizures and headaches. Seizures occur in 85% of sporadic MA patients, and comprise of complex partial (47%), simple partial (34%), and general tonic clonic seizures (11%)[25]. Seizures are characteristically intractable in many cases, and there has been report of a sudden unexplainable death in one patient[26]. As the mechanisms involved in the development of MA is not clear, immunohistochemistry is of little value. Pathologically, meningiovascular proliferation, perivascular cuffs of spindle cells proliferation, and perivascular connective tissue proliferation may be seen, but without malignant features. Neurofibrillary tangles seen during degeneration is observed in 1/5-5/6 of patients, there is almost no growth of the tumor prior to surgery, and there are no recurrences after surgical removal. These characteristics suggest that MA is more related to a malformation or hamartoma, rather than a true neoplasm[5,11,23]. Consi-

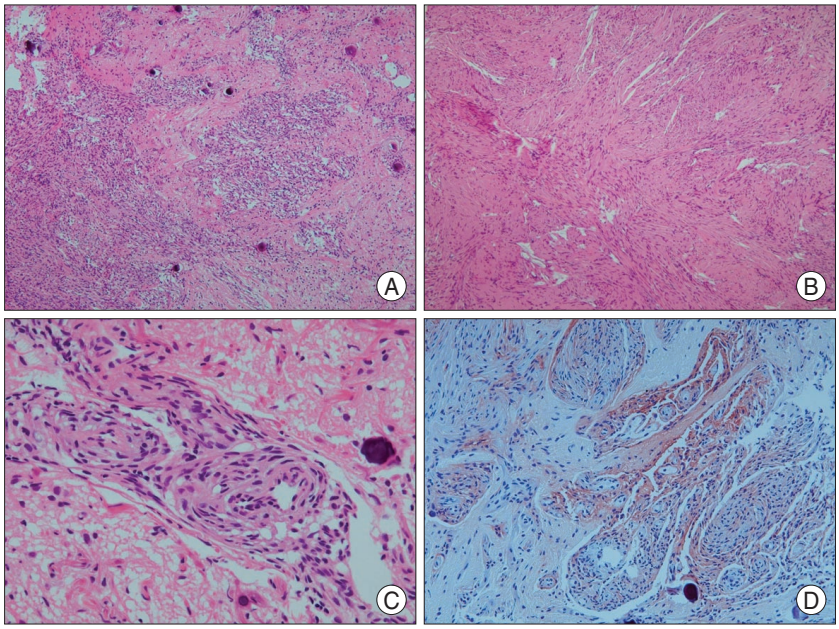

Fig. 3. Pathologic findings with immunohistological study. A : Pathologic examination of the tissues including the cerebral cortex shows meningioma-like cellular areas with occasional psamomma bodies (×100). B : Fibrous area with an interaced fascicular pattern is noted (×100). C : Perivascular meningothelial cell proliferation and psammomatous calicfication are seen (×400). D : Weakly positive staining for epithelial membrane antigen is observed at the perivascular area (×200).

Table 1. Summary for meningioangiomatosis developed in infants and toddlers less than 3 years of age. No case was associated with neurofibromatosis

Authors	Age/Sex	Symptoms and signs	Radiologic findings	Operation	Postoperative results
Aizpuru et al.[1]	2.5 yrs / M	Seizures	Dense calcification lesion with dense enhancement on the right parietal lobe	Biopsy	NA
Oka et al.[15]	16 mos / F	Seizures	Densely enhanced lesion in the left temporoparietal area	Total removal	No deficit; no seizure nor recurrence for 3 yrs and 5 mos
Tien et al.[22]	20 mos / F	Seizures	Low signal intensity on T1- and high signal intensity on T2-weighted MRI in the right occipital lobe	Partial resection	No deficit
Blumenthal et al.[2]	11 mos / M	First seizure attack	Focal calcified enhanced mass in right frontal lobe	Total removal	No deficit
Authors' case	2 yrs / F	First seizure attack	Focal calcified mass with separated cyst in right parietal lobe	Total removal of calcified mass except cyst	No deficit; no seizure for 2 yrs

mos : months, NA : non-available, yrs : years

dering the fact that growth is present in NF2-associated MA, mutation of the somatic or germline NF2 gene is suspected in sporadic MA. Even though Stemmer-Rachamimov et al.[20] confirmed that there were no NF2 gene mutations among 12 sporadic MA patients, a recent study reported 1 case of NF2 gene deletion, and therefore this needs to be further elucidated by future studies[19,20].

MA may be accompanied by other tumors such as astrocytomas, ependymomas, oligodendrogliomas, primitive neuroectodermal tumors, schwannomas, and hamartomas. Other accompanying vascular lesions are anterior cerebral aneurysms, venous angiomas, and vascular malformations, and occasionally encephaloceles[6,8,13,14,24]. The most common lesion reported are meningiomas, which was seen in 16 of MA patients. Although these meningiomas are not histologically malignant, the presence of brain invasion makes it essential to differentiate from a malignant meningioma[3,4,9]. It has been observed that there are no specific characteristics of MA on MRI or CT[10,18,25]. The most common site of occurrence is the frontotemporal region, and the appearance of the lesions has ranged from no contrast enhancement to strong enhancement[10,25]. Consequently, MA lesions have often been confused pre-operatively with low-grade astrocytomas, meningiomas with cortical invasion, and vascular malformations, and it has been stressed that the benign nature of this condition should proscribe unnecessary treatment[5]. In general, most CTs show a calcified enhancing lesion with surrounding low density, while the T1- and T2-weighted image MRI show hypointense signal and surrounding hyperintense signal on the T2-weighted image. In most patients, gadolinium enhancement has been reported to be minimal[7,16].

After surgical removal of the tumor, long-term seizures disappeared in 43% of patients, 30% of patients showed improvement, and 20% of patients did not require antiepileptic drug administration[25]. Partial removal of tumor has been shown to improve symptoms, but still total removal seems to be more effective[7,25]. If post-operative seizure control is not successful, then extension of the epileptic foci was seen to develop with time[21].

There have been 6 cases (including the present case) of MA accompanied by cysts which have all been reported to be located within or attached to the MA lesion (Table 2). In 2 cases the cyst was microcystic and which was visible only on microscopic examination, 2 cases were sufficiently large to be detected by MRI, while 1 case was a large cystic mass comprising of multiple cysts. However, there have no reports of a large macro-cyst that was completely separate from the main tumor, as in our case[11,12,17,23]. It has been suggested that the formation of this cyst is due to collection of trapped CSF by the tumor as in cystic meningiomas, but communication between the cyst and the subarachnoid space was not confirmed[11]. In our patient, pathologic examination demonstrated no tumor tissue present in the

Table 2. Summary for 6 cases of meningioangiomatosis accompanied with cysts. All cases were not associated with neurofibromatosis

Authors	Age (yrs) /Sex	Symptoms and signs	Radiologic findings	Characteristics of cyst	Operation	Postoperative results
Park et al.[17]	47/F	Headache and generalized seizures for 5 years	2×1×1 cm and 0.2×0.1×0.1 round calcified masses and eccentriccysts with edema in the left frontoparietal and right parietal lobes	Eccentric cyst within left lesion	Total removal of only left lesion	Free of seizures and headaches forthe 15 months since surgery
	53/M	Headache and generalized seizures for 2 years	Dense round calcified lesions with eccentric cysts in the left frontal and parietal lobes	Multiple macrocysts within lesions	Total removal of both frontal and parietal lesions	Free of seizures and headaches for the 7 months since surgery
Kuchelmeister et al.[12]	58/M	Headache, forgetfulness for 10 years	Multicysts with meningioma in the right frontal lobe	Septated large multiple cysts	Total removal	Forgetfulness, no recurrence for the 2 years since surgery
Kobayashi et al.[11]	14/M	Intractable seizure (complex partial and generalized tonic-clonic) for 3 years	2-3 cm mass of hypointensity on T1 and isointensity on T2 in the left frontal lobe with small cyst	Small cyst in the periphery of the lesion	Total removal	Seizure controlled with two antiepileptic drugs for 10 month
Wang et al.[23]	12/M	Intractable seizure (complex partial) for 7 years	4×3×2 cm cystic lesion in the left frontal lobe	Large cyst within lesion	Lesionectomy	Seizure controlled with antiepileptic drug for 10 month
Authors' case	2/F	First partial and general seizure attack	Plate-like, gray-white, slightly hard calcified 1.6 cm× 1.2 cm × 0.5 cm mass	Separated 1.8 cm× 1.5 cm ×1.5 cm cyst	Total removal of calcified mass except cyst	No recurrence for the 2 years since surgery

cyst wall. The cyst was separated from the MA, completely separated within the parenchyme and with no communication between the cyst and subarachnoid space. The above findings suggest that this cyst was not formed by the tumor or a portion of the tumor trapping the CSF. Therefore, considering the above findings, we propose that MA is a developmental anomaly rather than a neoplasm, and that the formation of the cyst is also an accompanying developmental anomaly. If this is true, then the cyst forming around the tumor would not be a true tumor, and removal of the cyst may not be essential, especially if it is located in the eloquent area, and also if there are no demonstrable tumor cells in the cyst wall biopsies.

CONCLUSION

Although rare in infants and toddlers, MA should be considered as a differential diagnosis, and as accompanying cysts in the eloquent area surrounding the tumor may not be a tumor, careful consideration should be given with respect to removal of the cyst.

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