Case Report

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Imaging Features and Pathological Correlation in Mixed Microcystic and Angiomatous Meningioma: A Case Report

미세낭종성 혈관종성 혼합 수막종의 영상 및 병리 소견: 증례 보고

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Meningiomas are the most common intracranial tumors. However, microcystic and angiomatous meningiomas are very rare subtypes that present unusual imaging findings. Hence, radiological diagnosis of these tumors can be challenging. We herein describe a case of mixed angiomatous and microcystic meningioma in an 81-year-old male. MRI revealed an extra-axial mass with high T2 signal intensity, measuring 1.5 cm in diameter, with multiple tiny intralesional cysts and entrapped peritumoral cyst formation. After tumor resection, a histopathological diagnosis of mixed angiomatous and microcystic meningioma was made.

Index terms Meningioma; Microcystic Meningioma; Angiomatous Meningioma; Magnetic Resonance Imaging

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INTRODUCTION

Meningiomas are the most common brain tumors, accounting for more than one-third of all primary intracranial neoplasms (1). The World Health Organization subdivides grade I meningiomas into nine subtypes according to the cell type (1). Microcystic and angiomatous meningioma are rare histological subtypes, and they can be difficult to diagnose because they show unusual radiological findings, such as cystic change and a higher T2 signal intensity (SI) solid portion than ordinary meningiomas (2, 3). Here, we

describe a case of mixed microcystic and angiomatous meningioma and discuss the radiological findings.

CASE REPORT

An 81-year-old male presented to our clinic with a chief complaint of slowly progressive cognitive impairment over several months. He also experienced intermittent headaches. Neuropsychological tests (Seoul Neuropsychological Screening Battery) showed reduced function of the bilateral frontal lobes and right parieto-temporal lobe. He had no notable medical history other than the use of a positive airway pressure device to treat obstructive sleep apnea.

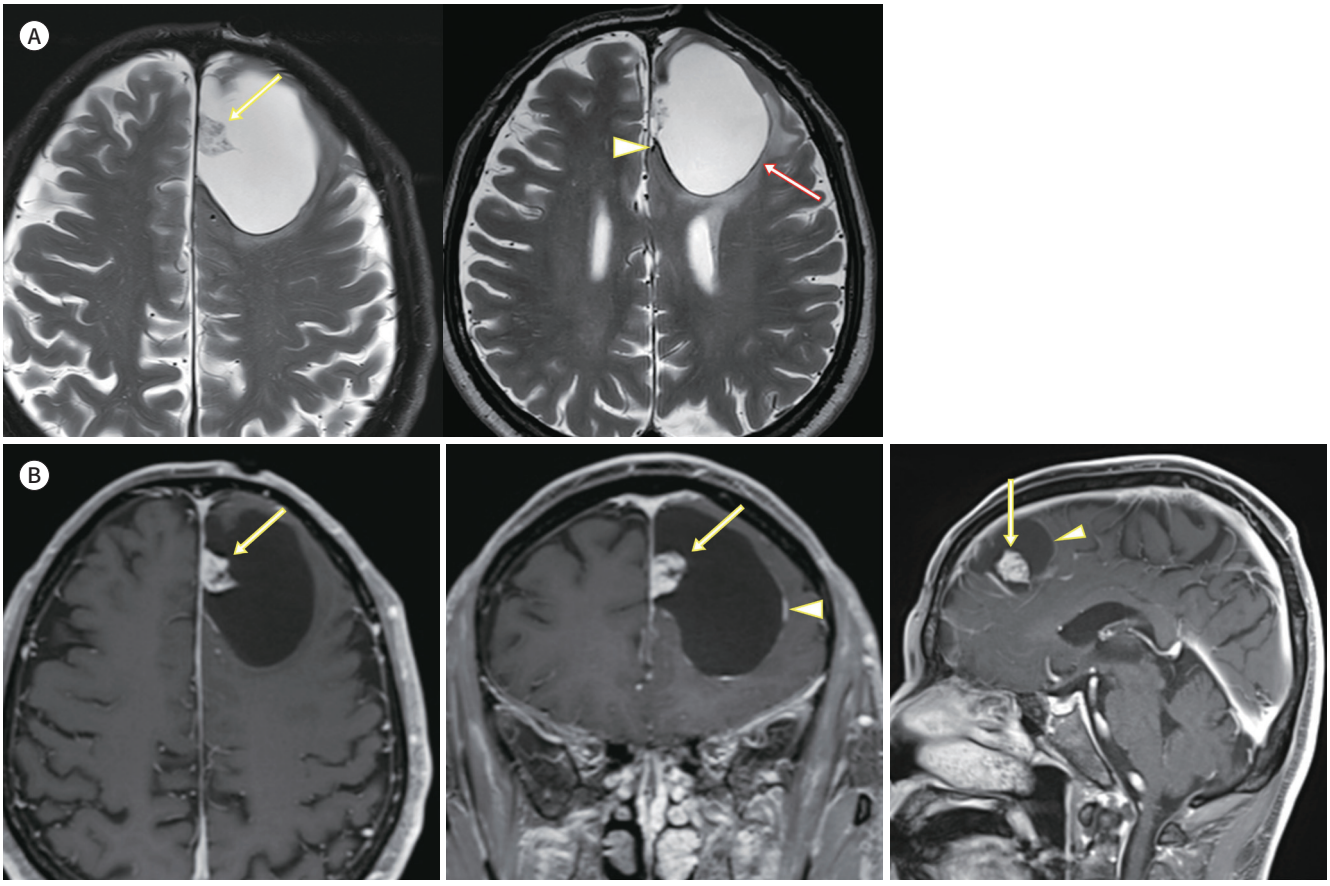
Brain MRI showed a well-defined, predominantly cystic, partially solid mass with mild peritumoral brain edema. The mass measured 6.0 cm × 5.7 cm × 4.2 cm and was located in the left frontal lobe. Although the mass was broadly attached to the anterior part of the falx cerebri, there were no other imaging findings of extra-axial tumors, such as cerebrospinal fluid (CSF)/vascular cleft or dural tail sign. A peripherally located, large cystic portion around the mass (Fig. 1A) was invaginated into the brain parenchyma like a mushroom. Therefore, it was difficult to identify whether the origin of the lesion was extra-axial, and we suspected that the mass to be an intra-axial tumor such as pleomorphic xanthoastrocytoma, even though the adjacent left anterior cerebral arteries seemed to be located between the mass and the cerebral cortex (Fig. 1A). However, a retrospective review after more comprehensive information available showed that the mass was more likely to be an extra-axial mass originating from the falx cerebri as some atypical meningiomas invaginate into the brain parenchyma like a mushroom (4).

T2-weighted imaging showed that the large cystic portion around the mass was a homogeneous fluid with high SI (Fig. 1A). It showed slightly a higher SI than the CSF on fluid attenuated inversion recovery (FLAIR) images (not shown here), but no solid mural nodules or enhancing solid portion were identified. Surgical inspection and pathological examination revealed that the thin-walled, peripheral, low SI rim (Fig. 1A), which had several small thin enhancing portions of the cyst wall on MRI (Fig. 1B), was a reactive fibrous membrane rather than the wall of the cystic tumor. Therefore, it was presumed that the cyst was formed by entrapment of peritumoral fluid.

The intracystic enhancing solid tumor measured 1.5 cm × 1.4 cm × 1.3 cm, was broadly attached to the falx cerebri, and presented as a heterogeneously mixed lesion composed of a bright T2 SI solid portion with multiple tiny intralesional cysts (Fig. 1A, B). The solid portion showed strong enhancement in all areas, except the small cystic portions, leading to a bubbly or reticular appearance (Fig. 1B). This was unlike typical meningioma solid portions, which show a homogenous, low T2 SI solid mass with homogeneous enhancement.

Tumor removal was planned because the cyst had a mass effect in the adjacent left frontal lobe and because the patient had intermittent headaches and progressive cognitive impairment. Median frontal craniotomy was performed. The cyst was exposed to the medial part of the left superior frontal gyrus. The cyst wall was a thickened fibrous membrane. The cyst wall was incised and cystic fluid was drained to relieve pressure. The cystic fluid was clear

Fig. 1. An 81-year-old male with mixed angiomatous and microcystic meningioma in the left frontal region.
A. T2-weighted axial MR images show a well-defined, predominantly cystic, partially solid mass with mild peritumoral brain edema. The mass measures 6.0 cm × 5.7 cm × 4.2 cm in the left frontal lobe and its large, peripherally located cystic portion shows a slightly higher SI than the cerebrospinal fluid on fluid attenuated inversion recovery sequence (not shown here), with invagination into the brain parenchyma like a mushroom. The wall of the mass shows a thin rim with low SI (red arrow). The 1.5 cm × 1.4 cm × 1.3 cm, intracystic, solid mass (yellow arrow) shows a solid portion with high T2 SI and small intratumoral cysts. Surgical inspection identified the peripheral cystic portion to be an entrapped peritumoral cyst formation with a reactive fibrous membrane. Note that the signal void (arrowhead) by the left anterior cerebral artery is located between the cerebral cortex and the mass. When the signal void is traced, it turned out to be a displaced left anterior cerebral artery. A more comprehensive retrospective review of this information indicated that the tumor is completely extra-axial, with broad attachment to the falx cerebri.
B. Post-contrast, T1-weighted, axial, coronal, and sagittal images show strong enhancement of intracystic tumor solid portion, while the small cystic portions, with a bubbly or reticular appearance (arrows), show no such enhancement. The large cyst wall with several thin, linear enhancements (arrowheads) was later identified as a fibrous membrane.
SI = signal intensity



xanthochromic fluid. A soft pinkish mass was found on the falx cerebri within the cyst. It was broadly attached to the falx cerebri and was an entirely extra-axial mass. The mass was removed *en bloc* and the underlying adjacent falx was resected. Several specimens were obtained from the inner part of the peripheral cyst for frozen section examination, which showed fibrous tissues without tumor cells.

On histopathological examination, the mass comprised two distinct histological features. There were areas showing numerous densely packed blood vessels that were positive for CD34 on hematoxylin & eosin and immunohistochemical analysis (Fig. 1C, D). These were admixed with meningothelial cells that were positive for epithelial membrane antigen and

were wrapped around the blood vessels (Fig. 1C), consistent with angiomatous component. The other areas revealed a loosely reticular, lace-like appearance with a myxoid extracellular component and numerous extracellular microcystic spaces (Fig. 1E), compatible with their identification as the microcystic meningioma component. These two components were intermixed with each other; therefore, a diagnosis of mixed angiomatous and microcystic meningioma was made.

One day after surgery, the patient experienced a seizure episode; hence, anticonvulsive treatment was initiated. The patient was discharged from the hospital 15 days after surgery without complications. No remnant tumor or recurrence at the operation site was observed at the 2-month follow-up MRI.

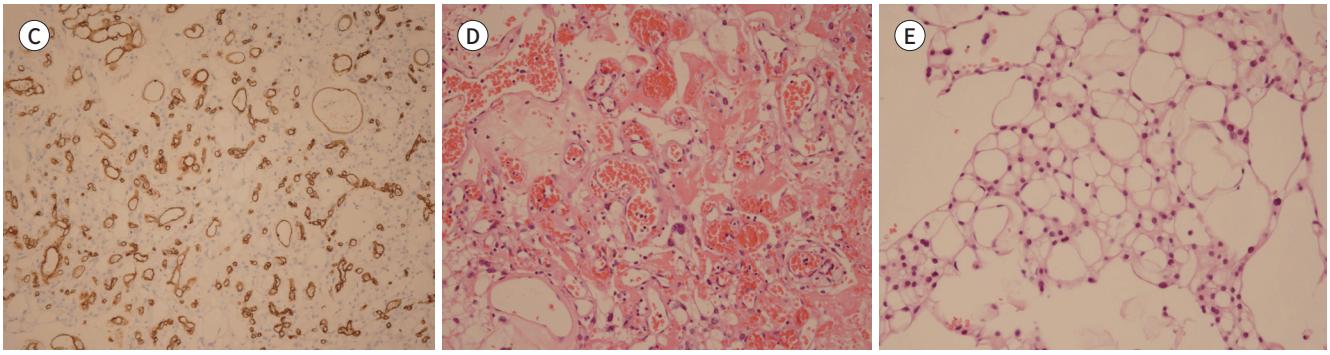
This case report was approved by the Institutional Review Board of our institute (IRB No. DMC 2021-03-016).

DISCUSSION

Microcystic meningioma is the rarest subtype of meningioma. It is characterized by extracellular myxoid background and extracellular microcysts. Microcystic meningioma was first described by Masson in 1956, who labeled the lesion as "humid" meningioma because of its gross morphology, which is usually soft, occasionally with a glistening cut surface (2). It was classified as a distinct subtype of meningioma by the World Health Organization in 1993 (5).

Microcystic meningiomas can be difficult to diagnose radiologically because they are rare and show unusual imaging features, such as a higher T2 SI solid portion than ordinary meningiomas, which show homogeneous low T2 SI solid masses with homogenous enhancement. These unique high T2 SI solid portions are thought to originate from the abundant extracellular myxoid background, low cellularity, and extracellular micrometer-scale microcysts. Angiomatous meningiomas also show high SI on T2WI, similar to microcystic meningioma, due to their numerous dilated blood vessels and vacuoles (3). Their intrapixel or intravoxel partial volume average of microcystic or angiomatous components can lead to T2 prolongation, un-

Fig. 1. An 81-year-old male with mixed angiomatous and microcystic meningioma in the left frontal region.
C. An angiomatous meningioma area shows numerous densely packed blood vessels admixed with meningothelial cells wrapped around the blood vessels (H&E stain, × 100).
D. Vessels are immunostained for CD34 (× 100).
E. A microcystic meningioma area shows a loosely reticular, lace-like appearance with numerous extracellular cystic spaces (H&E stain, × 200).
H&E = hematoxylin & eosin



like ordinary meningiomas, which are highly cellular. When we encounter high T2 SI solid portions in meningiomas or extra-axial masses, we should carefully check for the presence of the extracellular myxoid components or microcysts within the mass, which may indicate the rare microcystic meningioma. Additionally, prominent intratumoral signal voids and intratumoral cystic changes are can be observed in angiomatous meningioma (3). In our case, intratumoral signal voids was not observed.

The incidence of visible cysts in meningiomas is approximately 1.6%–11.7% (6). In addition to the previously noted microcysts, which are not observed on imaging, microcystic meningiomas also can have relatively large, visible intratumoral cysts or cystic change (7). These radiologically visible cysts can contribute to the characteristic reticular enhancement of meningiomas (8). Two patterns of contrast enhancement in microcystic meningiomas have been described. The first and most common is homogeneous avid enhancement similar to that of ordinary meningioma (7) which presumably occurs in the absence of larger, visible cysts. A second, less frequent pattern of marginal and reticular enhancement is seen in approximately 27% patients (7). This pattern is thought to be due to variably sized, larger cysts measuring several millimeters, which are not enhanced on the MRI contrasted with strong enhancement of its solid portion. Thus, the reticular enhancement of microcystic meningiomas may be due to the enhanced solid tumor portion combined with the variably sized, relatively small, visible cysts. This reticular enhancement is considered a unique finding of microcystic meningioma, with a specificity of nearly 94% (2).

Peritumoral edema is a frequent finding in microcystic meningiomas, occurring in 87.5% cases compared to that in 40%–60% ordinary meningioma cases (7); in our case, only mild peritumoral edema (Fig. 1A) was observed.

Peritumoral cysts formation of meningiomas are often reported (6). Various explanations for them have been suggested, including presence of reactive arachnoid cyst, direct secretion and its encapsulation of fluid by tumor cells, and loculated or entrapped CSF by scar tissue between the brain surface and the meningioma (9). Nauta et al. proposed four relationships between a meningioma, its associated cyst, and the surrounding brain (10). In the present case, reactive fibrous membranes of the peritumoral cyst wall entrapped fluid between the tumor and brain, corresponding to Nauta type IV. Reactive fibrous membrane formation could be explained by long-standing irritant secretion from ruptured intratumoral cyst, as in our case, or oozing fluid from the loose tumor surfaces with high interstitial pressure like that from hemangioblastomas or some meningiomas. Eventually, resultant CSF confinement or entrapment of peritumoral fluid with enough positive pressure to form a space may occur due to this fibrous adhesion. In our opinion, that irritant fluid can lead to fibrous membrane formation as we usually observe it in the case of empyema or chronic subdural hematoma, although we didn't find exactly same case reported. According to our literature review, no reports also have described microcystic meningiomas associated with peritumoral cysts.

In conclusion, we have reported a rare case of mixed microcystic and angiomatous meningioma, with a high T2 SI solid portion and intratumoral tiny cysts that have reticular enhancement. We should consider the possibility of microcystic meningioma when we encounter an extra-axial mass or meningioma with a high T2 SI solid portion or small cysts that show reticular enhancement. In the present case, it was difficult to determine whether the mass was

intra-axial or extra-axial due to the accompanying peritumoral cyst, which invaginated into the brain parenchyma like a mushroom.

Author Contributions

Conceptualization, C.K., K.D.Y., K.H.J.; data curation, CK., K.D.Y., H.G., P.S.; investigation, C.K, K.D.Y.; methodology, C.K., K.D.Y.; project administration, C.K, K.D.Y.; resources, C.K., K.D.Y., H.G., P. S.; supervision, K.D.Y.; visualization, C.K., K.D.Y., P.S.; writing—original draft, C.K., K.D.Y., H.G., P.S.; and writing—review & editing, all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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미세낭종성 혈관종성 혼합 수막종의 영상 및 병리 소견: 증례 보고

최건우[1] · 김대윤[1*] · 김현진[1] · 황교준[2] · 김미경[2] · 김현곤[2] · 백소야[3]

수막종은 가장 흔한 두개내 종양이지만, 그 아형인 미세낭종성 및 혈관종성 수막종은 매우 드물고 특이한 영상 소견으로 인해 영상의학적 진단이 어려울 수 있다. 이에 저자들은 미세낭종성 혈관종성 혼합 수막종으로 진단된 81세 남자 환자 증례를 보고하고자 한다. 자기공명 T2 강조영상에서 병변은 왼쪽 전두부에 약 1.5 cm 크기의 고신호강도의 축외 종양으로 보이고 있었고, 종양 내에 다수의 아주 작은 낭종들이 관찰되었으며, 종양 주위로 포획된 큰 낭을 형성하고 있었다. 수술적 절제를 통한 병리 검사상 미세낭종성 혈관종성 혼합 수막종으로 최종 진단되었다.

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