

Microcystic Meningioma - Unusual Variant of Meningiomas

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Microcystic meningioma is a rare variant of meningiomas. This unusual variant was originally described by Masson, who labeled it "humid". The computed tomographic scan or magnetic resonance images of these tumors resemble those of a glial or metastatic tumor with cystic or necrotic changes. There is no definitive method for differentiating cystic meningiomas from these more common tumors. But immunohistochemically, they share a similar pattern of positive staining for epithelial membrane antigen and vimentin with other meningiomas. Our case was a 34-year-old woman with a tumor mass on the right frontal area. She was admitted to hospital because of generalized tonic seizure. Grossly all of the tumor could be removed, and histopathologically this tumor was revealed to be a microcystic meningioma.

KEY WORDS : Microcystic meningioma · Cystic tumor.

Introduction

Meningiomas are common benign tumors accounting for 13% to 18% of intracranial neoplasms²⁷. They are generally known to be solid tumors and their classical appearance on the computed tomography(CT) scan and magnetic resonance(MR) image usually lead to a correct diagnosis.

Meningiomas have been subdivided into a variety of types, including meningotheliomatous, fibrous, transitional, psammomatous, angiomatous, and others. Microcystic meningioma is a rare variant of meningiomas showing microcyst formation.

The incidence of cysts in association with meningiomas is between 4%~7%^{23,28}. In 1956, Masson¹⁹ described similar lesions as humid and myxomatous meningiomas and recently WHO classification of brain tumors included the microcystic meningioma as a subtype of meningiomas¹⁶. The authors encountered a case of microcystic meningioma. So, we report this case with clinicopathological features.

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Case Report

A 34-year-old woman was admitted to our hospital because of generalized tonic seizure. It was the first time seizure and other neurologic examinations were normal. She did not have any other past medical history.

Axial CT scan with enhancement showed a large cystic lesion at right frontal area with densely enhanced intramural solid portion. Surrounding cerebral edema was noted(Fig. 1). MR image also displayed a large cystic lesions at the right frontal area with a small enhanced component(Fig. 2A).

Coronal T1-weighted MR image with gadolinium enhancement showed a lesion attached to the falx cerebri with surrounding cerebral edema of right frontal lobe and corpus callosum(Fig. 2B).

Right carotid angiogram revealed a parasagittal blush fed by the external carotid artery(Fig. 3).

At surgery, all tumors were tightly attached to the falx cerebri and were extra-

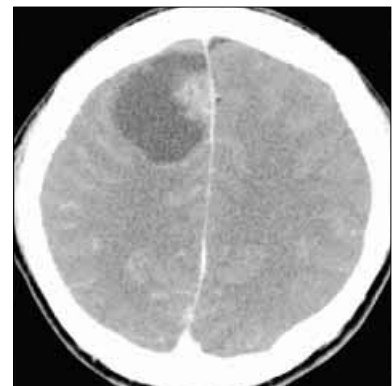


Fig. 1. Brain enhanced computed tomography. About 4cm sized cystic mass with cerebral edema at the right frontal lobe and genu of corpus callosum. No visible associated hypo-ostosis of calvarium.

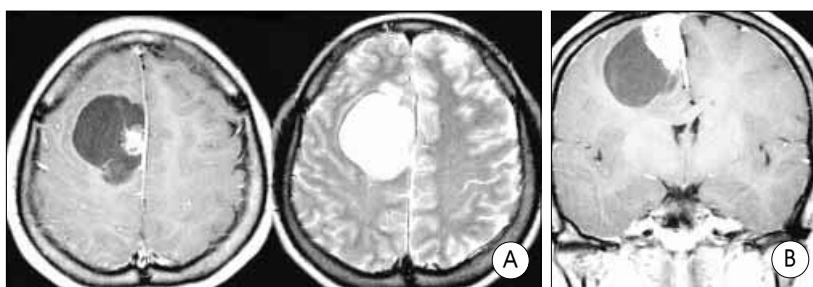


Fig. 2. Brain magnetic resonance(MR) imaging with gadolinium enhancement. About 5.3 x 4.3cm sized cystic tumor at the right frontal lobe with intramural solid portion showing strong enhancement on postcontrast scan. Coronal T1-weighted image with gadolinium enhancement shows a lesion attached to the falx cerebri with surrounding cerebral edema of the right frontal lobe and corpus callosum(A : axial view, B : coronal view).



Fig. 3. Right carotid angiogram revealing a parasagittal blush fed by the external carotid artery.

axially located.

Tumor was totally excised. The tumor had cystic component filled with yellowish fluid and the tumor margin was well demarcated.

On histologic examination, the tumor cells having xanthomatous cytoplasm and long cytoplasmic process are observed.

The background is loose mucinous with many small cysts formed by cytoplasmic process (Fig. 4A) .

The tumor cells reveal immunoreactivity for epithelial membrane antigen(EMA) along the cytoplasmic membrane (Fig. 4B). Postoperatively, she was discharged 9 days later without neurologic deficit.

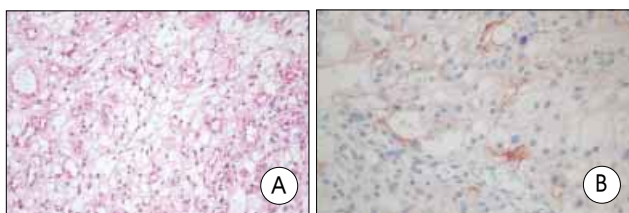


Fig. 4. A : Histologically characteristic loose mucinous stroma by many small cysts is observed. The tumor cells have xanthomatous cytoplasm and long cytoplasmic processes constituting microcysts (H&E, x200). B : On immunohistochemical stain, the tumor cells reveal immunoreactivity for EMA along the cytoplasmic membrane (x400).

Discussion

Meningiomas show various histological features, and many subtypes. Cystic meningiomas are rare

subtypes. Although this feature is unusual as meningiomas, this type of tumor has been described as one of the histological variants of meningiomas under the term of micro-cystic, humid, myxomatous, or vacuolated meningioma. Penfield⁵⁾ was the first to describe cyst formation in a meningioma. Previous descriptions of ‘humid’ and myxomatous meningioma probably represent microcystic meningiomas¹⁹⁾. The term ‘microcystic’ was suggested by Kleinman et al at 1980¹⁷⁾. Ito et al proposed to call this type of meningioma ‘arachnoid trabec-

ular cell meningioma’ to avoid confusion and to clarify the nature of the tumor at 1991¹²⁾. But according to the new WHO classification of brain tumors, the term microcystic meningioma is available¹⁶⁾.

Cystic meningiomas are more common in the pediatric age group than in adult. They are seen in 10%~19% of all pediatric meningiomas, compared with only 2%~4% in adult^{1,13)}. The most frequent location of cystic meningiomas is on the cerebral convexity and the parasagittal region is the second most frequent location^{9,18)}. These are also typical locations for other usual types of meningiomas. There is a slight female preponderance, similar to most other meningiomas²¹⁾. The clinical behavior of these tumors appears to have no difference from other benign meningiomas. The symptoms of meningiomas are variable according to the location and size of tumors. Increased intracranial pressure sign, motor weakness, seizure, visual disturbance and cerebellar signs are relatively common symptoms¹⁵⁾. Microcystic meningioma also shows the slow progression of symptoms²⁶⁾. Some authors reported the rapid onset of symptoms²⁶⁾. But it might be due to enlargement of cyst and edema rather than the tumor itself. Four main theories of edema formation exist ; disturbed blood brain barrier, mechanical compression, vascular compression and secretion of edematogenic factor⁸⁾. It means that peritumoral edema in meningiomas is not related with the grade of malignancy such as in gliomas. Cystic meningiomas are difficult to diagnose pre-operatively because their radiologic finding mimic that of glial tumors or metastatic tumors with a cyst and contrast enhancing tumor nodule. In one report, correct pre-operative diagnosis was possible in only 50% of cases¹¹⁾. The occurrence of a cystic tumor in a typical location for a meningioma may be helpful in the diagnosis, and the recognition of a relatively broad surface of contact with the dura is of great value. A coronal MRI will help to visualize the enhancing

mural nodule and its attachment to the falx and the dura^{6,26}. Additional diagnostic clues are the presence of a meningeal vascularization. The diagnosis of a cystic meningioma can be made from angiographic study that includes an external carotid injection, as gliomas are rarely fed via the external carotid artery⁵. In this case, the meningioma was preferred due to angiographic findings. The origin of meningiomas has generally been considered to be the arachnoid cap cells, with constitute the arachnoid membrane and trabeculation in normal situations¹⁴.

Characteristic features include prominent interdigitation of cell membranes of adjacent cells, frequent desmosomes and the presence of intracytoplasmic filaments, measuring approximately 10nm in thickness^{3,4}. The cell whorls of concentrically arranged polygonal cells are encountered. Psammoma bodies, which are round, laminated, calcified bodies, are occasionally seen in connection with the cell whorls. Microcystic meningiomas reveal some different findings from those of classic meningiomas. Microcystic meningiomas possess distinct histological features, with numerous cystic spaces filled edematous fluid and lined by stellate-shaped meningothelial cells²². Fukuoka et al believed that the cystic spaces within the cell clusters or sheets are characteristic of microcystic meningioma¹⁰. The immunohistochemistry of microcystic meningiomas is similar to other meningiomas. Meningiomas are generally considered to be mesenchymal origin, arising from the arachnoid membrane, although the tumors show distinct epithelial properties of interdigitating cytoplasmic processes and desmosomes. Their dual mesenchymal and epithelial properties are reflected by staining for both vimentin and EMA (epithelial membrane antigen) in most major types of meningiomas^{14,22,25}.

Concerning the pathogenesis of the marked extracellular dilatation in microcystic meningiomas, several theories have been proposed². Some studies indicated secretory activity of tumor cells or degenerative processes²². Michaud and Gagne suggested that the transduction of low-protein fluid could be responsible for the cyst formation²⁰.

As this report, meningiomas are richly vascularized and the vascular endothelium is partially fenestrated, so the transduction is possible. Some reports claimed that the formation of microcysts mimics the developmental process of the subarachnoid space in the embryo^{7,24}. But all of these reports say that the precise mechanism of microcystic formation remains unknown^{20,22}.

Histologically, microcystic meningioma may resemble hemangioblastoma with lipidication, pilocystic astrocytoma with a prominent microcystic component, and metastatic

adenocarcinoma^{10,20}. The astrocytoma presents stellate cells and PTAH (phosphotungstic acid-hematoxylin)-positive intracytoplasmic fibrils²⁰. The absence of malignant cytological features such as mitosis, necrosis and cerebral invasion is worth mentioning as it helps to exclude clear cell malignant tumors of other cellular origin²⁰. Unfortunately, sometimes even the intraoperative pathologic examination (frozen section) is unclear and only the postoperative histopathologic study will provide an accurate diagnosis²⁹. Nishio et al said an excessive subclassification of meningiomas may be undesirable, because the biological behavior of microcystic meningiomas corresponded to those of meningiomas in general²².

Conclusion

Cysts associated with meningiomas, though very uncommon, should be considered in the differential diagnosis of cystic lesions. Unfortunately, there is no definitive method for pre-operatively differentiating cystic meningioma from gliomas or metastatic carcinomas. External and internal carotid angiography has proved to be very useful, when in doubt. Recently, we experienced this uncommon type of meningiomas, so reported it with literatures.

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