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ORIGINAL ARTICLE

Characteristic features and proposed classification in 69 cases of intracranial microcystic meningiomas

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Abstract

Microcystic meningioma (MM) is a rare subtype of intracranial meningiomas, with clinical and radiologic features not well characterized in the literature. Based on our experience, we propose a classification system of intracranial MMs. We reviewed the medical records, radiographic studies, and operative notes of a group of consecutive patients with intracranial MM. The mean age of the 69 patients was 46.8 ± 10.6 years (range, 21–75 years). Three types of intracranial MMs could be identified. Type 1 MMs presented as a solid lesion, hypointense or isointense on T1WI, hyperintense on T2WI, and homogeneous or heterogeneous enhancement, and were found in 43 patients (67.2%). Type 2 MMs represented signals similar to CSF both on T1WI and T2WI, and faint reticular enhancement with marginal enhancement, and these were found in 7 patients (10.9%). Type 3 MMs consisted of cystic-solid or cystic lesion and were found in 14 patients (21.9%). Significant differences were observed among the different types of MMs for the following variables: sex, presence of severe peritumoral brain edema (PTBE), and extent of tumor resection. Females were found in all of patients with type 2 MMs, but were only 35.7% of those with type 3 MMs ($P = 0.018$). Severe PTBEs were more common among patients with type 1 MMs (55.8%) than among those with type 2 (14.3%) and type 3 MMs (14.3%) ($P = 0.007$). Type 1 MMs (97.7%) were associated with a significantly higher rate of gross total resection compared with the other two types (71.4 and 78.6%) ($P = 0.019$). Total length of hospital stay after craniotomy ranged from 4 to 30 days (median, 8 days). There were no significant differences in progression-free survival among the three types of MMs ($P = 0.788$). The current classification identifies three distinct types of intracranial MM based on their radiological findings and growth patterns. The type 1 MMs are more commonly associated with severe PTBE. Type 2 and Type 3 MMs have a higher predilection towards parasaggital location with venous involvement and therefore have a lower rate of gross total resection.

Keywords Microcystic meningioma · Classification · Radiology · Clinical features

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Introduction

Meningiomas are the most common intracranial brain tumors and account for more than a third of primary brain neoplasms [23, 36]. Microcystic meningioma (MM) is a rare benign variant of intracranial meningiomas. It was first described in the literature in 1983 [20] and was added to the World Health Organization (WHO) classification of brain tumors in 1993 [13].

Radiologically, there were a few reports dealing with the characteristics of MMs. The specific magnetic resonance image (MRI) findings include hypointensity on T1-weighted images (T1WI), hyperintensity on T2-weighted images (T2WI),

marginal and reticular enhancement, and high incidence of peritumoral brain edema (PTBE) [7, 12, 14, 19, 25, 30, 32].

However, these previous clinical and radiological observations were obtained by examining only small numbers of cases of this rare meningioma subtype (mostly case reports). Few systematic studies of the different types of intracranial MMs has been reported, and a classification system of these MMs is not available. We collected 69 patients with intracranial MMs from one of the world's largest neurosurgical center for a systematic investigation. Based on the results of this study, we propose a comprehensive but simple classification of intracranial MMs.

Method

Patient population

This study included 69 consecutive patients harboring intracranial MMs that were surgically treated at the Beijing Tiantan Hospital from 2008 to 2016. The clinical data, surgical records, and follow-up information were reviewed retrospectively. The following information was recorded: patient age, sex, clinical manifestation, duration from onset to admission, lesion size, location, CT and MRI features, extent of resection, and surgical outcomes. The Beijing Tiantan Hospital Research Ethics Committee approved the study.

Image evaluation

Two radiologists independently evaluated the radiological data. Signal intensities of the meningiomas on T1WI and T2WI were recorded as hypointense, isointense, or hyperintense relative to the intensity of the gray matter. The pattern of contrast enhancement after Gd administration was divided into homogeneous or heterogeneous involving some or all of the tumor, and reticular or marginal enhancement involving most of the tumor. Tumor size was recorded as the measurement of maximal diameter based on radiology. PTBE was evaluated on T2WI and graded as absent (edema index [EI] < 0.1), mild ($0.1 \le EI < 1.0$), moderate ($1.0 \le EI < 2.0$), or severe ($EI \ge 2.0$) [24, 35].

Pathological examination

Fresh paraffin-embedded tumor tissue was cut into 5-mm sections and stained with hematoxylin and eosin. The microscopic pathologies of 69 cases were reviewed by two experienced neuropathologists according to the criteria of the 2016 WHO classification of meningiomas [18].

Surgical outcome and postoperative follow-up

The extent of resection was graded based on operative notes and confirmed with postoperative MRI (whenever available) and was classified according to Simpson's grading system [33]. Simpson grade I/II was defined as gross total resection (GTR) and grade III/IV was defined as subtotal resection (STR). Patients were followed up after surgery. Postoperative complications, progression-free survival (PFS), and overall survival (OS) were recorded. Progression of MM was defined according to the radiological findings after tumor removal. PFS was defined as the time between initial surgery and tumor progression on radiology. OS was defined as the time between initial surgery and death.

Statistical analysis

IBM SPSS statistics for Windows version 19.0 (IBM Corporation, Armonk, NY, USA) was used for all statistical analyses. Comparisons of three types of MMs were carried out using the chi-square test for dichotomous variables and the Mann–Whitney U test for continuous non-parametric data. Probability values were obtained from two-sided tests, with statistical significance defined as $p < 0.05$.

Result

Incidence and clinical features of intracranial microcystic meningiomas

Sixty-nine cases of pathologically confirmed intracranial pure MM were identified. Approximately 11,000 patients with MMs were surgically treated in our center from 2008 to 2016. The incidence of intracranial MM was approximately 0.63%.

Patient demographics and baseline characteristics are summarized in Table 1. The mean age of these 69 patients was 46.8 ± 10.6 years (all means are expressed ±SD) (range, 21–75 years). There were 41 females and 28 males. Histologic examination revealed 68 WHO grade I meningiomas (98.6%) and one grade II meningioma (1.4%) with brain invasion. MIB-1 labeling index in most of the patients was < 3.0%. The duration from onset of symptoms to admission ranged from 1 week to 8 years (median, 3 months). The chief complaints included headache or raised intracranial hypertension in 24; seizures in 17; limb weakness in 13; numbness in 9; dizziness in 8; blurred vision in 7; aphasia and memory impairment in 4, respectively; behavioral change, hyposmia, drinking cough, and hearing loss in 2, respectively; dysphagia and ataxia in 1, respectively. Six patients (8.7%) were asymptomatic; their tumors were found incidentally on

Table 1 Clinical and demographic characteristics of 69 intracranial microcystic meningiomas

Characteristic	No. (%)
Age at diagnosis (years)	
Median (IQR)	47 (12.0)
Mean*	46.8 ± 10.6
Range	21–75
Sex	
Male	28 (40.6)
Female	41 (59.4)
Histology	
WHO grade I	68 (98.6)
WHO grade II	1 (1.4)
Duration from onset to admission	
Median	3 months
Range	1 week–8 years
Primary or recurrent lesion	
Primary	66 (95.7)
Recurrent	3 (4.3)
Preop neurological deficit	
Yes	26 (37.7)
No	43 (62.3)
Preop KPS score	
Median	90
Range	50–100
Duration of follow-up	
Median	49 months
Range	5 months–9 years
Recurrence	1

IQR, interquartile range; *WHO*, World Health Organization; *KPS*, Karnofsky performance status

*All means are expressed ±SD

neuroimaging, such as CT and MRI; they underwent operations because of marked PTBE, or tumor diameter greatly exceeded 3 cm with/without progressive growth. Of the 69 MMs, 66 were primary lesions and 3 were recurrent lesions. Two MMs were associated with hydrocephalus, while 1 MM was accompanied with cerebellar cavernous angioma. The median preoperative KPS score was 90 (range, 50–100).

Radiological findings of intracranial microcystic meningiomas

The radiological data are summarized in Table 2. The lesion was located in the convexity in 32 cases, in the parasagittal in 21 cases, in the falx in 5 cases, in the tentorial in 2 cases, and in the pineal region in 1 case. The skull base MMs were located in the anterior skull base in 3 cases, in the petroclival region in 2 cases, in the tuberculum sellae in 1 case, in the sphenoid ridge in 1 case, and in the cerebellopontine angle in 1 case, respectively.

Table 2 Radiological characteristics of 69 intracranial microcystic meningiomas

Characteristic	No. (%)
Location	
Non-skull base	61 (88.4)
Convexity	32 (46.4)
Parasagittal	21 (30.4)
Falx	5 (7.2)
Tentorial	2 (2.9)
Pineal region	1 (1.4)
Skull base	8 (11.6)
Anterior skull base	3 (4.3)
Petroclival	2 (2.9)
Tuberculum sellae	1 (1.4)
Sphenoid ridge	1 (1.4)
Cerebellopontine angle	1 (1.4)
Side	
Left	33 (47.8)
Right	27 (39.1)
Bilateral/midline	9 (13.1)
Single or multiple tumor(s)	
Single	68 (98.6)
Multiple	1 (1.4)
Max tumor diameter (cm)	
Median (IQR)	4.4 (1.95)
Mean	4.6 ± 1.4
Range	2.2–8.0
CT findings*	
Density	
Hypodensity	23 (33.4)
Isodensity	6 (8.7)
Mixed	3 (4.3)
Hyperdensity	1 (1.4)
NA	36 (52.2)
Calcification	
Yes	0
No	33 (48.8)
NA	36 (52.2)
Bone destruction	
Yes	5 (7.2)
No	28 (40.6)
NA	36 (52.2)

NA data not available

*CT scan was available in 33 patients

The tumor was located on the left side in 33 cases and on the right side in 27 cases; it was bilateral in 9 cases. Multiple lesions were revealed in 1 case, whereas a single lesion was revealed in the other 68 cases. The mean maximal tumor diameter was 4.6 ± 1.4 cm (range, 2.2–8.0 cm). CT scans were available in 33 cases, and no calcification was observed.

Table 3 MRI findings of intracranial microcystic meningiomas

Characteristic	No. (%)
Solid/cystic growth pattern	
Solid lesion	50 (72.5)
Cystic-solid lesion	12 (17.4)
Cystic lesion	2 (2.9)
NA	5 (7.2)
T1-weighted imaging	
Hypointense	62 (89.9)
Isointense	2 (2.9)
Hyperintense	0
NA	5 (7.2)
T2-weighted imaging	
Hypointense	0
Isointense	0
Hyperintense	64 (92.8)
NA	5 (7.2)
The enhancement pattern	
Strong homogeneous enhancement	38 (55.1)
Heterogeneous enhancement	10 (14.5)
Faint reticular enhancement	7 (10.2)
Nodular enhancement	2 (2.9)
Other	7 (10.1)
NA	5 (7.2)
Tumor margin	
Smooth	34 (49.3)
Irregular	30 (43.5)
NA	5 (7.2)
Peritumoral brain edema	
No	11 (15.9)
Mild	16 (23.2)
Moderate	10 (14.5)
Severe	27 (39.2)
NA	5 (7.2)

MR image data was available in 64 patients

NA data not available

MRI data were available in 64 cases and summarized in Table 3. The tumor presented with solid in 50, cystic-solid in 12, and cystic in 2. The tumors (or solid component of cystic tumors) were either hypointense ($n = 62$) or isointense ($n = 2$) to gray matter on T1WI and hyperintense ($n = 64$) on T2WI. The tumor appeared with strong homogeneous enhancement in 38 cases, heterogenous enhancement in 10, reticular enhancement in 7, and nodular enhancement in 2.

Bone hyperplasia due to tumor invasion occurred in 5 cases. Regarding tumor margin, 34 tumors were smooth, and 30 were irregular.

PTBE was present in 53 patients (76.9%), as mild in 16 patients (23.2%), moderate in 10 patients (14.5%), and severe edema in 27 patients (39.2%).

Proposed classification and distribution of intracranial microcystic meningiomas

On the basis of the solid or cystic characteristic of growth and MRI finding, three types of MMs could be identified (Table 4).

Type 1 MMs presented as solid lesion, hypointense or isointense on T1WI, hyperintense on T2WI, and homogeneous or heterogeneous enhancement, and were found in 43 patients (67.2%) (Fig. 1). Type 2 MMs represented signals similar to CSF both on T1WI and T2WI, and faint reticular enhancement with marginal enhancement (Fig. 2), and these were found in 7 patients (10.9%). Type 3 MMs consisted of cystic-solid or cystic lesions and were found in 14 patients (21.9%) (Figs. 3 and 4).

Significant differences were observed among the different types of MMs for the following variables: sex, presence of severe PTBE, and extent of tumor resection (Table 5). All female patients were with type 2 MMs, but were only 35.7% of patients with type 3 MMs ($P = 0.018$). Severe PTBEs were more common among patients with type 1 MMs (55.8%) than among those with type 2 (14.3%) and type 3 MMs (14.3%) ($P = 0.007$). Type 1 MMs (97.7%) were associated with a significantly higher rate of GTR compared with the other two types (71.4 and 78.6%) ($P = 0.019$). There was a trend for type 1 MMs (mean, 4.3 ± 1.3 cm) to have smaller tumor size compared with type 2 (mean, 5.0 ± 1.4 cm) and type 3 (mean, 5.1 ± 1.4 cm) MMs ($P = 0.091$).

Table 4 Proposed classification and distribution of intracranial microcystic meningiomas

Classification	No. (%)	Solid/cystic growth pattern and/or MRI finding
Type 1	43 (67.2)	Solid lesion, hypointensity or isointensity on T1WI, hyperintensity on T2WI, and homogeneous/heterogeneous enhancement
Type 2	7 (10.9)	Signal equivalent to CSF both on T1WI and T2WI, and marginal enhancement with faint reticular enhancement
Type 3	14 (21.9)	Cystic-solid or cystic lesion

MR image data was available in 64 patients

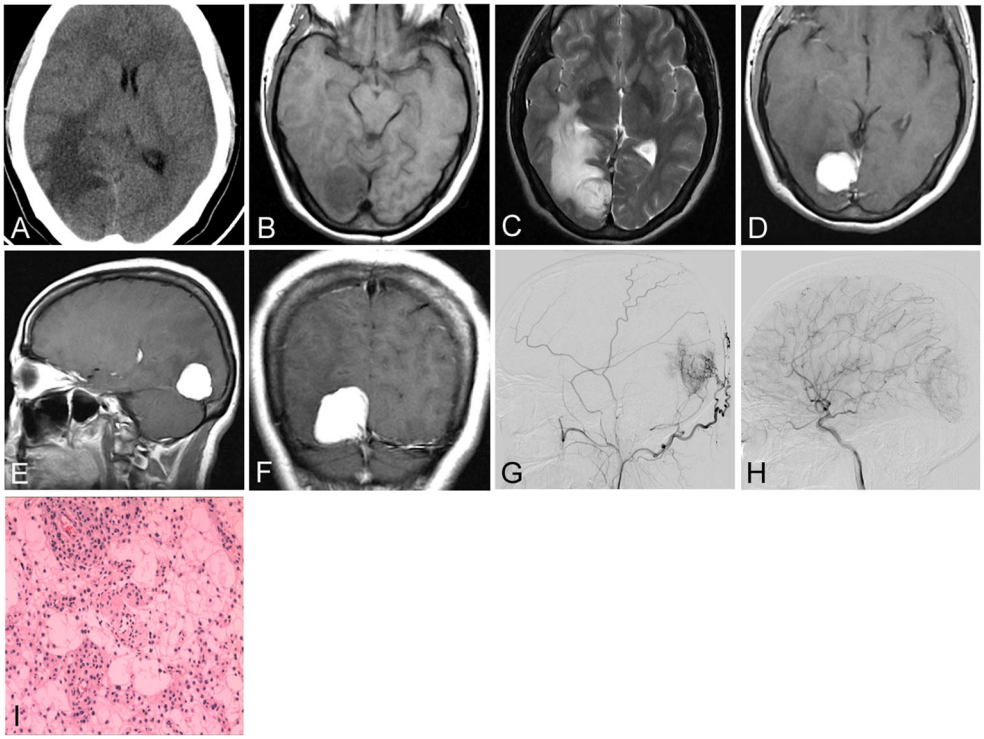

Fig. 1 Type 1 microcystic meningioma. Computed tomography **a** showed a slight low-density mass in the right occipital region with severe peritumoral brain edema. Magnetic resonance imaging revealed **b** hypointensity on T1WI, **c** hyperintensity on T2WI, and **d**–**f** strongly homogeneous enhancement. Cerebral angiogram showed **g** homogenous strong tumor stain on right external carotid artery angiogram and **h** faint stain on right internal carotid artery angiogram. **i** Pathological examination revealed the diagnosis of microcystic meningioma (H & E stain)

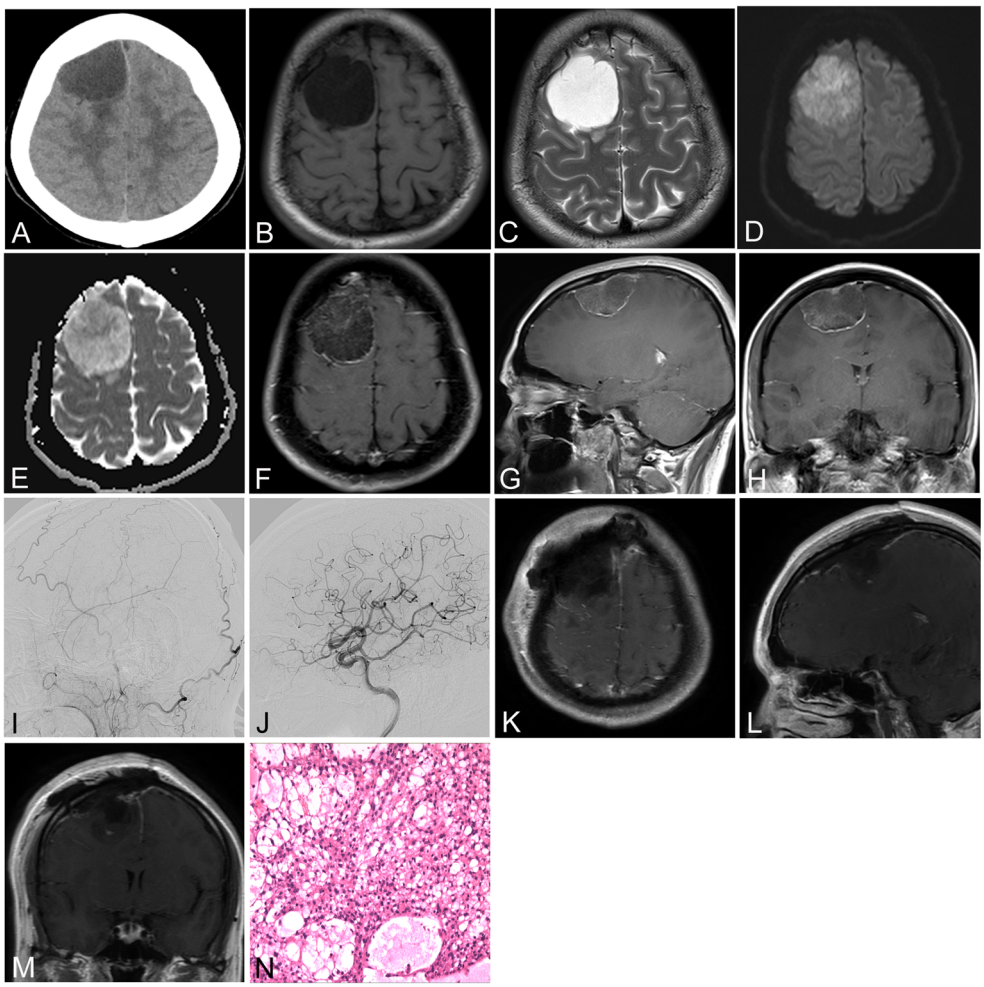

Fig. 2 Type 2 microcystic meningioma. Computed tomography **a** showing a hypodense mass in the right frontal region. The lesion showed **b** hypointense T1 and **c** hyperintense T2 signals equivalent to CSF. **d** Diffusion-weighted imaging and **e** apparent diffusion coefficient map showing local decreased diffusion. **f**–**h** Postcontrast T1WI showing marginal enhancement with faint reticular enhancement of the tumor. **i**, **j** Right external and internal carotid angiograms failed to reveal either tumor feeding arteries or tumor stain. **k**–**m** Postoperative MRI revealed gross total resection of the tumor. **n** Pathologic examination revealed the diagnosis of microcystic meningioma (H & E stain)

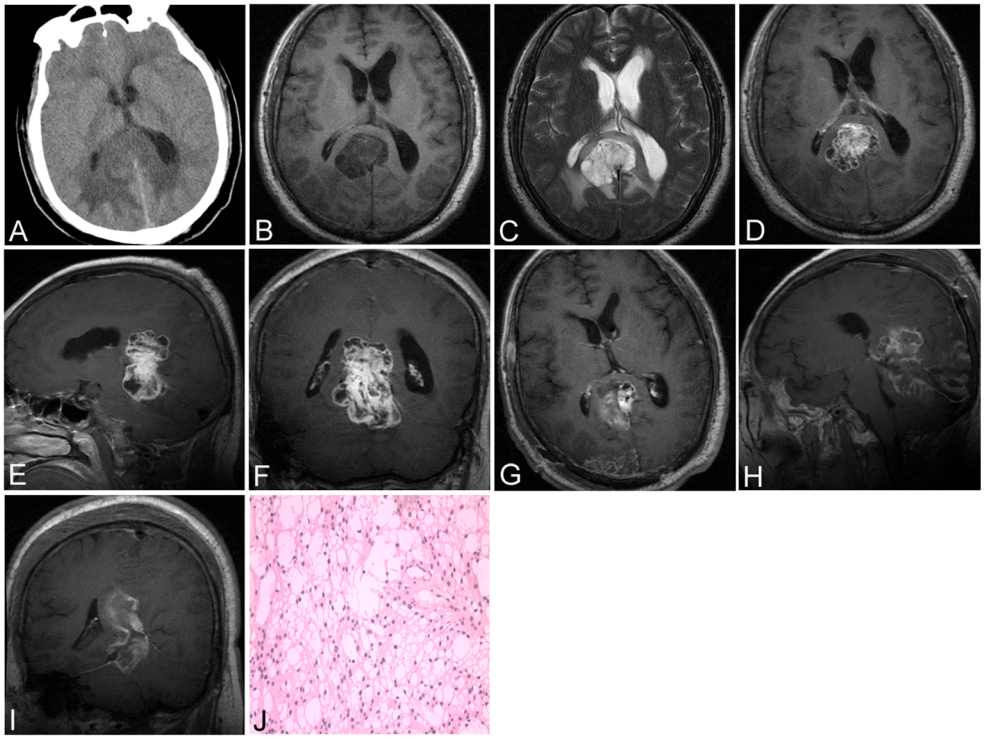

Fig. 3 Type 3 microcystic meningioma. Computed tomography **a** showing a hypodense mass in the splenium of the corpus callosum and superior vermis of cerebellum with peritumoral brain edema. Magnetic resonance imaging (MRI) revealing **b** hypointense T1 and **c** hyperintense T2 signals, as well as **d**–**f** heterogenous enhancement. **g**–**i** Postoperative MRI revealed subtotal resection of the tumor. **j** Pathological examination revealed the diagnosis of microcystic meningioma (H & E stain)

Surgical outcomes and follow-up

According to the Simpson grading system, patients treated by Simpson grade I, II, III, and IV resections were 47, 14, 2, and 6, respectively. Blood loss during surgery ranged from 100 to 8000 mL and the median loss was 300 mL. Total length of hospital stay after craniotomy ranged from 4 to 30 days (median, 8 days).

During the follow-up, four cases were missing. Sixty-five patients were evaluated clinically and by serial imaging (CT or MRI) for a median follow-up period of 49 months (range, 5 months–9 years). At the latest follow-up, KPS score showed

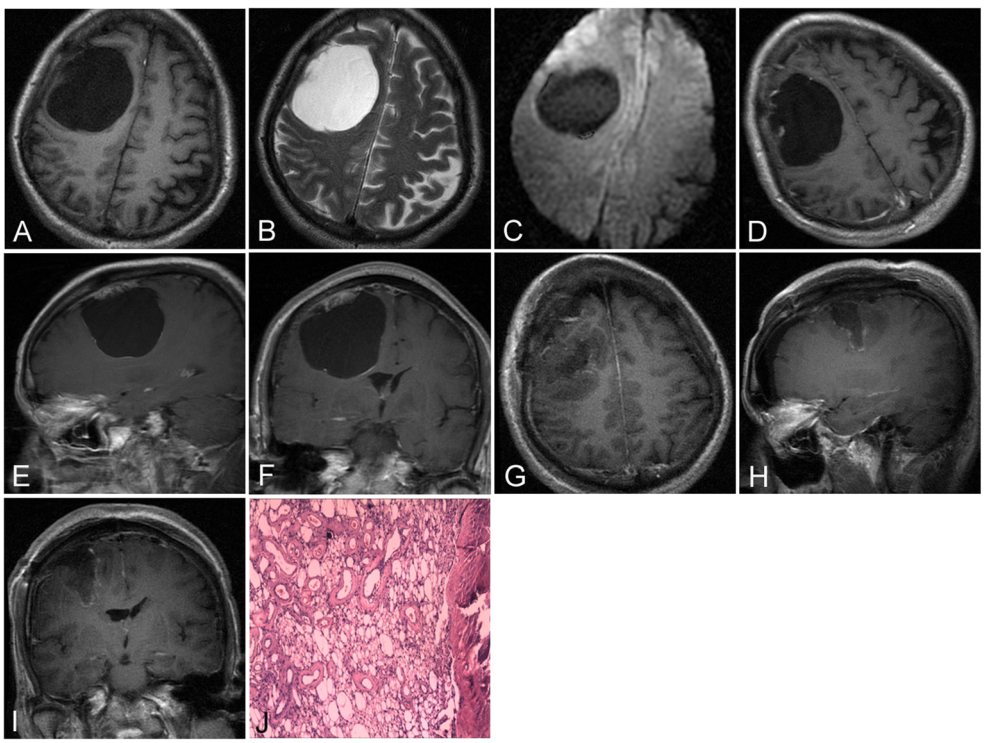

Fig. 4 Type 3 microcystic meningioma. **a**, **b**, **d**–**f** Magnetic resonance imaging (MRI) revealing a giant cystic mass with an enhancing mural nodule in the right frontal lobe. **c** Diffusion-weighted imaging showing increased diffusion of the cyst. **g**–**i** Postoperative MRI showed total resection of the tumor. **j** Pathologic examination revealed the diagnosis of microcystic meningioma (H & E stain)

Table 5 Clinical characteristics of intracranial microcystic meningiomas based on classification

	Type 1	Type 2	Type 3	*P* value	All patients
No.	43	7	14		64
Female sex	26 (60.5)	7 (100.0)	5 (35.7)	0.018	38 (59.4)
Mean age (years)	46.7 ± 10.7	51.9 ± 6.6	45.3 ± 10.4	0.451	47.0 ± 10.3
Seizure	11 (25.6)	1 (14.3)	4 (28.6)	0.767	16 (25.0)
Preop neurological deficit	16 (37.2)	2 (28.6)	5 (35.7)	0.907	23 (35.9)
Parasagittal	11 (25.6)	4 (57.1)	6 (42.9)	0.181	21 (32.8)
Skull base location	4 (9.3)	1 (14.3)	1 (7.1)	0.869	6 (9.4)
Mean tumor size (cm)	4.3 ± 1.3	5.0 ± 1.4	5.1 ± 1.4	0.091	4.6 ± 1.4
Peritumoral brain edema	36 (83.7)	6 (85.7)	11 (78.6)	0.885	53 (82.8)
Severe PTBE	24 (55.8)	1 (14.3)	2 (14.3)	0.007	27 (42.2)
Length of stay after craniotomy, days	9.2 ± 4.9	8.3 ± 1.9	8.2 ± 3.2	0.781	8.9 ± 4.3
Gross total resection	42 (97.7)	5 (71.4)	11 (78.6)	0.019	58 (90.6)
Patient with follow-up	42 (97.7)	6 (85.7)	13 (92.9)	0.412	61 (95.3)
Tumor recurrence	1 (2.4)	0	0	0.795	1 (1.6)

Classification was available in 64 patients. Values are presented as the number of patients (%) unless indicated otherwise. Mean values are presented as the mean ± SD

improvement in 63 cases and worsening in 2 cases, respectively. One patient with type 1 MM had developed tumor regrowth 4 years after surgery, gamma radiosurgery was performed, and the tumor remained stable in the latest follow-up of 7 years. The 3-, 5-, and 8-year PFS rates were 100.0, 98.5, and 98.5%, respectively. There were no significant differences in PFS among the three types of MMs (Fig. 5; $P = 0.788$). The median PFS for these three types MMs were both unavailable. The 3-, 5-, and 8-year PFS rates for type 1 MM were 100.0, 97.6, and 97.6%, respectively. The 3-, 5-, and 8-year PFS rates for type 2 and type 3 MM were 100.0, 100, and 100%, respectively. No patient died of recurrence. The median OS was also unavailable. There were no significant differences in OS among the three types of MMs.

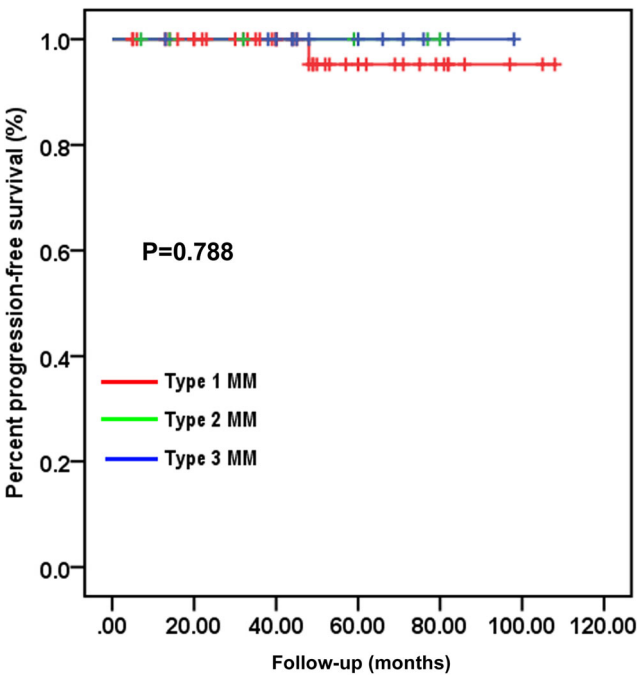

Fig. 5 Kaplan–Meier plots for PFS between type 1, type 2, and type 3 microcystic meningiomas were not significantly different ($P = 0.788$)

Discussion

Clinical characteristics of intracranial microcystic meningiomas

MM is a rare and benign subtype of intracranial meningioma. To date, the clinical features and radiological findings of MMs have been rarely addressed in the literature due to its extremely low incidence. A brief review of comparison of clinical features observed in different series is shown in Table 6. The proportion of MMs among the intracranial meningiomas in our study was approximately 0.63%. The female-to-male ratio was 1.46:1 in our series, which was consistent with that reported in other series but still lower than a ratio of approximately 2:1 among patients with intracranial meningiomas [29]. In addition, MM showed strong preference of supratentorial locations.

CT scan data were available in less than half of the cases, but over two thirds (23/33) of them presented as hypodense. Pathologically, MM can have intercellular, pale, and mucin-containing microcysts along with vacuolated, xanthomatous cells interspersed among hyalinized blood vessels [28]. Other typical features such as lobules, whorls, and psammoma bodies are absent [3, 7, 15]. Owing to the particular pathologic characteristics, radiologically, none of intracranial MMs showed calcification on CT, while all of them characteristically demonstrated distinct hyperintensity on T2WI in the present study.

Table 6 Comparison of clinical features observed in different series

	Kim et al. [12]	Paek et al. [25]	Kalani et al. [10]	Present study
Total no. of cases	11	16	25	69
Frequency	NA	1.75% (16/915)	NA	0.63% (69/11,000)
Age range (years)	37–67	38–71	24–76	21–75
Mean age	56.7	55.9	53.8	46.8
Female to male ratio	1.4:1	1.5:1	1.5:1	1.46:1
Location				
Supratentorial	100%	100%	56.0%	88.4%
Infratentorial and skull base	0	0	44.0%	11.6%
Mean tumor size	NA	34.8cm^3	NA	4.6 cm
Peritumoral brain edema	100% (11/11)	87.5% (14/16)	NA	82.8% (53/64)*
Severe PTBE	81.8% (9/11)	68.8% (11/16)	NA	42.2% (27/64)*
Reticular enhancement	36.4% (4/11)	12.5% (2/16)	NA	10.9% (7/64)*
No. of cases with available follow-up	11	NA	23	65
Mean follow-up period (range)	38.8 (7–79 months)	NA	101.7 (16 months–18.4 years)	49 (5 months–9 years)
Recurrences	0	NA	3	1

NA, data not available; *PTBE*, Peritumoral brain edema

*MR image data was available in 64 patients

Peritumoral brain edema and prolonged hospitalization

Intracranial MM has been reported to sometimes cause severe PTBE, despite the benign pathology [22, 31, 35]. Although the small series of no more than five cases in these reports might make the views lack powerful convincingness, this trend was confirmed in our study. Various PTBEs were seen in over 80% (53/64) of MMs in our series, and half (27/53) of them presented as severe PTBE. In the series by Paek et al. [25], PTBE was observed in 87.5% (14/16) of cases, and the majority of them (78.6%, 11/14) experienced a severe degree. In our cohort, the long duration of hospitalization after craniotomy (median, 8 days) was primarily driven through the occurrence of profound PTBE. Intracranial hypertension continued or increased despite surgical removal of tumor. These disproportional PTBEs frequently lead to severe medical and neurological complications in postoperative management. In addition, aggressive and prolonged antiedematous therapy was also necessary during the following days after surgery. These might result in an increase of the hospitalization time.

Histopathologically, MM is characterized by vacuolation, myxomatous change, and microcysts with formation of extracellular spaces containing edematous fluid, and suspected tumor cell hypersecretory activity [6]. Vascular endothelial growth factor (VEGF) immunoreactivity is higher in the endothelial cells of MM than in other common meningiomas. Prominently extensive PTBE is associated with disturbances of local vascular permeability [31], which are attributable in part to VEGF [6]. Secretion of other cerebral edema-related proteins, such as matrix metalloproteinase (MMP) or hormone-receptor, etc., may also be involved. Unfortunately, immunohistochemical data of MMP for most cases were unavailable. At present, most investigations have focused on meningiomas as a group, and the presence of MMP and steroid receptors in MMs have not been previously reported. This may be an interesting issue to be investigated in the future.

A proposed classification of intracranial microcystic meningiomas

In the present study, we identified three types of MMs. The solid or cystic growth pattern and radiological finding form the basis of the classification system which will be helpful for diagnostic certainty.

Type 1 MMs were found in nearly 70% of our patients with intracranial MM, and often showed severe PTBE. The majority of type 1 MMs appeared as extraaxial, solid, and homogeneously enhancing lesions with the dural tail sign. In addition, their findings suggestive of extraaxial localization included the presence of CSF cleft, broad base, and displacement and compression effect of the tumor on the brain parenchyma. Then, type 1 MMs can easily be distinguished from intraaxial tumors by virtue of their characteristic radiological findings.

Type 1 MM could be easily differentiated from intracranial psammomatous meningioma (PM). On T1WI, PM typically demonstrates isointensity with or without hypointense signal. On T2WI, the tumor commonly shows hypointense but may show isointense to hyperintense as well. CT reveals calcification diffusely or at the periphery of the tumor [16].

Type 2 MM showed a striking female predominance and accounted for approximately one tenth of patients in our study. Differential expression of hormone receptors or sex hormone in this subtype may partially contribute to this phenomenon. CT scan data were available in only three cases, but all of them showed obvious hypodensity. The MRI findings of type 2 MM are extremely characteristic. Tumors presented with signals similar to CSF both on T1WI and T2WI, and faint reticular enhancement. Authors suggested that this reticular enhancement was the characteristic signs highly suggestive of MM [5, 12, 14, 32]. This type of meningioma was most frequently reported in the previous literature. However, our study identified that this pattern was restricted to only minor cases which was classified as type 2 MM. Such MRI findings were not observed in non-microcystic meningiomas [2, 38]. The distinctive signal should be regarded as a suggestive and specific MRI sign of type 2 MM.

Type 3 MMs with atypical characteristics can be difficult to differentiate from malignant intraaxial tumors by conventional MRI. For example, in case 3 (see Fig. 3), the appearance on MRI, in particular, the lack of a typical CSF cleft or dural tail sign in addition to the cystic nature of the tissue enhancement and surrounding vasogenic brain edema, may lead to preoperative planning that presumes the tumor to be an intrinsic malignant high-grade glioma, rather than a benign extraaxial lesion. In case 4 (see Fig. 4), the tumor presented as an uncommon giant cystic lesion with an enhancing mural nodule. The differential diagnoses are wide, including hemangioblastoma, pilocytic astrocytoma, ganglioglioma, and pleomorphic xanthoastrocytoma. In these situations, radiological differentiation with conventional approaches is fairly difficult and may lead to false radiological reports. Hakyemez et al. [9] suggested that calculation of regional cerebral blood flow (rCBV) ratios via perfusion-weighted MRI and construction of signal intensity-time curves may contribute to the differentiation. The rCBV ratios of meningiomas were significantly higher than that of high-grade gliomas and metastases. However, dynamic contrast-enhanced perfusion-weighted MRI has some limitations. Calcium, hemorrhage, anterior skull base, and brain–bone–air interfaces may cause susceptibility artifacts [4]. Type 3 MMs should be differentiated from cystic atypical or malignant meningiomas. The appearance of lower apparent diffusion coefficient values were predictive of grade II–III meningiomas [1].

Peritumoral brain edema in different types of microcystic meningiomas

There was no significant difference with respect to the occurrence of PTBE between type 1, type 2, and type 3 MM. However, despite its smaller tumor size, type 1 MM tended to cause severe PTBE more often than did the other two types.

DSA data for a proportion of cases were available, and we found that the pial-cortical arteries were clearly involved in type 1 MMs (see Fig. 1g,h), while they were less involved in the blood supply to type 2 MMs (see Fig. 2i,j). Angiographic findings of MMs whose MRI showed dense enhancement demonstrated profuse dual blood supply from both pial and dural feeders [25]. The presence of vascular supply from the pial-cortical arteries in angiography is a significant association factor for severe PTBE [24, 35]. Increased rate of blood supply from the pial-cortical artery would lead to a higher risk to require blood transfusions [37]. The surgeon should take these facts into consideration.

Potentially resulting in significant midline shift and more severe postoperative complications, type 1 MM can pose a major challenge in clinical perioperative management and lead to prolonged hospitalization time. In the case of type 1 MM, early recognition of the extensive brain edema can be relevant for the clinical practice and should serve as an important indicator for potential perioperative hazards. Because of their long-proven efficacy in reducing vasogenic edema in brain tumors, the most widely used antiedematous agents in clinical practice are corticosteroids. Preoperative corticosteroid combined with dehydrant administration can be considered to reduce intraoperative swelling, and continuation of these medications is mandatory for the first few days postoperatively.

The previous study suggested that MM was known for hypervascularity [21]. However, according to clinical-radiological findings and classification, the present study notes that this theory is not quite right, and it should be refined. Hypervascularity may be more accordant with type 1 MMs. VEGF is the most potent conventional factor inducing PTBE in meningioma or other brain tumors [8, 11, 26]. Hypervascularity may be correlated with expression of VEGF in meningiomas [26]. The expression of VEGF for most cases was unavailable. Further studies are needed to clarify whether the expression of VEGF is varied among these three types of MMs.

Surgical outcomes of intracranial microcystic meningiomas

Our analysis demonstrates that patients with type 1 MM have significantly a higher GTR rate compared with the other two types of counterparts. Type 2 and type 3 MMs showed stronger preference of parasagittal locations than type 1 MM. Parasagittal meningiomas attached to and invading the superior sagittal sinus present a unique set of challenges. The treatment of these tumors poses the risk for catastrophic complications because of the intimate relationship between these tumors and the venous sinus with critical tributary bridging veins [27]. Radical resection of meningiomas involving major dural sinuses or bridging veins may be limited, in particular,

when these venous channels are patent. Postoperative long-term follow-up with radiological examination is proposed for type 2 and type 3 MMs with incomplete resection.

Despite classification as an established subtype of meningioma, large series with long-term follow-up of MMs are lacking. At a mean follow-up of 101.7 months (range, 16–221), the 3- and 5-year PFS rates for MM were reported to be 96 and 88%, respectively [10]. Due to the relatively short follow-up, we did not observe a statistical difference in the rates of tumor recurrence among the three types of MMs. Tumor recurrence was dependent on extent of tumor and dural resection. In addition, the rates of meningioma recurrence increased when the follow-up period was extended. Therefore, the proposed classification could still help to predict potential outcomes of tumor recurrence and progression.

Following surgical resection, there was no mortality related to surgery, and new temporary neurological deficits were limited to a small minority of cases [10]. MM showed strong preference of supratentorial locations. The operations were comparatively simple with a low risk of debilitating neurologic deficits. The consistency of MM may be another major indicator of surgical outcomes in addition to tumor location. Firm tumor consistency may present a risk primarily because of the increased need for sharp dissection and tumor manipulation during debulking. On T2WI, MM commonly shows hyperintensity. Hyperintensity on T2WI in meningiomas was advocated to be correlated with soft tumor consistency [34].

Recently, we reported surgical outcomes of 65 patients with intracranial PM, and the rate of GTR was 86.2% [17]. PMs had less extensive resections compared with MMs. This trend may also be explained by the tumor consistency. PM frequently shows hypointensity on T2WI. The stony tumor is laborious to remove and correlated with less favorable outcome of surgical resectability [34].

Study limitations

There were multiple limitations of this study. First, this was a retrospective review of a rare intracranial tumor. Typical biases existed, and the statistical analysis was limited given the small number of cases. Second, the available clinical follow-up period, ranging from 5 months to 9 years, was not adequate for the assessment of recurrence of benign meningiomas. Third, some immunohistochemical data, such as MMP, VEGF, and steroid receptors, for most cases were unavailable in our series.

Conclusions

We identified three types of intracranial MMs in this study. The solid or cystic growth pattern and radiological finding form the basis of the classification system which will be helpful for diagnostic certainty. Neurosurgeons should be aware of the characteristic features of this rare subtype. Preoperative attenuation of PTBEs is desirable to decrease intraoperative and postoperative complications, especially for type 1 MM. Type 2 and type 3 MMs predicted a less extensive resection, and close follow-up with radiological examination is proposed.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. For this retrospective study formal consent was not required.

Informed consent Informed consent was obtained from all individual participants included in the study.

Abbreviations *MM*, microcystic meningioma; *T1WI*, T1-weighted images; *T2WI*, T2-weighted images; *WHO*, World Health Organization; *MRI*, magnetic resonance image; *PTBE*, peritumoral brain edema; *EI*, edema index; *KPS*, Karnofsky performance scale; *GTR*, gross total resection; STR, subtotal resection; *VEGF*, vascular endothelial growth factor; *PFS*, progression-free survival; *OS*, overall survival; *PM*, psammomatous meningioma

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