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Clinical Study

# Long-term follow-up of surgical resection of microcystic meningiomas

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ABSTRACT

Microcystic meningioma is a rare tumor with myxoid and microcystic features. Our objective was to evaluate the efficacy of surgical resection of microcystic meningioma. Between December 1985 and October 2000 we treated 25 microcystic meningioma patients with surgical resection. We retrospectively analyzed the results including the long-term follow-up of this patient population. We identified 15 women and 10 men with a mean age of 53.8 years (24–76 years) who had microcystic meningiomas treated with surgery. Based on the Simpson grade, we found four Grade I (16%), 16 Grade II (64%), three Grade III (12%) and two Grade IV (8%) resections. The mean preoperative Karnofsky Performance Scale (KPS) score was 80.3 (range 60–100). The mean postoperative KPS score was 90.4 (range 60–100). At a mean follow-up of 101.7 months (range 16–221) the KPS score improved to a mean of 93.8. The recurrence/progression free survival (RFS/PFS) rates at 3 and 5 years were 96% and 88%, respectively. The 3 and 5 year RFS/PFS rates based on the Simpson grade were evaluated. The 3 year RFS/PFS rates for Grade I, II, III and IV were 100%, 100%, 66.6% and 100%, respectively. The 5 year RFS/PFS rates were 66.6%, 90%, 66.6% and 100%, respectively. Microcystic meningioma is a rare tumor, which is characterized by extracellular microcystic spaces filled by edematous fluid and peritumoral edema. Following surgical resection these tumors have a positive prognosis with a benign course. The surgical outcomes seem to be associated with the risks related to the surgical procedure.



## 1. Introduction

Meningiomas are the most common benign intracranial neoplasm constituting approximately 20% of all primary brain tumors [1–3]. Incidence increases progressively with age and usually middle-aged and elderly persons are more affected. Meningioma incidence is much higher in women than men and has been reported as high as 38% for intracranial tumors overall [1–3]. Meningiomas have been classified into three different grades by the World Health Organization (WHO). The vast majority of tumors (92%) are benign Grade I meningiomas whereas only 8% are atypical Grade II or anaplastic Grade III meningiomas [4,5]. Grade I meningiomas present nine subtypes of different histological patterns including microcystic meningioma [6], a rare histological variant presenting remarkable morphological features such as microcysts and a myxoid appearance (Fig. 1). Our study focuses on a long-term follow-up of patients who underwent surgical resection of this subclass of tumor.

## 2. Methods

Patient data obtained between December 1985 and October 2000 were retrospectively analyzed. Clinical and operative records of 25 consecutive patients undergoing surgical resection for histologically diagnosed microcystic meningioma were reviewed. Each patient's clinical status was assessed using the Karnofsky Performance Scale (KPS) [7] during the neurological examination at admission.

## 3. Results

In total we identified 25 patients (15 women and 10 men) with a diagnosis of microcystic meningioma. The mean age of the patients

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was 53.8 years (range 24–76). Patient characteristics and tumor location are described in Table 1. Twenty-three out of 25 patients had only a single meningioma whereas two patients had multiple meningiomas. One patient had also previously had a meningioma.

Symptom duration varied at presentation with a mean of 11.8 months (range 1–72). The most common presentations in our study population were motor seizures in seven patients (with one patient developing progressive epilepsy). Three patients presented with contralateral weakness due to prerolandic cortex involvement, four with speech disturbances and aphasia because of the location of the tumor in an eloquent area (Broca's and Wernicke's areas), four with visual impairment with diplopia or visual field decline, two with lethargy and cognitive decline due to frontal lobe location of the lesion, and two with memory difficulties.

The preoperative KPS score was measured on admission with a mean of 80.3 (range 60–100). Deficits and neurological symptoms on admission typically reflected the location of the tumors. Regardless of the tumor location, the most common symptoms were headache (n = 8), seizures (n = 6) and confusion (n = 4). Most symptoms improved after surgery. Common location-related symptoms were weakness (n = 3), hemiparesis and tremors (n = 3) and cognitive decline (n = 2).

Among the patients included in the study population there were significant differences in the extent of surgical resection (Fig. 2). The extent of surgical resection was evaluated according to the Simpson grade. In our cohort, four resections were Grade I, 16 were Grade II, three were Grade III and two were Grade IV. No resections were classified as Grade V. The mean Simpson grade scale for these patients was 2.12. Blood loss was a mean of 450 mL (range 100–1200), one patient required a blood transfusion during surgical resection. The mean hospitalization period was 10.6 days, and all patients were discharged.

The mean postoperative KPS score was 90.4. Preexisting symptoms worsened in one patient with increased dysphagia, concomitant cerebrospinal fluid (CSF) leakage and tetraparesis that improved on follow-up. Temporary new neurological deficits were reported in six patients.

At a mean follow-up of 101.7 months (range 16–221) the KPS score improved to a mean of 93.8. In two patients, follow-up data beyond the postoperative period were not available. Two patients died during the follow-up period for reasons unrelated to their tumors. Two patients who had grade IV resections required postoperative radiation.

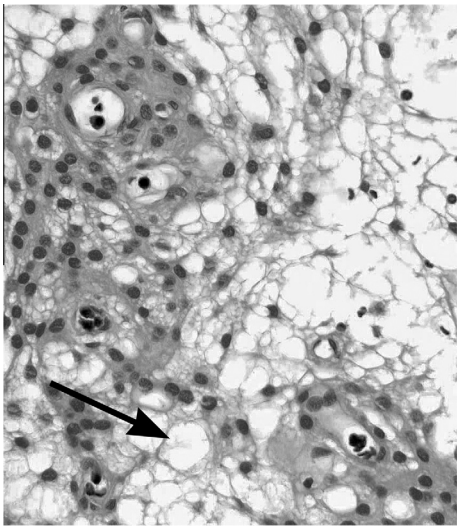

**Fig. 1.** Hematoxylin and eosin stained sections of the tumor reveal the microcystic features (arrow) of this lesion. *Used with permission from Barrow Neurological Institute.*

Three patients developed recurrence or progression of their tumor. One patient with a Grade II resection and one with a Grade III resection exhibited disease progression and one with a Grade I resection had recurrence of his tumor. The recurrence/progression free survival (RFS/PFS) rates at 3 and 5 years were 96% and 88%, respectively. The relationship between RFS/PFS and the Simpson grade was investigated. Because the long-term follow-up was extremely variable in our cohort, we evaluated the RFS/PFS of only those patients who underwent clinical and radiological evaluation at 3 and 5 years, respectively. We did not evaluate those with a shorter follow-up. The 3 year RFS/PFS rates were 100%, 100%, 66.6%, and 100%, respectively. The 5-year RFS/PFS rates for Grades I, II, III and IV resections were 66.6%, 90%, 66.6%, and 100%, respectively.

## 4. Discussion

Microcystic meningioma is a rare WHO Grade I subtype of meningioma characterized by a myxoid appearance and the presence of microcysts. These tumors constitute approximately 1.6% of all intracranial meningiomas [8–19]. If cases are included in which an association between microcysts and meningioma has been reported, the incidence remains only 4–7% of all cases of meningiomas [20,21].

Masson was the first to report this tumor as a unique histological entity [19]. Subsequently, Penfield suggested a mechanism of microcyst formation in these tumors [22]. In 1980, Kleinman et al. proposed the name "microcystic meningioma" to describe this subset of tumors [23]. In 1993, this histological subtype was acknowledged by the WHO and included in the classification of brain tumors [15]. Despite classification as an established meningioma subtype, large data series with long-term follow-up of microcystic meningiomas are lacking.

This tumor variant has a slight female preponderance reflecting the sex incidence of the common benign meningiomas [24]. Some series suggest that this variant may be more common in children than adults, but others report this variant to be exceedingly rare in children [25–28]. Our patients had a mean age of 53.1 years and the youngest was 24 years old. The scarcity of children in this series likely represents the referral pattern to our hospital which mainly treats adult patients. Other findings reported in some papers indicate the cerebral convexity as the most common location for microcystic meningioma followed by the parasagittal region [29,30]. When these studies are considered together these locations accounted for 56% of all patients.

Several hypotheses have been proposed concerning the pathogenesis of this type of tumor. Masson proposed that CSF transudation causes imbibition of the tumoral tissue [19]. Michaud and Gagne emphasized the potential movement of low-protein fluid from fenestrated vessels of the vascular network of the lesion [31]. Even vascular endothelial growth factor has been suggested to play a role in the pathogenesis of the microcysts by Christov et al. [32]. However, these theories have not been substantiated by clinical or molecular evidence [33,34]. Other researchers have postulated that an imbalance between matrix metalloproteinases and tissue inhibitors of matrix metalloproteinase play a role in microcysts formation and peritumoral edema [34]. Another hypothesis derives from the observation of gap-junctional features in the ultrastructure of the microcystic meningioma. They have also been thought to occur as a result of abnormal tumor cell differentiation, sharing some similarities with arachnoid cells and their physiological CSF regulation, and to be responsible for the accumulation of fluid in the microcystic pattern [35]. In summary, the mechanism of microcyst formation has not yet been clearly elucidated but it is supposed to be of arachnoid trabecular cell