Case Report

Rare Morphological Variant of Meningioma: A Case Report of Microcystic Meningioma

Muhammad Z. Arifin, Roland Sidabutar, Rachmanda H. Wibisono and Ahmad Faried*

Department of Neurosurgery, Universitas Padjadjaran–Dr. Hasan Sadikin Hospital, Indonesia

Abstract

A rare morphological variant of meningioma, known as microcystic meningioma, was originally described as “humid” or vacuolated meningioma. It is difficult to distinguish this clinical entity through computed tomographic scans or magnetic resonance images, as they appear similar to glial or metastatic tumor with cystic or necrotic areas. Their similar imaging characteristics make it difficult to differentiate microcystic meningiomas from these more common pathologies. We describe the case of a 33 years old female who presented with generalized seizures, and was found to have a mass of the left parieto occipital region. Preoperative diagnosis based on imaging studies was pilocytic astrocytoma. A craniotomy tumor removal was performed, and tumor was excised. Microscopic and histopathological results established a diagnosis of microcystic meningioma. Cystic meningiomas are difficult to diagnose accurately using pre-operative imaging. Our case is unique because it presented as a rare morphological variant of meningioma that is extremely rare in our institution. Its unusual imaging might lead to confusion between extra- and intra-axial tumors. In our case, a definitive diagnosis was only possible using a histopathological examination.

INTRODUCTION

Meningiomas are common tumors of the central nervous system that are generally slow growing, non-infiltrating and benign lesions, and they account for 13-26% of intracranial neoplasms [1,2]. Meningiomas are extra-axial neoplasms [3] (i.e., they arise from arachnoid cap cells, not the brain or spinal cord) that are generally solid lesions. Pathognomonic findings on Computed Tomography (CT) scans and Magnetic Resonance Imaging (MRI) are usually sufficient to establish a correct diagnosis. The WHO classifies meningiomas into several subtypes based on histological parameters, which are meningotheliomatous meningiomas, fibrous meningiomas, transitional meningiomas, psammomatous meningiomas, angiomatous meningiomas and others [4]. Microcystic meningiomas are rare morphological variants with microcystic formation.

Microcystic meningiomas were separately classified from other meningiomas [5], with the term “microcystic” coined by Kleinman et al. in 1980 [6]. These tumors are characterized by finding numerous large formations of extracellular microcystic spaces, containing edematous fluid, creating a stellate, vacuolated cellular appearance of with occasional large hyperchromatic and pleomorphic nuclei. Its unusual microscopic findings might be mistaken for hemangioblastomas, astrocytomas, schwannomas, chordomas, and angioblastic meningiomas. However, the clinical features and prognosis of the tumor do not differ from the usual benign meningiomas [7]. The presence of an associated cyst is an uncommon imaging feature that may make it difficult to distinguish the tumor from a primary intra-axial glial neoplasm. The presence of peritumoral edema can also be a misleading finding.

CASE PRESENTATION

We present the case of a 33-year-old woman who suffered from brief episodes of seizures and loss of consciousness over a period of three months. She also complained of vision deterioration in her left eye over the past 4 years that was associated with progressive intermittent headaches in the past 5 years. Neurological findings give a visual acuity of 1/60 in the right eye and 1/300 in the left eye, and funduscopic examination shows early papillary edema of both eyes. Other cranial nerves were within normal limits, with no motoric or sensoric deficits. Laboratory examinations showed normal findings. CT scan images show an iso hypodense lesion of the left parieto occipital region that enhanced in homogeneously with contrast, with peritumoral edema and compressed sulcus and gyrus (Figure...
1) MRI examination shows extension of the tumor, which has an iso hypointense appearance on T1-weighted images, peritumoral cystic (+), ring enhancement (+), mural nodule (+), without dural tail signs after gadolinium administration, and an iso hyperintense with peritumoral edema (+) in T2-weighted images, with a small solid component in a predominantly cystic lesion (Figure 2). Preoperative working diagnosis of pilocytic astrocytoma was made. A craniotomy tumor removal from a prone position was performed. Intraoperatively, the tumor contained xanthochromic fluid in its cystic component. The mass was reddish, soft, spongy, easily aspirated, moderately vascular and measured 5 x 4.5 x 4 cm. Adequate tumor decompression with excision of the tumor capsule was performed, and the solid component was biopsied. The tumor and cyst wall were completely removed. The resected tumor was well encapsulated by a thin fibrous capsule with focal areas of fibrous adhesions. The cut surface was a yellow, light brown color and slimy (Figure 3). Hemorrhage and necrosis were absent. The final histopathological report revealed microcystic meningioma (Figure 4).

**DISCUSSION**

Microcystic meningiomas are known to be unusual morphological variation of meningiomas. These tumors were originally described by Masson who labeled them “humid” or vacuolated meningiomas because of their gross appearance [5]. Similar lesions have been subsequently reported under various designations, such as humid and myxomatous [8], vacuolated [9], and microcystic [7]. They were established as a separate subgroup of meningiomas by the WHO classification of central nervous system tumors [1]. Sporadic case reports of microcystic meningiomas that include radiological findings are present in the literature [10,11].

The pathogenesis of these tumors is unclear, although transudation of low-power protein fluids was previously suggested [5]. Histochemical stains in our case indicated that the extracellular fluid was neither mucin nor glycogen. The age of affected patients ranges are 3 years old to 74 years old, with slightly more female predominance. Most lesions manifest the cerebral convexities or parasagittal region [6,12]. Grossly, microcystic meningiomas are well delimited with a smooth bosselated or lobulated external surface. The texture is generally soft and spongy. The cut surface discloses a yellowish, light brown homogenous color with a glistening appearance. Hemorrhages and necrosis are never found [5,13,14]. The clinical behavior of these tumors is not different from other benign meningiomas. Microscopically, there are numerous large formations of extracellular microcystic spaces, containing edematous fluid, creating a stellate, vacuolated cellular appearance of with occasional large hyperchromatic and pleomorphic nuclei. Some areas show conglomeration of the smaller cystic spaces, which creates a vacuolated, myxoid and loosely reticular appearance [5,6,15]. Foci of nuclear pleomorphisms are occasionally noted,

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**Figure 1** Non-contrast axial-computed tomography (CT) scan showed an iso hypodense lesion in the left parieto-occipital region (A) that enhanced in homogeneously with contrast (B), peritumoral edema (+) as a marker of vasogenic edema and compressed sulcus and gyrus.
but this finding does not indicate aggressive behavior [6,14]. Cystic meningiomas occur more in the pediatric patients than in adults. They occur in 10-19% of all pediatric meningiomas compared to only 2-4% in adults [16,17]. Rengachary et al., [18] classified two types of cysts, which are intra-tumoral and peritumoral, distinguished by the cyst was lined with meningothelial cells. Nauta et al., [19] also classified several types of cysts in meningiomas: (1) central intra-tumoral cyst, (2) peripheral intra-tumoral cyst, (3) peritumoral cyst of the adjacent brain tissue, (4) peritumoral cyst between the tumor and adjacent brain tissue. The presence of peritumoral edema, as in the case presented, can also be a misleading finding. There are four main mechanisms thought to be responsible for edema formation: mechanical compression, disturbed blood-brain

Figure 2 Axial magnetic resonance imaging (MRI) demonstrated an iso hypointense appearance on T1-weighted images (A), peritumoral cystic (+), ring enhancement (+), mural nodule (+) with gadolinium administration (B), iso hyperintense with peritumoral edema (+) on T2-weighted images, predominantly cystic with a small solid component and FLAIR images.

Figure 3 Resected tumor was well encapsulated by a thin fibrous capsule with focal areas of fibrous adhesions. The cut surface was a yellow, light brown color and slimy.
barrier, edemagenic factor secretion and vascular compression [20]. Peritumoral edema in meningiomas is not related to the overall malignancy, in contrast to gliomas. It is difficult to preoperatively diagnose cystic meningiomas, as its radiological findings are similar to glial or metastatic tumors with cystic or necrotic areas and contrast-enhancing tumor nodules.

CONCLUSION

Cystic meningiomas are unusual and distinct morphological variant of meningiomas which are difficult to diagnose accurately, as they have similar clinical and histological appearance as a normal meningioma. Its unusual imaging characteristics might lead to confusion between extra- or intra-axial tumors. In our case, a definitive diagnosis was only possible using a histopathological examination.

CONSENT

Informed consent was obtained from the patient for publication of this case report and any accompanying images. His family was present at the time.

AUTHORS’ CONTRIBUTIONS

MZA, RS, RHW, and AF had examined, treated, observed, and followed up the subject of this research. RS, RHW and AF performed the operation on the patient. All authors participated in writing the manuscript. All authors have read and approved of the final manuscript.

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REFERENCES


