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Case report

Cystic meningioangiomatosis with enhancing mural nodule on MRI and no calcification on CT



AND NEUROSURGERY

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ABSTRACT

The neuroradiological features of meningioangiomatosis (MA) are non-specific. We report a young man with sporadic MA. The plain computerized tomography (CT) demonstrated a deep located right cystic lesion without calcification. On magnetic resonance imaging, the cystic mass lesion was confirmed with a mural nodule with significant enhancement on contrast-enhanced images.

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Clinical details

A 23-year-old man presented with 2-years paroxysmal numbness of the left limb and slow aggravation for 1 year. Each episode lasted for 40–50 s. Over the past 2 years, seizure frequency gradually increased and symptoms got worse. On physical examination, the patient manifested clear language, normal binocular vision, and no optic papilla edema or stiff neck. Lower limb activity was found to be associated with grade V muscle strength, normal muscle tension, positive tendon reflexes, and no pathological neurologic signs. He had discrete hypoesthesia of the left limb, which presented reduced two-point discrimination capacity and negative Romberg's sign. Routine blood examination revealed a higher lymphocyte percentage (42.5%; (reference value = 20–40%), lower values of hematocrit (41.5%; reference value = 42–49%) and total cholesterol (3.31 mM; reference value = 3.57–5.1 mM), and a higher anion gap (22.12 mM; reference value = 4–20 mM).

Plain computerized tomography (CT) (Fig. 1) demonstrated a deep located right cystic lesion of almost 5 cm with an average CT value of 8.7 HU. Magnetic resonance imaging (MRI) confirmed the cystic mass, which presented an isointense mural nodule on T_1WI , slightly hyperintense on T_2WI and diffusion-weighted images (DWI), with significant enhancement on contrast-enhanced T_1WI (Fig. 2).

At surgery, the cystic tumor was probed through the insular lobe, and about 40–50 mL yellowish transparent fluid was aspirated. An 8 mm aubergine colored tumor nodule was

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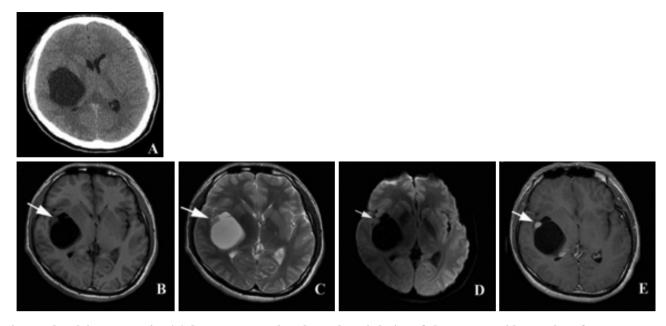


Fig. 1 – The plain CT scanning (A) demonstrates a deep located cystic lesion of almost 5 cm with CT value of 8.7 HU. MRI confirms the cystic nature of the lesion on T_1WI (B), T_2WI (C), DWI (D), and contrast-enhanced T_1WI (E), and with a mural nodule that enhances significantly after using gadolinium (arrows).

found on the capsule wall at the insular side. The tumor nodule and surrounding adhesion tissue were removed.

The pathological diagnosis (Fig. 3) after surgery confirmed the meningioangiomatosis (MA). The CT and MRI follow-up up to 3 months after surgery confirmed the complete removal of the cystic mass.

Discussion

MA is a rare, benign focal disorder that usually affects the leptomeninges and the cerebral cortex [1]. There are only 136

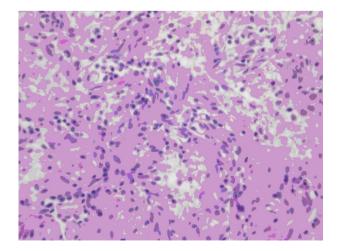


Fig. 2 – The pathological image (Hematoxylin and Eosin staining at $400 \times$ magnification) shows that the hyperplasia of leptomeninges epithelial cells and meningeal vascular proliferation are surrounded with fibroblasts proliferation of spindle cells in a swirl or sarciniform pattern.

cases reported in the literature at present. Most sporadic MA involved children and young adults. Our patient was 21 years old when he first had symptoms. About 90% of MA affects the cerebral cortex of frontal and temporal lobes, and only occasionally involves deep locations, such as the third ventricle, thalamus, cerebral peduncle, and brainstem [1].

Preoperative accurate diagnosis of MA is crucial. However, both clinical and neuroradiological features of MA are nonspecific. The imaging findings of MA can be diverse on CT and MRI because of different morphological features. CT shows different characteristics from hypodense, isodense, or slightly hyperdense round masses surrounded by areas of low density and with or without calcifications [2]. MA manifestations on MRI are various, which can be isointensity or hypointensity on T₁WI, and a heterogeneous cortical mass surrounded by an area of increased intensity on T₂WI, probably due to either edema or gliosis [2]. Contrast enhancement is variable.

Our patient had a well-defined cyst on plain CT, without calcification. On MRI, the cystic lesion associated with a mural nodule which manifesting isointense on T_1WI , slightly hyperintense on T_2WI and DWI, with significant enhancement on T_1WI . Only 6 cystic MA cases have been previous published, but none were associated with an enhancing mural nodule [3]. Therefore, our case is the first cystic MA with an enhancing mural nodule and without calcifications.

The differential diagnosis of MA includes vascular malformations, cystic meningioma, hemangioblastoma, pilocytic astrocytoma, pleomorphic xanthoastrocytoma, ganglioglioma, and cystic brain metastases [2]. The cystic meningioma is quite rare, accounting for 2–4% of intracranial meningiomas, which is more common in childhood. Hemangioblastomas are preferentially located in posterior fossa and rarely in supratentorial region, and typically manifest as a well-defined cyst associated with a solid enhancing mural nodule. Pilocytic astrocytomas occur more often in children and young adults, and usually manifests as a well-circumscribed macrocyst lesion associated with an enhancing mural nodule. About 50–60% of pleomorphic xanthoastrocytomas present as a cystic component with an enhancing mural nodule. Half of gangliogliomas present as a cystic mass with a mural nodule, mostly located in the temporal lobe. Cystic metastases in the brain are relatively rare. Some breast ductal carcinomas and lung adenocarcinomas may be more frequently associated with metastatic cyst formation.

In conclusion, our case demonstrates that MA can present as a cystic lesion with an enhancing mural nodule. Although clinical and imaging findings are non-specific, MA should be included in the differential diagnosis of such brain lesions.

Conflict of interest

None declared.

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None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical

Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

Authors' contribution

Y.F. carried out the molecular genetic studies, participated in the sequence alignment and drafted the manuscript. Z.H.S. carried out the immunoassays. J.Z. participated in the sequence alignment. H.T.L. participated in the design of the study and performed the statistical analysis. Z.H.S. conceived of the study, and participated in its design and coordination and helped to draft the manuscript. All authors read and approved the final manuscript.

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