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Case Report

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Giant Cavernous Malformation : A Case Report and Review of the Literature

Giant cavernous malformations (GCMs) occur very rarely and little has been reported about their clinical characteristics. The authors present a case of a 20-year-old woman with a GCM. She was referred due to two episodes of generalized seizure. Computed tomography and magnetic resonance image demonstrated a heterogeneous multi-cystic lesion of 7×5×5 cm size in the left frontal lobe and basal ganglia, and enhancing vascular structure abutting medial portion of the mass. These fingings suggested a diagnosis of GCM accompanying venous angioma. After left frontal craniotomy, transcortical approach was done. Total removal was accomplished and the postoperative course was uneventful. GCMs do not seem differ clinically, surgically or histopathologically from small cavernous angiomas, but imaging appearance of GCMs may be variable. The clinical, radiological feature and management of GCMs are described based on pertinent literature review.

KEY WORDS : Cavernous hemangioma · Venous angioma · Seizure.

INTRODUCTION

The cavernous malformation (CM), also known as cavernous angioma or cavernoma, is a vascular malformation characterized by the presence of sinusoid-like capillary vessel containing blood in very sluggish circulation[5].

CMs vary in size from a few millimeters to a few centimeters. Few data can be found about the size of these malformations. Kim et al.[12] reported the size of cavernoma between 1mm and 75 mm, with a mean size 14.2 mm. The majority of cavernous malformations are small but it may reach significant size. Unlike giant aneurysms, defined as having a diameter of at least 25 mm, no threshold dimension has been accepted for giant cavernous malformation (GCM). Lawton et al.[15], although arbitrary, defined a GCM as a cavernoma with a diamter greater than 6 cm. GCMs are very rare and usually not considered in the differential diagnosis of large tumor. Recently we experienced a case of cerebral giant cavernous malformation with a diameter of 7 cm. In this report, the clinical, radiological feature and the surgical management and prognosis of this vascular malformation are described.

CASE REPORT

A 20-year-old, right-handed woman was refered due to two episodes of generalized seizure. She had no significant past and family history. There wereno neurological deficits on admission. Computed tomography (CT) scan revealed a 7×5×5 cm sized mixed density lesion with multifocal calcification in the left frontal and basal ganglia region. On contrast enhanced CT scan, the mass showed heterogeneous enhancement and traversing vascular structure was found in posteromedial portion of the mass. The mass was multicystic and mixed intensities in magnetic resonacne (MR) image and surrounded by a low signal intensity rim on T2-weighted images, representing hemosiderin. On enhanced MR images, there was venous angioma abutting medial portion of the mass (Fig. 1). Based on CT and MR finding, we diagnosed this lesion as GCM with venous angioma.

After left frontal craniotomy, transcortical approach was done. The mass appeared brownish, and had multiple cysts, "caverns". Each cavern had brownish liquified blood product. Cysts were coagulated and the contents were removed by suction. The margins were distinct, allowing the lesion to be peeled away from the surrounding tissue by repeated coagulation and piecemeal removal. There was no significant bleeding. The venous angioma was found in posteromedial portion of tumor and left untouched. After the operation the patient made a

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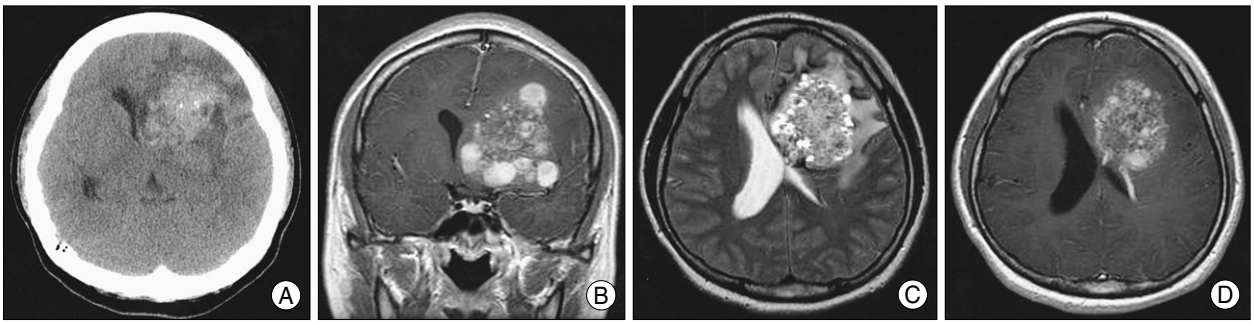

Fig. 1. A : Non-enhanced computed tomography (CT) scanning showing a 7×5×5 cm sized mixed density lesion with multifocal calcification in the left frontal and basal ganglia region. B, C : The mass was multicystic and mixed intensities on gadolinium-enhanced coronal magnetic resonance (MR) image (B) and surrounded by a low signal intensity rim on T2-weighted axial images (C), representing hemosiderin. D : On enhanced axial MR images, there was venous angioma abutting medial portion of the mass.

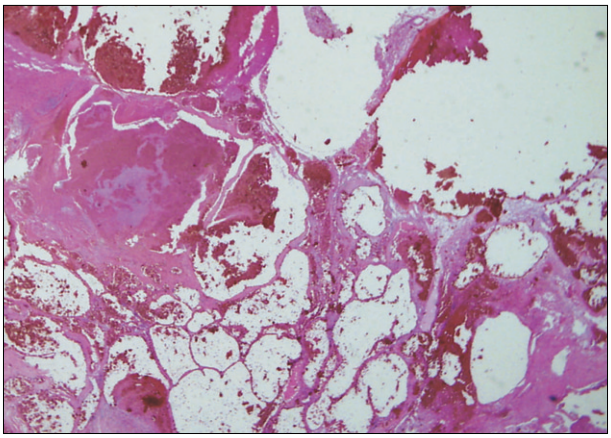

Fig. 2. Low-power photomicrographs showing various thrombosis and calcifications within the cavernous vascular spaces. Also noted are thin-walled vascular channels with little intervening brain (H&E, ×100).

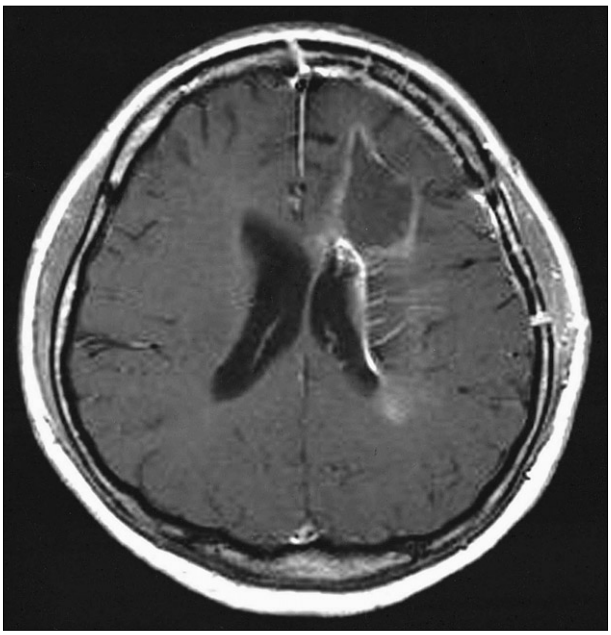

Fig. 3. Postoperative gadolinium-enhanced axial magnetic resonance image showed no residual cavernous malformation with venous angioma being intact.

fast recovery without any neurological deficits. Histological examination revealed a CM (Fig. 2). Follow-up MRI showed no residual lesion (Fig. 3).

DISCUSSION

CMs account for approximately 5 to 10% of all vascular malformations[19,20,22,25]. GCMs are very rare and little has been reported about their clinical characteristics[7,14,23,24]. But, the definition of GCMs is arbitrary. We defined GCMs as cavernoma with a diameter greater than 6 cm in accordance with Lawton et al. With this definition, we found that only 14 cases have been repoted in the literature[7,14,23,24].

Growth of CMs

Cerebral CMs rarely attain large dimension. The mechanism by which they enlarge is probably recurrent bleeding, followed by organization of the clot, pseudocapsule formation, and secondary expansion[3]. However, it was also reported that CMs can show expansile growth without any evidence of a hemorrhagic event and mimic neoplasm[21].

Clinical features

Although patients with CMs typically present between the second and fourth decades[1,5,12,22,25], the majority of GCMs has occurred in children, with youngest one being 3.5 months of age[3,4,8-11,24]. The overall prevalence among males and females is equal in the majority of CMs[5,20], but, in GCMs, there seem to be a female preponderance[24]. Familial CMs account for 20% to 50% of patients presenting with CMs[17,19]. In review of GCMs , no familial occurrence has been reported[24]. Multiple CMs may occur in 10% to 30% of sporadic cases and in up to 84% of familial cases[6,26], but it was not reported in any of GCMs[24].

The usual symptoms of a cavernoma are seizure, progressive neurologic deficit, hemorrhage[5,20]. Usually, the presentation of the GCMs is not different from that of usual CMs[7,24].

Our case also presented with seizure. But, presenting a large intracranial mass with signs of increased intracranial pressure in children was reported in some cases[4,8,10,11,24]. Hemorrhage of the CMs is reported to be 8% to 37% in adults and 36% to 78% in children[16,17]. However, true hemorrhage occurrence is relatively rare in GCMs[3,15,24].

Neuroimaging

Diagnosis is mostly straightforward in typical cases of cavernous malformation. CMs usually have little or no surrounding edema nor mass effect[7,18]. CMs may rarely be in the form of a cystic growth with a well-defined capsule[18]. On the other hand, diagnosis may be challenging in GCMs, which are rare lesions[7]. Imaging appearance of GCM is variable, ranging from completely cystic lesion[4,11] to those resembling neoplasms with striking contrast enhancement and mass effect[4,24], and finally to heterogenous lesion with peripheral hemosiderin rim and without significant contrast enhancement and mass effect[2,7]. Therfore, despite its varity, the possibility of CMs should be considered in the case of large intracranial tumor. In our case, preoperative diagnosis of GCM was made without difficulty because of the presence of hemosiderin, blood breakdown products, calcification and surrounding gliosis and accompanying venous angioma.

Treatment and outcome

The current, well-established indications for surgical resection of CMs are recurrent hemorrhage, progressive neurologic deterioration, and intractable epilepsy, unless the location is associated with unacceptably high surgical risk[5,20]. Despite its large size, good surgical outcome also has been reported in the reports of GCMs[2,4,8,10,11,24]. Therefore, complete surgical removal should be attempted when the operation is considered. In the present case, contrast-enhanced MRI showed venous angioma abutting medial portion of the mass. It is known that venous anomalies are often associated with solitary cavernous angiomas[13]. The presence of a venous anomaly in close proximity to a cavernous malformation is important for the surgeon, because injury to such veins can cause devastating venous infaction. In our case, complete removal was accomplished without injury to venous angioma.

CONCLUSION

We report a rare case of GCM that was completely removed by microsurgical treatment. This case provides important points for the practicing neurosurgeon to consider when making a differential diagnosis of large intracranial tumors. Since Imaging appearance of GCMs is variable, the possibility of CMs should be considered in the case of large tumor.

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