Giant cerebral cavernous malformation causing raised intracranial pressure in an adult: Case report and review of literature

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ABSTRACT

Giant cerebral cavernous malformations (GCCMs) are very rare malformations in adults, and they rarely present with raised intracranial pressure due to obstructive hydrocephalus. Around 20 cases of GCCMs have been reported in available literature. We report such a case, discuss and review the available literature regarding this eminently treatable group of vascular malformations.

Key words: Cavernoma, giant cerebral cavernous malformation, seizure

INTRODUCTION

Cavernoma is a congenital hemorrhagic vascular malformation characterized by the presence of sinusoid-like capillary vessels containing blood in very sluggish circulation.[1,2] Cavernomas vary in size from a few millimeters to a few centimeters. The majority of cavernous malformations are small, but it may reach significant size. Lawton *et al*. defined a giant cerebral cavernous malformation (GCCM) as a cavernoma with a diameter greater than 6 cm.[3] GCCMs are extremely rare[2] and only few cases have been reported in the literature.[1,2] In this article, we report such a case and review the available literature.

CASE REPORT

A 23-year-old lady presented with two episodes of sudden-onset, moderately severe holocranial headache over the past 4 years. The first episode subsided gradually over 1 month and she was not investigated. The second episode of holocranial headache was progressive and associated with repeated vomiting without any visual disturbances. Except papilledema, her neurological examination was normal. Magnetic resonance imaging (MRI) brain showed a deep seated left frontal periventricular mass lesion of size 6.5 cm × 4.8 cm × 4.2 cm causing hydrocephalus. Lesion was heterogenous on T1 [Figure 1a] and T2 [Figure 1b], with a peripheral hypointense rim on T2, with heterogenous contrast enhancement [Figure 1c]. She underwent a left frontotemporal craniotomy, middle frontal gyrus approach and gross total excision of the lesion. Lesion was soft, irregular and bluish-black, resembling bunch of black grapes containing areas of organized and liquefied blood clot. Yellowish pigmentation was noted in white matter surrounding the lesion. Post-operative recovery was uneventful. Post-operative imaging [Figure 2] showed total excision of the lesion. Histological examination was confirmatory of a cavernoma. Patient is asymptomatic at last follow-up of 2 years.

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DISCUSSION

Cerebral cavernous malformations (CCMs) account for approximately 5-13% of all cerebral vascular malformations.[1,2] Giant intracranial intraaxial cerebral parenchymal cavernomas are extremely rare, most commonly located in the parietal lobe and thalamus.[2]

The mechanism by which they enlarge is probably recurrent bleeding, followed by the organization of the clot, pseudocapsule formation and secondary expansion.[1] Re-endothelization of the hemorrhagic cavity, formation

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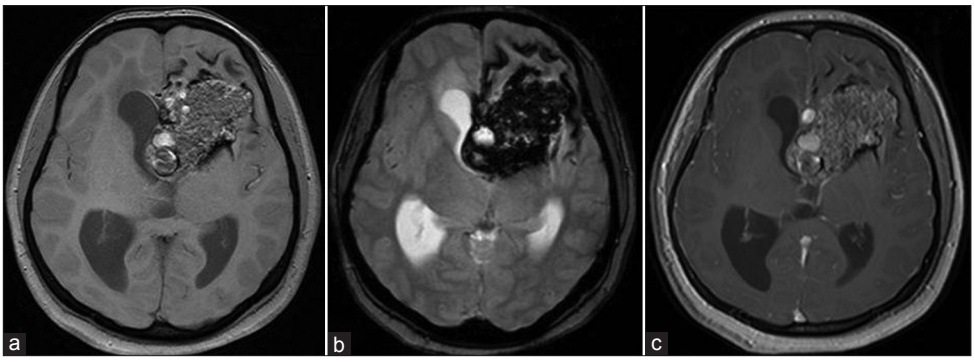

Figure 1: (a) T1 and (b) T2 weighted images showing a well-circumscribed heterogenous left basifrontal mass lesion of size 6.5 cm × 4.8 cm × 4.2 cm protruding into the left frontal horn with effacement of the left frontal horn and obstructing foramen of monro causing dilatation of the right lateral ventricle, with minimal surrounding edema; and (c) T1 contrast images showing heterogenous contrast enhancement without any prominent flow voids within or outside the lesion

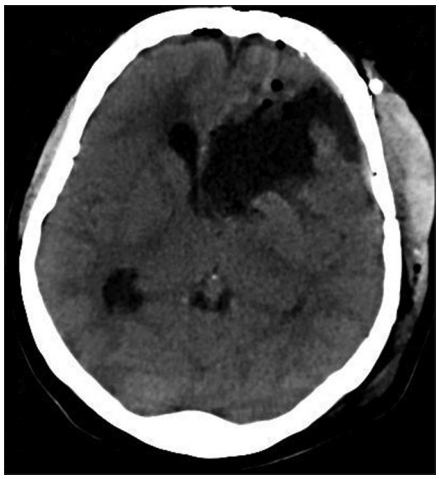

Figure 2: Post-operative computed tomography scan showing gross total excision of lesion

of new blood vessels and proliferation of granulation tissue increases the size of these lesions.[4] Although cavernomas typically present between the second and fourth decades of life, the majority of GCCMs reported in literature have occurred in children.[1] There is also a possibility of accelerated growth due to hormonal changes during puberty[2] and pregnancy.[5] Giant cavernomas do not differ from average-sized cavernomas in clinical, surgical or histopathological presentation but may differ radiologically.[5]

The annual clinically significant risk of hemorrhage in CCMs has been estimated at 0.7-1.1% per lesion per year.[2] However, true hemorrhage occurrence is relatively rare in GCCMs.[1] The risk of hemorrhage increases with lesions involving posterior fossa structures, females and previous history of hemorrhage.[4] Furthermore, there is a high risk (20-80%) of recurrent hemorrhage and progressive neurological decline after initial bleeding from a cavernoma.[2] There is no correlation between size of cavernoma and risk of hemorrhage.[5] The precipitation of acute symptoms occur when the bleed in a cavernoma ruptures through the perilesional hypointense hemosiderin rim seen on MRI.[2]

Imaging appearance of GCCM is variable. They exhibit a range of dynamic behavior including increase or decrease in size, *de-novo* formation as well as progression.[2] The MRI findings of cavernous malformations are classified into four categories. These include Type 1: Subacute hemorrhage; Type II: Mixed lesion consisting of loculated hemorrhage areas encircled with gliosis and hemosiderin; Type III: Chronic hemorrhage areas containing hemosiderin; Type IV: Small cavernous malformations resembling telangiectasia.[4] In most cases of intraparenchymal cavernous malformation, only a faint enhancement appears when contrast agent is used, although delayed images may show intense contrast enhancement.[5] Secondary changes such as calcification, hyalinization, thrombosis, cyst formation and cholesterol crystal formation has been reported in literature.[2] Cystic cerebral cavernomas are well-known.[2] Solitary cavernomas are often associated with venous anomalies. 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 infarction.[1]

The current, well-established indications for surgical resection of cavernomas are recurrent hemorrhage, progressive neurologic deterioration and intractable epilepsy, unless its location is associated with unacceptably high surgical risk.[1] Complete en-masse surgical excision should be attempted,[2] but can be removed piece-meal also. Usually, there are no enlarged vascular feeders. Accompanying, venous anomalies if any should be left undisturbed. Despite its large size, good surgical outcome also has been reported in the case reports of GCCMs.[1]

Although perilesional edema and mass effect is usually not seen with cavernous malformations,[1] presence of

papilledema suggesting raised intracranial pressure in our patient can be explained by obstructive hydrocephalus due to obstruction of foramen of Monro. Hence, strategic location of the lesion is as important as the nature of the lesion for causing its manifestations.

This report highlights a rare, but eminently treatable condition. The rarity of the condition along with its adult onset and presentation with raised intracranial pressure makes this case unique. Thus, possibility of GCCM should be considered in patients with large heterogenous intracranial tumors.

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