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Case Study

Giant Cerebral Cavernoma: A Case Study

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Abstract

Background: Cavernoma is known as cavernous malformation or cavernous angioma. It accounts for 0.5% of brain mass lesions. Giant cavernomas of the central nervous system is quite rare, only 65 cases of cerebral giant cavernous angioma have been included in literature over the last 62 years. They are more common in children and may be misdiagnosed as other intracranial neoplasms. This study presented a very rare giant cavernoma extended from right basal ganglia to the sylvian fissure in a 7-year-old female.

Case description: A 7-year-old female presented with the new onset of recurrent attacks of seizures, with progressive left-sided hemiplegia for the last month. The clinical examination showed that the patient was sleepy and had left-sided hemiplegia. A non-contrast CT scan revealed a spherical slightly hyperdense intraaxial lesion at the right basal ganglia extended to the sylvian fissure measuring 5x4.5x5 cm surrounded by moderate perifocal edema. A brain CT scan, with contrast, revealed slight patchy enhancement. MRI revealed a single large lesion occupying the right basal ganglia extended to the sylvian fissure measuring 5x4.5x5 cm and showed a patchy enhancement. The patient underwent craniotomy through the right fronto-temporal and transsylvian approach, under surgical microscope, with total en bloc resection of lesion. The histopathologic examination revealed cavernous hemangioma (cavernoma). After surgery, she was conscious alert, with no new neurological deficit apart from the pre operation Left-sided hemiplegia. The postoperative follow-up was uneventful with a significant improvement in her left-sided hemiplegia after 3 months.

Conclusion: Pediatric giant cavernous angioma is a rare intracranial lesion that may be best diagnosed with MR/CT, but sometimes, confirmation requires histopathological examination. It should always be included in the differential diagnosis of spontaneous intracerebral hemorrhages or large tumor. The best outcomes correlate with surgical excision, but may be, limited by eloquent tumor location.

In our case, we report a rare case of giant cavernoma 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 giant cavernoma is variable, the possibility of cavernoma should be considered in the case of a large tumor.

Keywords: Cavernoma, cavernous malformation, cavernous angioma, hemiplegia, seizure.

Introduction

Cavernoma, also known as cavernous malformation (CM) or cavernous angioma, is a benign occult vascular lesion, characterized by the presence of sinusoid-like capillary vessel containing blood in very sluggish circulation [1] that may occur in the CNS as well as in other organs such as the liver or skin [2,3]. The CNS cavernoma accounts for 5-13% of all intracranial vascular anomalies, 70–80% are supratentorial [4] and they vary in size from a few millimeters to a few centimeters in diameter [3,5]. However, giant cavernomas defined by Kan et al. [6] as a cavernoma with a diameter greater than 4 centimeters (cm) on preoperative MRI.

Most CMs are small and remain asymptomatic for long periods. Occasionally, supratentorial CMs precipitate the new onset of seizures and headaches, while infratentorial CMs more typically lead to acute/progressive neurological deficits [7, 8].

Few giant CMs (GCMs) have been reported in the literature and may mimic intracranial neoplasms or other vascular malformations. In this study, the Recently we experienced a case of cerebral giant cavernous malformation with a diameter of 5*4.5*5cm. In this study, the clinical, radiological features, the surgical management, and prognosis of this vascular malformation are described.

Case description

A 7-year-old female presented with the new onset of recurrent attacks of seizures, with progressive Left-sided hemiplegia for the last month. The clinical examination showed that the patient was sleepy and had left sided hemiplegia. A non-contrast CT scan revealed a spherical slightly hyperdense intraaxial lesion at the right basal ganglia extended to the sylvian fissure measuring 5x4.5x5 cm surrounded by moderate perifocal edema (Figure 1A). A brain CT scan, with contrast, revealed slight patchy enhancement (Figure 1B).

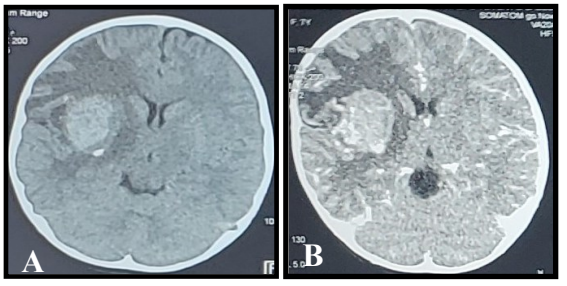

Figure 1: **(A)** Non-contrast CT scan revealed a spherical slightly hyperdense intraaxial lesion at the right basal ganglia extended to the sylvian fissure measuring 5x4.5x5 cm surrounded by a moderate perifocal edema and **(B)** Brain CT scan with contrast revealed slight patchy enhancement.

MRI revealed a single large lesion occupying the right basal ganglia extended to the sylvian fissure measuring 5x4.5x5 cm and showed an isointense signal on T1-weighted image (Figure 2A) and heterogeneous signal on T2-weighted images with peripheral hypointense rim (Figure 2B), with contrast, show patchy enhancement (Figure 2C).

The patient underwent craniotomy through the right fronto-temporal and transsylvian approach, under surgical microscope, which revealed a large dark brownish mass (Figure 3) with distinct margins from the surrounding tissues accompanied by gliosis. The lesion was totally resected en bloc with good control of bleeding (Figure 4),

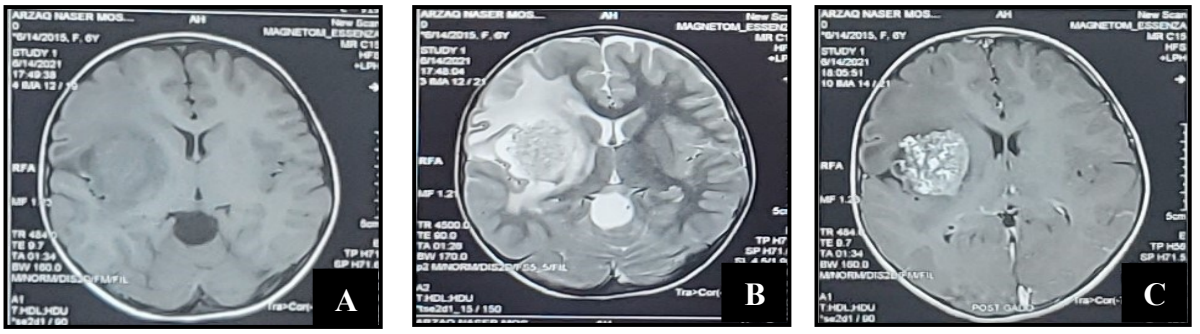

Figure 2: (A) T1-weighted MR image shows isointense signal, **(B)** T2-weighted MR image shows heterogeneous signal, and **(C)** contrast T1 weighted MR image shows patchy enhancement.

Figure 3: Dark brown cavernoma after en bloc excision.

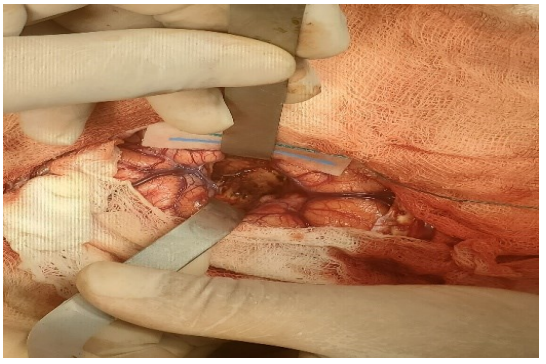

Figure 4: Cavity of tumor through the sylvian fissure.

and a follow-up CT-scan confirmed the total removal (Figure 5). The histopathologic examination revealed a multiple variable sized thin wall, and dilated blood vessels lined by endothelium resulting in cavernous hemangioma (cavernoma) (Figure 6).

After surgery, she was conscious alert, with no new neurological deficit apart from the pre operation left-sided hemiplegia. The

postoperative follow-up was uneventful with a significant improvement in her left-sided hemiplegia after 3 months.

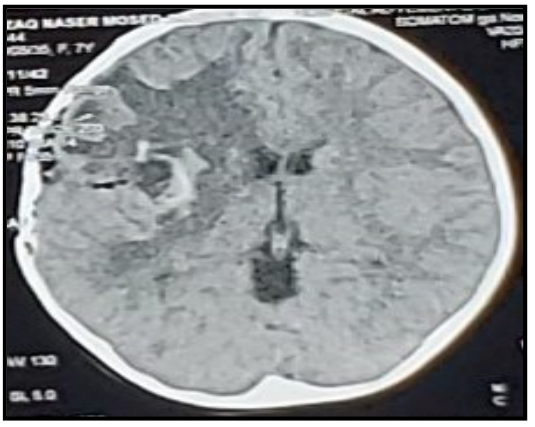

Figure 5: Post operation control brain CT.

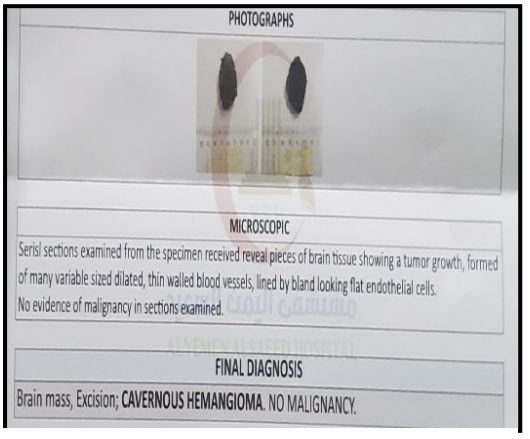

Figure 6: Histopathology contain description of cavernoma.

Discussion

Cavernoma, also known as cavernous angioma or cavernous malformation, is occult vascular benign malformations of the central nervous system [4]. It varies in size from a few millimeters to a few centimeters in diameter [3, 5]. However, a giant cavernoma is defined by Kan et al. [6] as a cavernoma with a diameter greater than 4 centimeters (cm) on preoperative MRI. In 2004, Lawton et al. defined a giant cavernous angioma as a lesion with a diameter over 60 mm [9]. CMs account for approximately 0.5% of brain mass lesions [8], and approximately 5 to 10% of all vascular malformations [10-13]. Only 65 cases of cerebral giant cavernous angiomas have been included in literature over the last 62 years and since the first case of Penfield et al. in 1948 [14]. At least half of them have been reported from 2008 to date. Jhawar et al. [8] reviewed 16 cases of giant cavernomas in all age groups but ranging in size from 4 cm and 6 cm. All lesions were reported as solitary and most frequently were found in the supratentorial white matter [4] (e.g., parietal lobe and thalamus) [8]. Our case study was found in the right basal ganglia extended to the sylvian fissure.

Cavernous angiomas have become more common and frequent in adults in the last two to four decades, but some researchers advocated that a quarter of cavernous angiomas are seen in the pediatric population [15]. While 59.4% of giant cavernomas occur in the pediatric population (less than 15 years old), in our case the patient was in the age of 6 years.

Growth and Size of Cavernoma

Cerebral cavernomas are typically 9–20 mm in size, and rarely attain larger dimensions. The mechanism by which they enlarge is probably repeated intralesional micro hemorrhages followed by the organization of the clot, pseudocapsule formation, and secondary enlargement [3,6,16]. However, it was also reported that CMs can show expansible growth without any evidence of a hemorrhagic event and mimic neoplasm [17].

The large size of the cavernoma in our case may be explained by the means of this hypothesis.

There is also a possibility of accelerated growth due to hormonal changes during puberty and pregnancy [2, 3].

Epidemiology

Although patients with CMs typically were presented between the last two to four decades [1,18,19], some researchers advocated that a quarter of cavernous angiomas are seen in the pediatric population [15]. The majority of giant cavernomas have occurred in children, with the youngest one being 3.5 months of age [20,24]. The overall prevalence among males and females is equal in the majority of cavernomas [1,11], but in giant cavernomas, there seems to be a female preponderance [24]. In our case study, the patient is female.

Familial cavernomas account for 20% to 50% of patients presenting with cavernomas [10], which is prone to be more symptomatic, likely due to the higher incidence of multiple lesions in these patients and the propensity for the novo cerebral cavernous malformation formation. In the review of giant cavernomas, no familial occurrence has been reported [24]. Multiple cavernomas may occur in 10% to 30% of sporadic cases and in up to 84% of familial cases [25], but it was not reported in any of the giant cavernomas [24].

For our patient, there was a single cavernoma with a negative familial cerebral cavernomatosis history.

Clinical Presentation

The most common symptoms of a cavernoma are seizure (ranging from 30-70% of cases) [6], followed by neurological deficit, hemorrhage, and headache [3]. Usually, the presentation of the giant cavernoma is not different from that of usual cavernomas [24-26]. Our case also presented with seizure and progressive neurological deficit. Yet, the presentation of large intracranial mass with signs of increased intracranial pressure in children was reported in some cases [21,23]. Hemorrhage of the cavernoma is reported to be 8% to 37% in adults and 36% to 78% in children [27]. However, true hemorrhage occurrence is relatively rare in giant cavernoma [9, 20, 24].

The risk of hemorrhage has been estimated at 0.7-1.1% per lesion per year [2, 7]. Fatal outcomes due to hemorrhage from cavernoma angioma are rare [5]. Most of the time, this extralesional hemorrhage is not immediately devastating [6], but there is a high risk of rebleeding with intervals ranging from weeks to years and may lead to catastrophe [3].

CT and MRI Appearance

On CT scan, giant cavernomas often present punctate or large calcification, and the mass effect is usually less than expected for the size of the lesion [3, 6, 7]. In our case, the right basal ganglia which extends to the sylvian fissure was isodense on CT scan with heterogeneous enhancement.

Typically, the MR imaging (MRI) appearance of a giant cavernous angioma is a heterogeneous “bubble of blood” which is named “popcorn-like” mass with/without cysts reflecting various states of degradation of blood [28], with a hypointense rim of hemosiderin on T2

weighted images, is nearly pathognomonic [6,7,16]. However, MRI variability is frequent [3,6,16]. Moreover, gradient echo MRI is useful to exclude other small cavernomas that may be occult on T1 and T2-WI.

Cavernomas usually have little or no surrounding edema nor mass effect [26,29]. Cavernoma may rarely be in the form of cystic growth with a well-defined capsule [29].

On the other hand, diagnosis may be challenging in giant cavernoma, which are rare lesions [26]. Imaging appearance of giant cavernoma is variable, ranging from completely cystic lesion [21, 23] to those resembling neoplasms with striking contrast enhancement and mass effect [21,24], and finally to the heterogeneous lesion with peripheral hemosiderin rim and without significant contrast enhancement and mass effect [26]. In our case, there was significant mass effect and patchy enhancement of cavernoma as discussed above.

Differential Diagnosis

The differential diagnosis of cavernoma includes not only pilocytic astrocytoma, hemorrhagic metastasis [4,17], low-grade or malignant cystic glioma, primitive neuroectodermaltumor, oligodendroglioma, or thrombosed arteriovenous malformation, but also spontaneous intracerebral hematoma [3, 7, 16]. In many instances, the final diagnosis of cavernous angiomas is based on histopathological examination.

Histopathological Appearance

The histopathological examination shows abnormally dilated blood vessels lined by a single endothelium layer without mural muscular or elastic fibers embedded within a matrix of collagenous tissue [8]. These vessels are in contact with each other without any intervening neural tissue and with no direct communication of the arterial with the venous system [3]. The vessels are separated by fibrotic tissue containing foci of calcification with hemosiderin deposition [3, 6], gliosis, and sometimes calcification or thrombosis eloquent [8].

Management

Surgical excision is the treatment of choice for cavernoma with recurrent hemorrhage, progressive neurologic deterioration, and intractable epilepsy, located accessible, non-eloquent [3], unless the location is associated with unacceptably high surgical risk [1,11]. The management of less accessible, deep cerebral, and brainstem lesions increasingly require stereotactic radiosurgery [3, 7], but the efficiency of radiosurgery for cavernous angioma remains uncertain. Thus, the surgical approach was justified in our case. According to many researchers, small cavernomas located in eloquent area or asymptomatic patients require observation [3, 6], however, there is no clear guideline for giant cavernoma in these cases as far as all the 65 cases were symptomatic.

Complete surgical removal should be attempted when the operation is considered [3].

Despite the large size of giant cavernoma, good surgical outcomes also have been reported in the literature of giant cavernoma [22, 23].

In our case, a complete removal was accomplished without post operation complication or new neurological deficit. In addition, the neurological deficit is improved after 3 months of physiotherapy post operation.

Conclusion

Pediatric giant cavernous angioma is a rare intracranial lesion that may be best diagnosed with MR/CT, although, sometimes, confirmation requires histopathological examination. It should always be included in the differential diagnosis of spontaneous intracerebral hemorrhages or large tumor. The best outcomes correlate with surgical excision but may be limited by eloquent tumor location.

In our case, we reported a rare case of giant cavernoma that was completely removed by microsurgical treatment. This case provides important ideas for the practicing neurosurgeon to consider when making a differential diagnosis of large intracranial tumors. Since imaging appearance of giant cavernoma is variable, the possibility of cavernoma should be considered in the case of a large tumor.

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