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Huge Cavernoma With Massive Intracerebral Hemorrhage in a Child

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ABSTRACT

Cavernous malformations are benign vascular lesions of the central nervous system that lack intervening normal brain parenchyma. They can be seen almost anywhere that normal vasculature is available. Lesions are raspberry-like, thin-walled vascular sinusoids without smooth muscles containing hemosiderin deposits. Cerebral cavernous malformations are characterized by small bleedings. Their size varies from a few millimeters to 2- 3 centimeters. Giant cases are rare. Also referred to as cavernoma, these lesions rarely lead to intracerebral hematomas that threaten life. In this report, we have presented a 14-year-old patient with a giant cavernoma leading to a life-threatening massive intracerebral hematoma.

KEY WORDS: Huge cavernoma, Child patient, Massive intracerebral hemorrhage.

INTRODUCTION

Cavernous malformations are benign hamartomas composed of vascular channels that lack intervening brain parenchyma (4). Although they can be seen at any age group, the peak age at occurrence varies between 20 and 40 years. They tend to behave according to age of the patient and the site of involvement. The risk of hemorrhage increases with lesions involving posterior fossa structures, female sex and previous history of hemorrhage (6). Their size varies from a few millimeters to 2-3 centimeters. Giant cases are rare. Supratentorial cavernous malformations leading to massive, life-threatening hemorrhage are also rare entities (2,3). The differential diagnosis is difficult in cases of calcified giant cavernoma that have previously bled. In this report, we have presented a 14-year-old child suffering from a deep coma due to massive intracerebral hemorrhage arising from a giant supratentorial cavernoma.

CASE REPORT

A fourteen-year-old male patient had initially been admitted to another medical facility where he had received medical therapy for the diagnosis of acute gastroenteritis due to complaints of a headache lasting for a week, nausea and vomiting. He was dispatched to our emergency room after magnetic resonance imaging (MRI) and computerized tomography (CT) investigations were performed due to rapid alteration in his level of consciousness. Physical examination was normal at the time of admission. Neurological examination revealed a decreased level of consciousness, namely severe confusion, in addition to orientation and cooperation disorders. He was localizing painful stimuli. The score on the Glasgow Coma Scale was 8 (E2 M4 V2). Cranial

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CT revealed a well-circumscribed mass of 6 cm in diameter, at the left fronto-parietal region, causing a shift in the midline and compression on the left lateral ventricle (Figure 1). There were hyperdense, lobulated areas of patchy calcifications at the lateral side of the mass. On the medial side of the lesion, there were bleeding areas, which were denser than the surrounding brain parenchyma, but less dense than previously described lesions. Additional to the CT findings, MRI demonstrated a lobulated mass of 42x35 mm with peripheral hemosiderin deposits on T2-weighted images. The hemorrhage in the medial portion of the lesion was thought to be in an early subacute stage (hyper-intense on T1-weighted and hypo-intense on T2-weighted images) (Figure 2A, B, C).

The subacute intracerebral hematoma was evacuated with a left fronto-temporal craniotomy and a well-circumscribed, purple, lobulated mass consisting of vascular structures was excised totally. The pathological diagnosis was cavernous hemangioma (Figure 3). The post-operative period was uneventful. The patient's level of consciousness gradually improved and he was discharged on the 10th post-operative day. There was no residual mass on the control CT performed three months later (Figure 4).

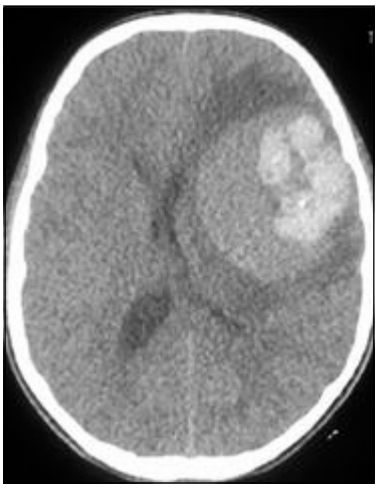
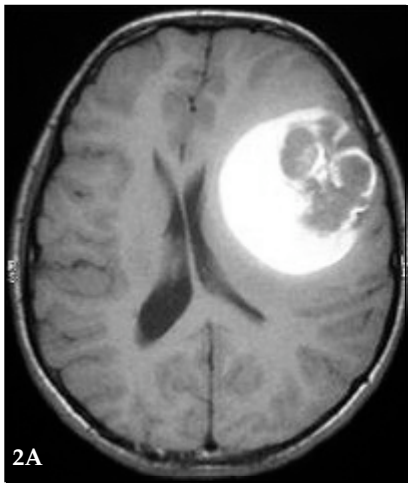


Figure 1: Non-enhanced CT demonstrating hyperdense, heterogenous lesion and areas of hemorrhage at the left fronto-parietal region including millimetric calcifications with lateral ventricle compression and midline shift.



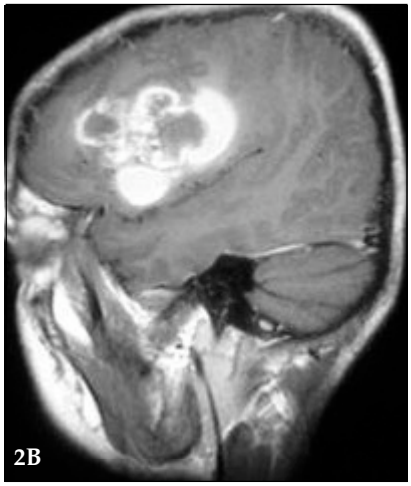


Figure 2 A-B: Non-enhanced axial and sagittal T1-weighted images (**2A-2B**) revealing hypointense lesions with peripheral hyperintensities at the left fronto-parietal region and subacute hemorrhage medially surrounding these lesions.

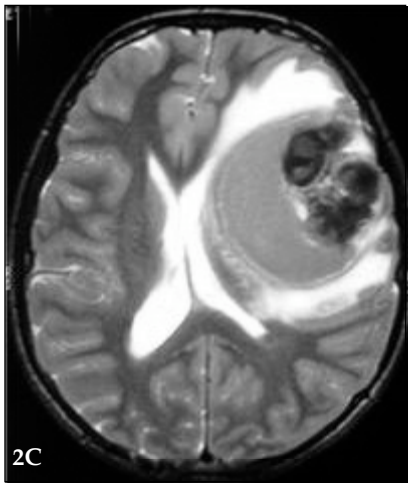


Figure 2 C: Axial T2-weighted image (2C) revealing low signal intensities around hypointense lesions originating from hemosiderin.

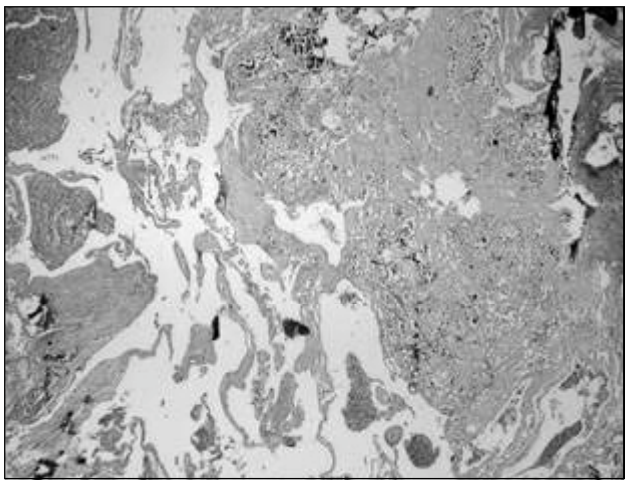


Figure 3: Cross-sectional evaluation of the specimen revealed a lesion consisting of randomly distributed telangiectatic and congested vascular channels in a stroma formed of hyalinized fibrous connective tissue. There was no intervening neural tissue (HE, 100X).

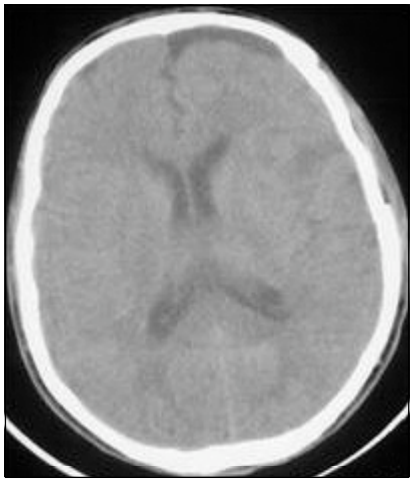


Figure 4: Follow-up CT at the 3rd post-operative month with no space occupying lesion and no pathological densities.

DISCUSSION

Cavernous malformations are benign vascular lesions of unknown etiology, with low pressure levels (1,4,6,7). Pathologically, these lesions do not contain smooth muscle or elastin, and lack intervening normal brain parenchyma. They are encircled by hemosiderin and gliosis. The vascular structures making up these lesions may or may not be thrombosed. They can be seen anywhere along the neuroaxis – brain and the spinal cord. Although they are usually single, multiple lesions can also be present. Twenty five percent of cavernomas are seen in the pediatric age group (4, 9).

Cavernous malformations constitute less than 1% of all intracranial mass lesions and 5-10% of all central nervous system vascular malformations. Eighty percent of the lesions are supratentorial. They can be asymptomatic; however, findings resulting from acute or chronic bleeding and mass effect can be observed. Epilepsy is the leading clinical finding in supratentorial lesions seen in 40-50% of the cases. On the other hand, 20% of the cases present with focal neurological deficits (7, 9). The annual bleeding incidence of cavernomas is 0.2–2%. Cases with previous bleeding history are at higher risk. Similarly, female sex and positive family history increase the risk for bleeding. Additionally, this risk is affected by site of involvement. Brain stem lesions, in particular, tend to bleed more severely than lesions at other sites (7). Although almost all of the cases have micro-bleedings, bleeding leading to clinical symptoms is a rare entity in cavernomas (0,25-6%). Since these lesions are small in diameter and their flow rates are low, they rarely cause life-threatening intracerebral hematoma (5, 8). Nevertheless, the case presented in this report had neither previous history of headache, epilepsy, nor any neurological deficits but presented with massive intracerebral hematoma resulting from a giant cavernoma.

The MRI findings of cavernous malformations are classified in 4 categories as: 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. Type I and II lesions are relatively large and they can be seen clearly on T1- and T2-weighted images, whereas Type III and IV lesions are best visualized in the gradient echo sequence. High tesla MR devices are much more sensitive and specific in the diagnosis of small and angiographic cryptic lesions (7). The case described in this report was a type II cavernous malformation according to the MR images. The dimensions of cavernomas vary from a few millimeters to centimeters; however, they are usually 2–3 cm in diameter and usually do not cause peripheral edema. On the other hand, the hemorrhage can increase the edema. Re-endothelization of the hemorrhagic cavity, formation of new blood vessels and proliferation of granulation tissue increases the size of these lesions. Thirty

percent of these lesions show calcifications on CT scans. Low-grade glial tumors, oligodendroglioma, thrombosed AVMs, metastatic melanoma, toxoplasmosis and cysticercosis should be included in the differential diagnosis of cavernous malformations (10). In our case, the unusual size of the lesion and the evident peripheral edema complicated the pre-operative diagnosis.

In conclusion, although cavernomas are small or medium-sized, benign vascular malformations with low flow rates, as in our case, they can be of giant dimensions leading to life-threatening massive intracerebral hemorrhages. Additionally, giant, heterogeneous lesions with marked peripheral edema resulting form massive hemorrhage can impede consistent pre-operative diagnosis.

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