CASE REPORT

Multiple supra- and infratentorial cavernous hemangiomas in a five year–old girl

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Abstract

Cavernous hemangiomas (CHs) are vascular structures comprising abnormally dilated blood vessel clusters. This anomaly is estimated to occur in approximately one out of every 500-600 people. Individuals often show the first sign of cavernous hemangiomas in their second or third decade. Therefore, the presentation of this disorder is not common in children. This study presents a five year-old-girl who developed abruptly nearly fixed right-sided eye deviation and incoordination after a short course of viral infection. In physical examination, she had left peripheral facial, right eye oculomotor, and left abducens nerve palsy. Her brain's computed tomography (CT) scan revealed hemorrhages in the posterior aspect of the pons and some areas of the hemispheres in different stages. Following brain magnetic resonance imaging (MRI), multiple popcorn ball low-signal T2* lesions with both supra- and infratentorial locations with marked peripheral hypo intensities were seen. These findings were in favor of multiple cavernous hemangiomas of the brain. The novelty of this case was due to observing cavernous angiomas in both supra- and infratentorial spaces in pediatric age.

Keywords: Cavernous Hemangiomas, Children, Magnetic Resonance Imaging

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Introduction

Cavernous hemangiomas (CHs), or cavernous malformations (CMs), cavernous angiomas, or cavernomas, are intracranial vascular malformations. They are cystically-dilated vascular spaces composed of a single endothelium layer without elastic lamina and smooth muscle cells embedded in a collagenous extracellular matrix (1). The disease occurs sporadically or in a familial pattern and has a similar distribution

between males and females. These lesions are histologically identical in association with all genotypes, Patients with cavernous hemangioma typically present during the 2nd to 5th decades of life. The most frequent clinical manifestations include seizures and symptomatic hemorrhage (2). Other related symptoms of this disorder are weakness, numbness, visual disturbance, and language difficulties (3). However, in some cases, no symptom is manifested (4). In population-based studies, the risk of the first symptomatic hemorrhage among incidentally identified lesions is shallow (0.08% per year). However, when a symptomatic hemorrhage occurs, the annual recurrence risk dramatically increases to 42% in five years. Brainstem lesions are associated with a greater risk of initial bleeding and recurrence (2,5). The best technique for the diagnosis of CHs is magnetic resonance imaging (MRI). CHs mostly have no observable symptoms or diagnostic criteria (5). However, their timely detection is an urgent problem for preventing secondary symptoms.

Case presentation

A five-year-old girl was referred to our center due to sudden deviation of both eyes to the right side, left-sided facial palsy, and incoordination after a short course of viral infection. The patient was the first child of non-consanguineous parents, born by normal delivery. She had no significant medical history. The nervous development of the patient was normal before the onset of the disease. Cranial nerve examinations showed the patient could not rotate her eyes and had right-sided oculomotor and left-sided abducens palsy. She was also unable to move the left side of her face and suffered from left peripheral facial palsy. She also could not walk independently. Other neurologic examinations revealed her patellar deep tendon reflexes decreased slightly, and her left side's plantar reflex was mute. Because of nearly fixed eye deviation and inability to cooperate for cerebellar tests, especially finger to nose, these tests were not performed. In addition, she had incoordination in her standing position.

On abdominal examination, there was no organomegaly, and heart and chest auscultation were normal. For more investigation, MRI was done. Multiple low-signal T2* lesions (varying from 1 to 8 mm) in both supra- and infra-tentorial locations (lesions in both cerebral hemispheres, caudate nucleus, left cerebellar hemisphere, and posterior aspect of the pons) with no surrounding edema in the parenchyma in favor of popcorn appearance were detected. However, some subacute to chronic hemorrhages were seen as well. These findings were highly suggestive of cavernous angioma (Figures 1 and 2).

No noticeable enhancement was seen in post-contrast images. Brain magnetic resonance angiography (MRA) revealed normal large and medium-sized vessels, so vasculitis and other significant vessel malformations were ruled out. A spinal MRI was requested to investigate other parts of the nervous system, and no abnormality or lesion was observed.

To determine whether the lesion was hereditary, MRI evaluated the parents; no lesion was detected in both parents' images.

After two weeks, the rotation of the eyes improved, and incoordination resolved within a month.

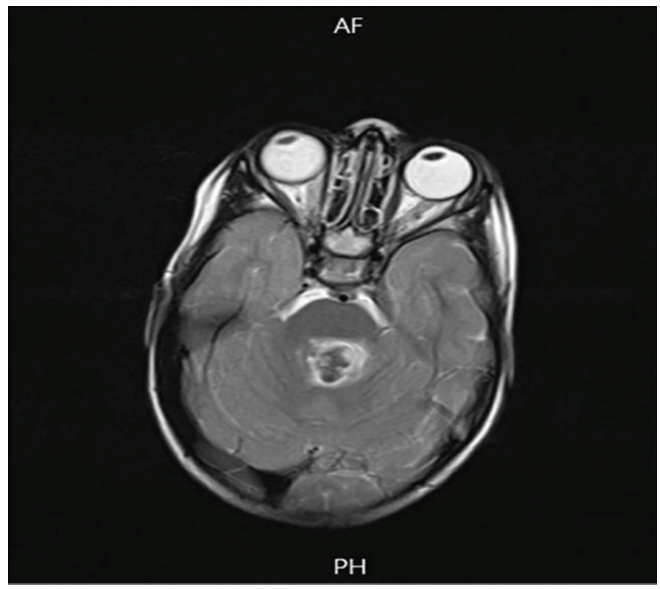

Figure 1. *Axial T2 Weighted image reveals a hypointense lesion with peripheral hypersignal rim in posterior aspect of pons*

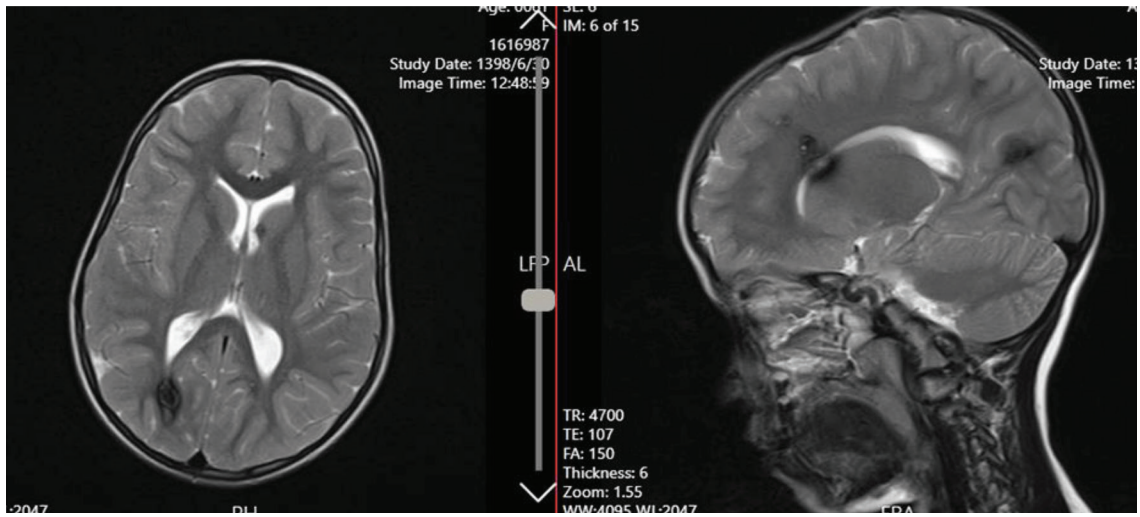

Figure 2. *Axial and sagittal T2- Weighted MRI show multiple pop-corn like lesions in supra-tentorial area (posterior periventricular area and perpendicular to corpus callosum).*

Discussion

Based on the previous reports, the prevalence of pediatric cavernomas is low, accounting for approximately 0.2% of infants and 0.6% of children (1, 6). Thus, these disorders are rare in children in comparison with adults. On the other hand, observing both supra- and infratentorial lesions in the pediatric population is rare. In this case, fixed eye deviation was due to dysfunction of cranial nerves IV and VI. These involvements were reported previously in some cases, predominantly in adults (7-10). These findings could be attributed to recent bleeding in the hemangioma in the posterior aspect of the pons. This case also could not walk independently, so its incoordination could be due to the pressure effect of bleeding and its edema on the middle cerebellar peduncle or cerebellospinal tracts.

Bleeding can cause local ischemia, edema, and permanent nerve damage. CHs are a kind of damage historically called (angiographical occult vascular malformations) (11). Therefore, MRI is

one of the best techniques for diagnosing CHs. CHs size, bleeding, and any other lesions can be detected by MRI (12). In this case, this technique was also used for the CHs detection. Production of a "salt and pepper or popcorn ball" appearance on a T2-weighted scan is a characteristic sign of CHs (13). Low-signal T2* margin, which surrounds popcorn ball lesions, is a hemosiderin deposition and can be seen in most lesions (15). Most studies mentioned that observing these low-intensity rims in T2-weighted images around the popcorn lesions is a significant hallmark of CHs (14-19). This case also had this characteristic pattern, especially in supratentorial lesions (Figures 1 and 2). Similar reports indicated that supra-tentorial location was the most common site for the lesions. However, in this case, lesions were seen in both supra- (basal ganglia and periventricular) and infratentorial (brainstem and cerebellum) sites. CHs with spinal involvement occur with the frequency of 3–5% (20, 21). In this case, there was no evidence for the spinal involvement.

It has been clarified that several items are related to the increased risk of later hemorrhage. These risk factors include infratentorial lesions, familial lesions, and recent hemorrhage (less than two years ago)(2,12). Because this patient had an infratentorial lesion that led to symptomatic manifestation, she is in the group with elevated risk.

In Conclusion

Despite of rarity of cavernous hemangiomas as an etiology for neurological disorders in children, they can be seen with exciting manifestations. In patients with cavernous hemangioma, observing both supra- and infratentorial lesions are infrequent, especially in children, and clinicians and radiologists should be aware of this diagnosis.

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Author's Contribution

Definite diagnosis of the case and critical revision of the manuscript for important intellectual content: Hosein Eslamiyeh, Zahra eslamiyeh

Conflict of Interest

The authors have declared no competing or potential conflicts of interest

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