Cerebellar cavernous hemangioma that presented with posterior neck myalgia

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Abstract

Cavernous hemangioma can occur in the entire brain but rarely in cerebellum, especially in the pediatric age group. Headache, seizure, gait disturbance, recurrent bleeding may be seen. This tumor is a relatively benign condition but if the lesion located in the posterior fossa or the brain stem bleeds, irreversible brain damage may occur because of its restrictive space. Moreover, it must be differentiated from malignant tumors. We report 12.6 year old boy who represented posterior neck myalgia as the presenting symptom. The pain continued for about a month despite analgesic medications. Brain MRI showed intracranial hemorrhage in the left cerebellum (4.5 cm) representing repeated hemorrhages at different times and originated from the cavernous hemangioma accompanied by mild hydrocephalus. The lesion was surgically removed successfully and the cavernous hemangioma was confirmed by pathologic findings. After the follow-up period of 14 months, he is in good condition without any complications. (Korean J Pediatr 2008;51:1363-1367)

Key Words: Cavernous hemangioma, Cerebellum

Introduction

Cavernous hemangioma, also known as cavernous angiomatous vascular malformation, accounts for 10-20% of vascular malformations. Cavernous hemangioma is found in all regions of the brain and usually located supratentorially, with 20.7% incidence in the infratentorial region including pons (14.6%) but rarely in cerebellum (6.1%) previously. The natural history of cavernous hemangioma was considered to be relatively benign, with significant hemorrhage being quite uncommon. However, recent experience has shown that cavernous malformation is not necessarily a benign clinical entity, as recurrent hemorrhage and subsequent neurological deficits may occur. Moreover, recent reports tend to demonstrate a higher hemorrhagic risk in the pediatric age group than adults.

Patients may have the signs and symptoms of headache, seizure, and the gait disturbance or the recurrent intracranial hemorrhage. Overall, cavernous hemangioma can be curable in complete extirpation if a surgical intervention is approached. And it should be differentiated by a diagnosis with malignant tumors through the obtained specimen.

We report a case of cerebellar cavernous hemangioma that presented with posterior neck myalgia as the chief complaint.

Case report

A 12-year 6-month-old boy had developed the posterior neck myalgia for a month. He visited a local clinic and was given analgesics but his symptoms did not improve. He was referred to our hospital to manage the severe neck pain and poor oral intake for about 3 days. He had difficulty controlling his head because of the deteriorating posterior neck pain. Upon neurological examination at admission, he was alert without neurological abnormalities. MR imaging performed on the following day of admission revealed intracranial hemorrhage in the left cerebellum (4.5 cm), most likely from the cavernous hemangioma. Mild hydrocephalus was shown (Fig. 1). This hemorrhage was presumed to have happened at different times. On the 3rd day of admission, the left suboccipital craniotomy was performed then the abnormal vascular structure was removed. The hemangioma on the left cerebellum was measured by 4.5×4.5 cm, the shape looked like the cluster of grapes at the surgical field (Fig. 2). Post-
operatively, his posterior neck myalgia improved and after ICU care, he was discharged on the 15th hospital day. The pathologists report identified cavernous hemangioma with old hemorrhage and organizing thrombus and reactive gliosis.

After one month, he recovered without any complications. The brain CT showed the resolution of intracranial hemorrhage in left cerebellum (Fig. 3). On a follow-up visit at 14 months, he is doing well without neurologic deficit.

**Discussion**

Cavernous hemangiomas belong to innate benign intra-cerebral vascular diseases and are characterized by the loss of muscular layer and elastic fibers within the lesion. Before the clinical application of CT, it was difficult to diagnose the disease because of negative angiography results. Scanning with high-field MRI shows that cavernous hemangiomas present specific images: there are reticulated, mixed signals
from the hemorrhage with breakdown products in the center of the lesion, around which a signal loop is shown due to hemosiderin pigmentation. The extensive application of MRI increases the diagnostic level, which makes it easy to diagnose more patients with cavernous hemangiomas. Recently, due to extensive application of magnetic resonance imaging (MRI), an increasing number of patients with cavernous hemangiomas have been able to be determined.

In one study, a sharply demarcated spherical intracerebral hematoma or heterogeneous lesion should always make one consider the hypothesis of a cavernoma in pediatric age group.

Cavernous hemangiomas can affect patients of all ages. From newborns to people aged 84 years old, the incidence rate reaches its peak in patients about 30 years old. In 1995, Maraire and Awad confirm that one fourth of the patients in the various series are children.

There are slightly male of female predominance, but majority of studies shows no differences in sex distribution. The distribution of cavernous hemangiomas is related to the volume of nerve tissue. Although cavernous hemangiomas located in the cerebrum are frequent, their predilection site is not obvious. In children, supratentorial locations account for about 80%, while the other 20% are located in the posterior cerebral fossa. In the pediatric age group, brainstem locations seem to be a little more frequent than in adults, and the pons (14.7%) is the most common site. Cavernous hemangiomas in the cerebellum in pediatric age group like our case is uncommon.

Cerebral hemorrhage, epilepsy, headache, and neurological impairment are the main clinical manifestations. Some patients with cavernous hemangiomas, however, do not have any related symptoms or present several symptoms simultaneously with the slow progression of the disease. Our patient presented posterior neck myalgia without neurological impairment as initial symptom, which is very unusual.

Since intrapachial cavernous hemangiomas are located deeply and even can be found in the brainstem and other important functional regions, it is not easy for a routine craniotomy to find the lesions accurately. The natural history of brainstem cavernous hemangiomas indicates a higher morbidity and mortality than cavernous hemangiomas in other locations. Radiosurgery appears to diminish the incidence of further haemorrhages but not remove the risk completely.

In patients with cavernous hemangiomas, moderately advanced or acute neurologic impairment is associated with hemorrhage of cavernous hemangiomas. Therefore, surgical treatment should be considered despite the certain risks of the operation. Surgical treatment is suitable for patients with cavernous hemangiomas suffering from hemorrhage and neurological impairment: the disability rate of the resection operation is very low.

Zimmerman et al. thought that if repeating hemorrhages of the brainstem and the progressive aggravation of symptoms of the nervous system were confirmed through MRI, patients with cavernous hemangiomas located in superficial positions that offend the surface of the pia mater and whose surrounding tissue can be separated should consider surgical treatment. Patients with cavernous hemangiomas that do not reach the surface of the pia mater and that do not display symptoms of the nervous system may be monitored without specific management. Sami et al. have made their opinions even clearer and go further in their recommendations. On the one hand, they recommend intervention in the case of superficial cavernous hemangiomas if the patient is young, even though it might be a serendipitous finding without haemorrhage. On the other, they recommend operation in case of progressive deterioration, with further haemorrhage, even though the cavernous hemangioma may not be superficial. The possibility of reaching deeply located cavernous hemangiomas and removing them with a very low morbidity have already been referred to by Mathiesen et al.
and females of reproductive age should actively adopt surgical treatment since there is more of an opportunity for occurrence of the lesions and hemorrhage in these patients\(^\text{20}\).

The prospective annual rate of hemorrhage from cavernous malformation is 4.5% in patients with previous hemorrhagic episodes, but only 0.6% in those without previous bleeding\(^\text{21}\). Early recurrent hemorrhage is thought to be unusual, but successive hemorrhage may occur at shorter time intervals. The interval from the initial symptomatic hemorrhage to the first recurrent hemorrhage ranged from 1 to 60 months and the second rebleeding occurred earlier; one patient experienced recurrent hemorrhages less than 1 week after the previous episode\(^\text{20}\). Some malformations which became hemorrhagic may continue to bleed intermittently\(^\text{20}\).

The indication and timing for surgery of cavernous malformations is still unclear. The risk of significant hemorrhage from the malformation is relatively low, so prevention of hemorrhage should not be an absolute surgical indication\(^\text{21}\). However, some patients certainly should undergo surgery to improve or halt neurological aggravation. Hemorrhage in the posterior fossa is more likely to manifest clinically than that occurring above the tentorium.

Cavernous malformations located within the brainstem are known to have an aggressive clinical course, but can sometimes be removed without serious surgical morbidity. In contrast, conservatively managed patients within the brainstem lesion may die from a subsequent hemorrhage\(^\text{17}\). The limited space of the posterior fossa cannot tolerate mass expansion, so the damage to the surrounding brain tissues will be irreversible. Therefore, we did not have any reason to delay the surgical operation in our patient.

Sandalcioglu et al.\(^\text{20}\), reported five cases whose cavernous hemangiomas were located in the cerebellum had favorable outcomes after surgery. Patients with cavernous hemangiomas accompanied by progressive neurological impairment or a recurrence should be treated through surgical means. If lesions are close to the brain surface, cutting open the pia mater should be avoided and therefore, the surgical risk is very low. If cavernous hemangiomas located within the brainstem are large in size but not accompanied by hemorrhage or extensive calcification, it is more difficult for surgeons to remove the lesions and therefore, there is a greater surgical risk\(^\text{20}\).

Our patient presented with symptoms of posterior neck myalgia not with neurologic symptoms, which may be due to recurrent hemorrhages from this cerebellar lesion. Because his neurologic signs were not obvious, we misunderstood his pain was originating from the cervical spine. Hence, his cerebellar lesion was found accidentally during the cervical spinal MRI imaging. After the successful surgical removal of cerebellar cavernous hemangiomas, he is in good condition without any complications after 14 months of follow-up.

### References

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