



Case Report

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A Case of Widespread Cavernous Malformations of the Central Nervous System Associated with Acute Neurologic Deficit

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A 45-year-old female visited our clinic due to sudden right leg weakness and sensory loss. Brain and spinal cord magnetic resonance imaging showed widespread cavernous malformations. Cavernous malformation in L1 spine area was accompanied by a subacute stage hematoma with perilesional edema. Sensory loss subsided after corticosteroid therapy. Usually, neurologic deficit by spinal cavernous malformation appears more chronically in the adults compared to children. Treatment options are difficult to establish in a case with multiple cavernous malformations. Identifying hemorrhagic lesions by extensive neuroimaging evaluation could be helpful to select the treatment target for cavernous malformation.

Keywords: Cavernous malformations; Hemorrhage; Acute neurologic deficit

INTRODUCTION

Cavernous malformations (CM) rarely involve the spinal cord. In an adult, CM accounts for 5% of the intramedullary mass. A single lesion is the most common presentation in the spinal cord. Rarely, multiple CMs can involve multiple segments of the spinal cord and cerebral hemispheres. In adults, diffuse CM involvement in the central nervous system (CNS) is a rare condition without a family history of CM and previous radiation therapy on the CNS (1).

If the cases are pediatric, or have a history of previous radiation therapy on the CNS, acute bleeding of the CMs commonly causes an acute neurologic deficit (2, 3). However, in adult cases, although the bleeding is commonly associated with CMs, hemorrhagic spinal cord CM is more likely to be associated with a slow or gradual worsening of neurologic deficit (1).

Herein we report a case with an acute neurologic deficit associated with multiple CMs of the CNS.

CASE REPORT

A 45-year-old female visited our clinic due to sudden weakness and paresthesia in

her right leg. She reported that the weakness of her right leg had become worse over the previous 15 days. She had been treated for essential hypertension. She was referred

to our hospital for multiple cerebral CMs identified in brain magnetic resonance imaging (MRI) performed in another hospital.

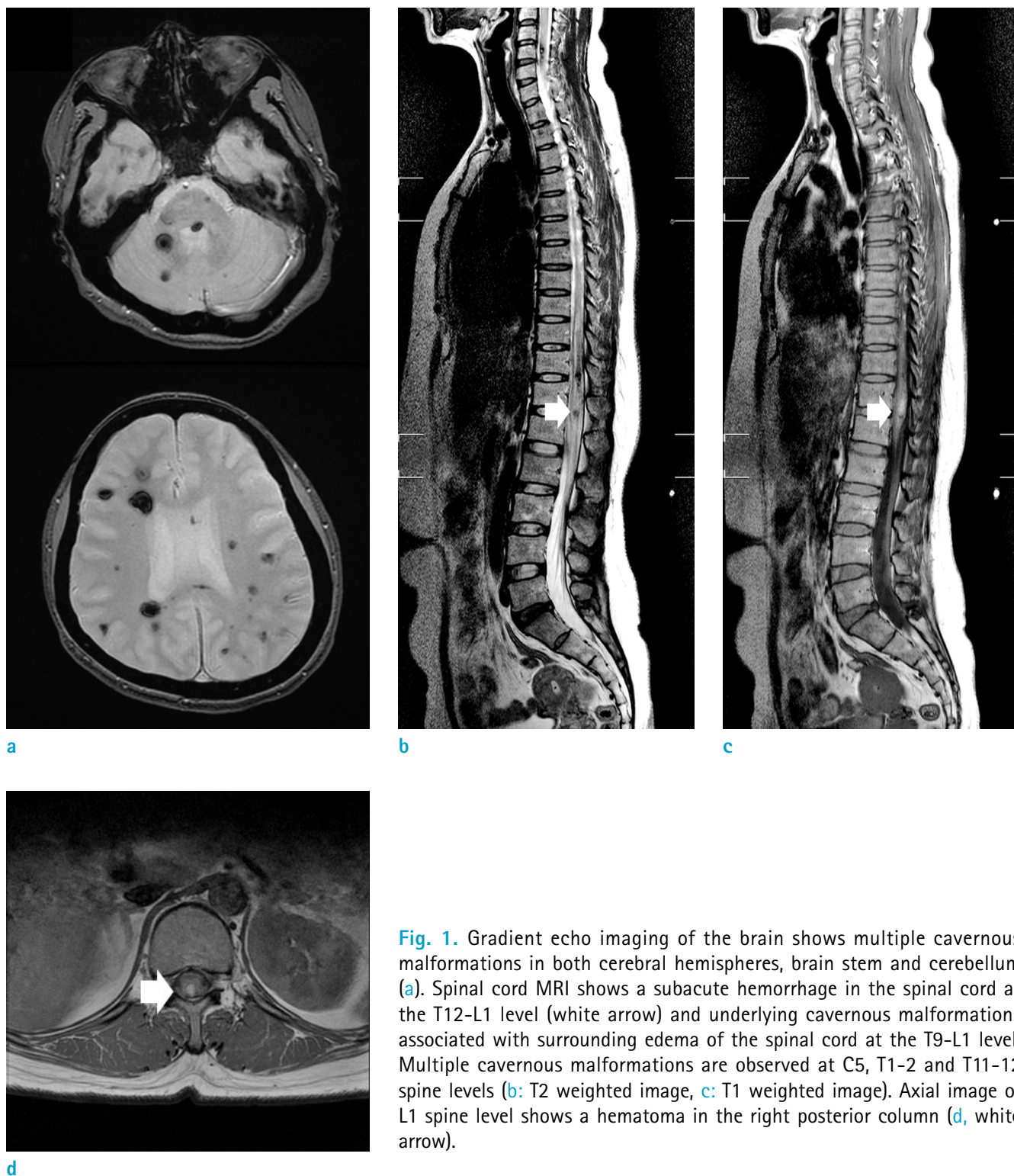


Fig. 1. Gradient echo imaging of the brain shows multiple cavernous malformations in both cerebral hemispheres, brain stem and cerebellum (a). Spinal cord MRI shows a subacute hemorrhage in the spinal cord at the T12-L1 level (white arrow) and underlying cavernous malformations associated with surrounding edema of the spinal cord at the T9-L1 level. Multiple cavernous malformations are observed at C5, T1-2 and T11-12 spine levels (b: T2 weighted image, c: T1 weighted image). Axial image of L1 spine level shows a hematoma in the right posterior column (d, white arrow).

Vital signs were within normal limits at the time of admission. Her right leg showed a normal muscle tone with mild motor weakness in hip flexion/extension, knee flexion/extension and ankle dorsiflexion/plantar flexion which all graded 4 using the Medical Research Council grade. Deep tendon reflex of right knee and ankle were slightly decreased. Pain, temperature, position and vibration sensory deficits were found in her right leg. Sphincter function was normal.

Brain MRI showed multiple CMs in the bilateral cerebral hemisphere, cerebellum, pons and medulla (Fig. 1a). All brain lesions showed no perilesional edema. To identify the culprit of her acute neurologic deficit, spinal cord MRI was performed. In the spinal cord MRI, a subacute hemorrhage associated with perilesional edema was found at the T12 ~ L1 spine level with an underlying CM. Multiple CMs were also found at C5, T1-T2, and T11-12 spine levels (Fig. 1b-d).

After intravenous administration of dexamethasone (10 mg loading, and 4 mg q 6 hours) for reducing vasogenic edema, sensory deficit had improved but motor deficit had not. However, she refused further surgical treatments.

DISCUSSION

A rapid progression of neurologic deficit associated with CM bleeding commonly appears in pediatric cases. The bleeding location is also well correlated with neurologic deficits in pediatric cases (2, 3). In adults, the neurologic deficit is presented more chronically (1, 4). This is because neurologic deficit is known to be triggered by perilesional gliosis, which is related to repetitive micro-bleeding,

In the present case, considering MRI findings of the bleeding spinal cord CM (high signal intensities in T1 weighted imaging, and low signal intensities in T2 weighted imaging), the bleeding was in a subacute stage that correlated with the time of the patient's neurologic deficit.

CM bleeding risk is differentiated by the CNS regions involved. Spinal cord or brainstem CMs are more likely to be associated with bleeding than CMs in the cerebral hemispheres. According to previous literature, the bleeding risks can be assumed to be 1.6-4.5% (per 1 CM, per 1 year) in the spinal cord, 0.7-1.3% in the cerebral hemispheres, and 2.7% in the brainstem (5, 6). In the present case, the patient had more than 40 CMs in the cerebral hemispheres, 4 CMs in the spinal cord, and 6 CMs in the brainstem and cerebellum. Therefore, she arithmetically had at least more than 50% bleeding risk in the CMs.

Complete surgical removal is only preventive for CM bleeding. Depending on the location, surgical removal can be impossible, or associated with high risk. Therefore, surgical treatment of multiple CMs is limited. In a previous study, sensory symptoms accompanied by a CM can be treated conservatively without a difference in prognosis with surgical treatment (7). Motor benefit from the surgical treatment was also not significant (7). Therefore, conservative treatment may be a more reasonable option for cases with less severe neurological deficit and high surgical risks (7).

Multiple globular lesions in the CNS need to be differentiated from cerebral amyloid angiopathy and hemorrhagic metastasis. However, the size of the lesions was larger than cerebral amyloid angiopathy and there was no evidence of malignancy.

Multiple CMs in the CNS are commonly associated with a family history of CMs or previous radiation therapy on the CNS. With the positive family history, genetic abnormalities can accompany CMs (2, 8, 9). However, genetic testing was not performed in this case. The CNS of her family members also was not screened by neuroimaging or genetic testing. Therefore, the experience from present case might not be applicable to non-genetic cases.

In the present case, for acute neurologic deficit associated with diffuse CMs in the CNS, treatment options are difficult to establish. Extensive neuroimaging evaluation for identifying symptomatic CAs needs to be considered in cases with diffuse CMs of the CNS.

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