Cryptic Arteriovenous Malformation of the Choroid Plexus of the Fourth Ventricle
—Case Report—

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Abstract

A 54-year-old female presented with a cryptic arteriovenous malformation (AVM) of the choroid plexus of the fourth ventricle causing intraventricular hemorrhage. Computed tomography and magnetic resonance imaging disclosed the lesion near the fourth ventricle, but bilateral vertebral angiograms showed no abnormalities. The preoperative diagnosis was cavernous angioma. The mass was removed completely, and histological examination demonstrated an AVM of the choroid plexus. Vascular malformations of the choroid plexus of the fourth ventricle are extremely rare. The possibility of this lesion being the cause of primary intraventricular hemorrhage of unknown origin should always be considered.

Key words: vascular malformation, choroid plexus, intraventricular hemorrhage, cryptic arteriovenous malformation

Introduction

Vascular malformations of the choroid plexus are rare and usually manifest as intraventricular hemorrhage.1,2,7,8,25> These lesions are often too small to be detected by angiography, but are visualized more frequently by computed tomography (CT),1,3,18,27> while magnetic resonance (MR) imaging can differentiate these lesions from neoplasms or granulomatous lesions.13,20>

Here, we report a patient with a cryptic arteriovenous malformation (AVM) of the choroid plexus of the fourth ventricle, manifesting only as a hematoma. Choroid plexus vascular malformations are reviewed, and the differences between these lesions and intraparenchymal vascular lesions and the limitation of MR imaging in diagnosing small or occult vascular malformations discussed.

Case Report

A 54-year-old female was admitted to our institution on May 15, 1991 because of dizziness and diplopia. She had been well until 2 months prior to admission when she vomited several times while at work. Subsequently, she began to feel dizzy, and developed headache and diplopia. She had experienced a similar episode in her twenties.

Fig. 1 Postcontrast CT scan, showing a small enhanced lesion (arrow) close to the fourth ventricle.
On admission, she was conscious. Neurological examination revealed right abducens nerve paresis, slight right cerebellar sign, horizontal nystagmus, and gait disturbance. Physical examination revealed no abnormalities. Postcontrast CT scans disclosed an enhanced lesion near the fourth ventricle (Fig. 1). Bilateral vertebral angiograms showed no abnormal vessels in the arterial phase and no staining in the venous phase (Fig. 2). T₁-weighted MR images demonstrated a high-intensity mass lesion, not enhanced postcontrast, adjacent to the fourth ventricle. T₂-weighted MR images showed a high-intensity area with a low-intensity core and prominent low-intensity rim. MR imaging also showed atrophy of the right cerebellum (Fig. 3).

On June 17, 1991, she underwent a suboccipital craniectomy in the prone position. A hard vascular tissue close to the old hematoma was identified through the foramen of Magendie in the right posterior brainstem (Fig. 4). This mass was removed completely.

Histological examination of the resected specimen revealed a choroid plexus with numerous abnormal blood vessels consisting of an arterial component with muscular and internal elastic lamina and a venous component with hyalinized walls (Fig. 5).

Fig. 2  Right vertebral angiograms, showing no abnormal vessels in the arterial phase (left: Towne view, center: lateral view) and no abnormal staining in the venous phase (right).

Fig. 3  MR images at the level of the pons. The T₁-weighted axial images reveal a high-intensity area in the brainstem near the right middle cerebellar peduncle (left) and no enhancement following gadolinium-diethylene-triamine penta-acetic acid administration (center). The T₂-weighted image reveals a high-intensity area with a central low intensity and surrounding prominent low-intensity rim (right). The right cerebellum is atrophic.
The histological diagnosis was AVM of the choroid plexus.

The postoperative clinical course was uneventful except for residual nystagmus, which soon disappeared after discharge.

Discussion

Table 1 summarizes the 41 reported cases of vascular malformations of the choroid plexus.1-8,10,12,14-19,23-28,30 The locations of these vascular malformations were the lateral ventricle(s) in 34 cases (83%), the third ventricle in five (12%), and the fourth ventricle in two (5%), including our case. There were 25 patients (61%) with AVM(s), four (10%) with cavernous angioma, three (7%) with venous angioma, one (2%) with capillary telangiectasia, and eight (20%) with unknown histology.

Thirty-two of the 41 patients (78%) developed intraventricular or subarachnoid hemorrhage, and only three presented with seizure or syncope as the initial symptom. The incidence and site of the hemorrhage contrast with those due to vascular malformations occurring in the cerebral parenchyma, where hemorrhage occurs less frequently and is primarily parenchymatous with infrequent ventricular penetration.

These vascular malformations are reported to become symptomatic in young adults and occur more often in females.7,10 However, our review recognized no female preponderance (20 males and 21 females) or age tendency (range 0-75 yrs, mean 32.8 yrs). Nineteen patients were 40 years old or over, while only 15 were 20 years old or under, indicating that vascular malformations of the choroid plexus may be a cause of intraventricular hemorrhage in adult patients with no aneurysm or thalamic hemorrhage.

Vascular malformations of the choroid plexus are extremely rare in the fourth ventricle. Sekiguchi et al.25 reported a patient in whom whole intraventricular hemorrhage caused bilateral gaze nystagmus, and CT scans 21 days after onset demonstrated a small enhanced lesion in the fourth ventricle, shown to be a small AVM by vertebral angiography. Our patient developed diplopia, dizziness, right cerebellar signs, and horizontal gaze nystagmus. Such clinical characteristics may indicate this lesion in cases of primary intraventricular hemorrhage.

In our patient, angiography showed no vascular staining, although MR imaging demonstrated a
small round mass in the pons which was an old hematoma due to bleeding of the AVM. The AVM was angiographically occult, probably because it was simply too small to detect or may have been destroyed by hemorrhage or thrombosis of the involved vessels after bleeding.\(^{23}\) Angiography showed no evidence of vascular malformation in 11 of 31 (35\%) previous cases with vascular malformation of the choroid plexus. In addition, six of 20 (30\%) AVMs of the choroid plexus were angiographically occult.

CT is better at detecting small vascular malformations of the choroid plexus than angiography.\(^{1,8,18,25}\) In the 41 reported cases, CT detected 74\% (17/23 cases) and angiography 65\% (20/31 cases). Moreover, eight of nine cases with angiographically occult vascular malformation had positive CT findings. Miyasaka et al.\(^{18}\) considered that CT was more useful for 1) precise determination of the anatomical location of the lesion, 2) detection of angiographically occult vascular malformations including cavernous or venous angioma, and 3)
recognition of the extent of the hematoma or the degree of hydrocephalus accompanying the small vascular malformation. Postcontrast CT showed a small enhanced lesion in our patient. CT is therefore very useful for detection of vascular malformations of the choroid plexus in patients with intraventricular hemorrhage when angiography fails to reveal the responsible lesion.

MR images of the intraparenchymal venous or cavernous angioma have been reported, but not those of vascular malformations of the choroid plexus. In our patient, MR imaging demonstrated only a small hematoma in the pons. The T1-weighted image showed a high-intensity area and the T2-weighted image showed a mixed intensity area with a prominent low-intensity rim. Based on these findings, cavernous angioma was diagnosed preoperatively. However, histological examination of the surgical sample discovered an AVM of the choroid plexus. The mistaken diagnosis was caused by the absence of the characteristic flow signal void demonstrated by AVMs, and the strong MR imaging indications of a cavernous angioma. Thus, MR imaging may be useful for identifying the anatomical location of the secondary lesion but may not always yield qualitative information about small or angiographically occult vascular malformations.

Vascular malformations of the choroid plexus are generally small enough to be removed safely by microsurgical techniques. However, a correct preoperative diagnosis is important. Nine of the 35 hemorrhaging cases (26%) including our patient had recurrent hemorrhage, ranging from mild to severe with an overall mortality of 11% (4/35 cases). Patients with primary intraventricular hemorrhage of unknown origin present infrequently in spite of repeated angiography. The possibility of a vascular malformation of the choroid plexus should be considered, and postcontrast CT or MR imaging should be repeated after the intraventricular blood disappears.

References


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