Massive Subependymal Hemorrhage Caused by an Occult Vascular Malformation

—Two Case Reports—

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Abstract

Two patients presented with massive subependymal hemorrhage caused by vascular anomalies occult to angiography, computed tomography (CT) scanning, and magnetic resonance (MR) imaging. A 47-year-old male initially presented with a headache and became comatose 3 weeks later. CT and MR imaging demonstrated a massive hematoma in the right caudate head projecting into the lateral ventricle. A 60-year-old male sustained heaviness of his left extremities 8 days prior to admission and his condition gradually deteriorated. Neuroimaging revealed a right thalamic hematoma adjacent to the lateral ventricle. Cerebral angiography, CT, and MR imaging failed to detect any vascular anomaly. The hematomas increased in size gradually and were surgically explored. The histological diagnosis was arteriovenous malformation for the first case and cavernous angioma for the second case that had ruptured repeatedly. These cases were unique in the unusual location in the subependyma, and diagnostic neuroimaging modalities could not disclose the vascular anomalies, and the hematomas enlarged progressively to become critically symptomatic. Surgical intervention is mandatory for mass reduction and correct diagnosis of such lesions, with favorable outcome as long as the surgery is not delayed until too late.

Key words: arteriovenous malformation, cavernous angioma, cerebral hemorrhage, magnetic resonance imaging, angiographically occult vascular malformation, subependyma

Introduction

Intracerebral hematomas due to angiographically occult vascular malformations are frequently reported, mainly located in the cerebral hemispheres,^{1-3,10,12,18,19,22)} and rarely in periventricular areas. Lobar hemorrhage is sometimes associated with angiographically occult vascular malformations.²¹⁾ Computed tomography (CT) scanning and magnetic resonance (MR) imaging can identify these lesions with improved accuracy in spite of negative findings by cerebrovascular angiography.^{11,17)} However, occult vascular malformations may not be detected by any neuroimaging modality.

We report two cases of massive subependymal hematoma due to occult vascular malformation located in the paraventricular area and projecting

into the lateral ventricles without intraventricular rupture. Cerebral angiography, CT, and MR imaging failed to disclose the coexistent vascular anomalies. Both patients presented with progressive deterioration of consciousness due to gradual and progressive enlargement of the subependymal hematoma. Surgical exploration proved the existence of these vascular malformations.

Case Reports

Case 1: A 47-year-old male suffered from gradually worsening headache for 3 weeks. On admission to the Kitano Medical Research Institute and Hospital, he was alert and had no nuchal rigidity. No neurological abnormalities were detected except for severe headache. He was normotensive and had no history of diabetes mellitus.

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Fig. 1 Case 1. Computed tomography scans on admission (left) and 16 hours later (right), showing progressive increase in the size of a hematoma in the right caudate head projecting into the lateral ventricle.

CT on admission revealed a 3 cm diameter hematoma in the right caudate head projecting into the lateral ventricle without intraventricular rupture (Fig. 1 left). He gradually deteriorated to score 11 on the Glasgow Coma Scale (GCS) or score 10 on the Japan Coma Scale (JCS). MR imaging 10 hours after admission showed an enlarged heterogeneous hematoma of 4 cm in diameter (Fig. 2). No abnormal flow voids or abnormal enhancement were observed with intravenous gadolinium injection. Digital subtraction angiography (DSA) visualized no abnormal vessels, but stretched arteries indicated progressive hydrocephalus (Fig. 3). His level of con-

sciousness continued to deteriorate to GCS score 7 or JCS score 200, and he manifested Cheyne-Stokes respiration. CT just before surgery, 16 hours after admission, showed further enlargement of the hematoma, to 5 cm in diameter, and obvious obstructive hydrocephalus (Fig. 1 right).

The caudate head was exposed through the middle frontal gyrus to visualize the hematoma just beneath the ependyma. The clot was carefully removed so no abnormal vessels or tissue were overlooked. After removal of most of the hematoma, a tiny tangle of arterioles was identified. In addition, one of the caudate veins appeared arterialized. These abnormal vessels were difficult to coagulate by bipolar cautery until a larger feeding artery from the deeper brain tissue had been coagulated. Histological examination verified arteriovenous malformation (AVM) (Fig. 4). Hemosiderin deposition in addition to fresh bleeding indicated repeated hemorrhage.

The postoperative course was uneventful. Although his intellectual ability was subnormal for several weeks after the operation, he recovered completely and went back to his previous work as a TV producer without neurological deficit 3 months later.

Case 2: A 60-year-old male was admitted to our hospital complaining of headache, nausea, and vomiting. He sustained heaviness of the left extremities 8 days prior to admission. He had undergone removal of a right convexity meningioma, of the angiomatous subtype, 13 years previously. He was alert but manifested left hemiparesis. Nuchal rigidity was not present.

Fig. 2 Case 1. Magnetic resonance images 10 hours later, T₁-weighted images without and with gadolinium enhancement (left and center), and T₂-weighted image (right), showing mixed intensity, indicating repeated hemorrhage and no abnormal flow voids. Intravenous gadolinium administration revealed no abnormal enhancement.

Fig. 3 Case 1. Digital subtraction angiograms, showing stretching and unrolling of the anterior cerebral artery indicating hydrocephalus, but no abnormal vessels.

Fig. 4 Case 1. Photomicrograph, showing clusters of abnormal arteries and veins in the hematoma. HE stain, original magnification $\times 10$.

CT on admission revealed a hematoma, 3 cm in diameter, in the right thalamus involving the internal capsule and adjacent to the lateral ventricle (Fig. 5 left). MR imaging on the next day disclosed a heterogeneous hematoma associated with peripheral edema (Fig. 6). There was no abnormal enhancement or abnormal flow voids suggestive of a vascular anomaly or tumor. DSA demonstrated no vascular anomaly or tumor stain (Fig. 7). His level of consciousness gradually deteriorated to GCS score 9 or JCS score 20, and a focal seizure that was restricted to his right leg suddenly began and continued for half a minute. Repeat CT 42 hours after admission showed the hematoma had enlarged to 5 cm in diameter (Fig. 5 right).

He underwent evacuation of the hematoma through the right sylvian fissure. Careful examina-

Fig. 5 Case 2. Computed tomography scans on admission (left) and 42 hours later (right), showing a progressively enlarging hematoma in the right thalamus just beneath the ependyma.

tion of the hematoma revealed a small bundle-like substance in the clot. Histological examination identified abnormal vessels with irregular and nonuniform walls that was compatible with cavernous angioma, in addition to old and fresh hematomas (Fig. 8). No tumor tissue was detected.

The patient became alert after the operation but needed rehabilitation for his left hemiparesis.

Discussion

The term angiographically occult vascular malformation is applied to a group of intracranial vascular hamartomas that cannot be visualized by serial cerebral angiography.^{11,12,20,22} Angiographically occult vascular malformation includes AVM, cavernous angioma, venous angioma, capillary telangiectasia, and mixed malformations.^{4,12,15,20} Clinical manifestations and outcome are not significantly different between these histological subtypes.^{12,13,20} Widespread availability of MR imaging has identified these angiographically occult lesions with increased frequency and accuracy. Accumulation of data on a large number of angiographically occult vascular malformations since the introduction of MR imaging has emphasized the predominance of cavernous malformations.^{9,13,20}

Hemorrhage due to angiographically occult vascular malformations usually occurs in the cerebral hemispheres.^{1,9,12,18,22} Several cases of angiographically occult vascular malformations have been located in the basal ganglia or paraventricular sites.^{1,2,7,10,12,19,23} However, life-threatening hemorrhage that requires emergent surgery, as in our cases, is unusual.²³ Six of 55 patients with angiographically occult vascular malformations

Fig. 6 Case 2. Magnetic resonance images 24 hours later, T₁-weighted images without and with gadolinium enhancement (left and center), and T₂-weighted image (right), showing a developing right thalamic hematoma adjacent to the lateral ventricle with signs of old and fresh bleeding. Peripheral edema surrounding the hematoma is notable but there are no abnormal enhancement or flow voids suggesting vascular anomaly.

Fig. 7 Case 2. Digital subtraction angiograms, showing no vascular anomaly.

Fig. 8 Case 2. Photomicrograph, showing irregular vessels with thickened walls, but no arterial component, in the hematoma. HE stain, original magnification $\times 10$.

suffered massive bleeding, two of which had to be treated on an emergency basis.¹³⁾

Subependyma is the most common site of intracranial hemorrhage in the premature infant and usually spreads throughout the ventricular system. The origin of the hemorrhage is the germinal matrix immediately ventrolateral to the lateral ventricle. A study of the microvasculature demonstrated that penetrating and medullary branches of the main cerebral arteries are distributed in the germinal matrix, forming a fine network of anastomosing arterioles. Many venules in the germinal matrix also converge at a few points and drain into subependymal veins.⁸⁾ This immature vascular rete is postulated to rupture in overloaded conditions, leading to germinal matrix hemorrhage in the premature infant. Vascular anomalies in subependyma, causative

lesions of adult subependymal hemorrhage, may be derived from the immature vascular rete in the germinal matrix.

Angiographically occult AVMs consist of two main types: lesions with small overall size and lesions with slow blood flow.¹⁸⁾ Small vascular malformations are associated with intracranial hemorrhage.¹⁴⁾ The term "cryptic" was proposed for small vascular anomalies that manifested as brain hemorrhage, emphasizing the latent clinical character and their pathological features.^{5,16)} In contrast, thrombosed AVMs are identified by MR imaging as a defined area of increased intensity, either homogeneous or mottled with patchy or homogene-

ous enhancement.⁶⁾

The two present cases were unique because of both the unusual location and the failure of neurological imaging to demonstrate the presence of the associated vascular anomalies. When paraventricular vascular anomalies rupture, the hemorrhage is quite likely to spread into the ventricles. The massive hemorrhage restricted beneath the ependyma may have compressed the small caliber of the component vessels of the AVMs, resulting in the occult appearance on angiography and MR imaging. The heterogeneous intensity on the MR images and the mixture of different stages of red blood cell degradation indicated repeated hemorrhage. Hemosiderin and recent hemorrhage were verified by intraoperative and histological findings. No abnormal enhancement or abnormal flow voids suggestive of vascular anomaly were detected. The vascular anomalies were finally identified during surgery.

The initial symptom was subtle in our cases, but gradual or stepwise worsening of the neurological symptoms in the subacute course was characteristic. Repeated hemorrhage and obstructive hydrocephalus are reflected by decreasing level of consciousness. Case 1 manifested symptoms associated with increased intracranial pressure due to hydrocephalus; and Case 2, in whom the hematoma involved the right internal capsule, manifested symptoms associated with left hemiparesis.

In addition to hematoma evacuation, thorough exploration of the hematoma cavity is essential to detect the source of hemorrhage. The transcortical intraventricular approach is safe and useful for hematoma associated with hydrocephalus. The trans-sylvian approach is alternative, as used in the second case in which the hematoma extended to the internal capsule. We do not know the exact incidence and etiology of hematoma located in subependyma. The neurosurgeon must bear in mind that a vascular anomaly is a possible causative lesion in a case of subependymal hematoma.

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