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Clinical Study

Intracerebral hemorrhage due to developmental venous anomalies

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ABSTRACT

Developmental venous anomalies (DVA) and cavernous malformations (CM) are a common form of mixed vascular malformation. The relationship between DVA, CM and hemorrhage is complicated. It is important to differentiate hemorrhagic CM and hemorrhagic DVA. A retrospective review of all patients with acute spontaneous intracerebral hemorrhages (ICH) between 1 May 2008 and 1 May 2013 was performed. ICH due to DVA or CM were identified and compared for demographic features, clinical symptoms, neurological deficits, and radiological findings. A total of 1706 patients with acute spontaneous ICH were admitted to our hospital during the study period. Among these, 10 (0.59%) were caused by DVA and 42 (2.47%) were caused by CM. No significant differences were found in age ($p = 0.252$) or sex ratio ($p = 1.000$) between the two groups. Compared with CM-induced ICH, DVA-induced ICH were characterized by cerebellar predominance ($p = 0.000$) and less severe neurological deficits ($p = 0.008$). Infratentorial hemorrhagic DVA are characterized by cerebellar predominance and benign clinical course. Infratentorial hemorrhagic CM are mainly located in the brainstem. DVA should be given suspected rather than CM when considering the etiology of a cerebellar hemorrhage, especially in young adults.

1. Introduction

Developmental venous anomalies (DVA) and cavernous malformations (CM) are a common form of mixed vascular malformations. The relationship between DVA, CM and hemorrhage is complicated. DVA are asymptomatic in most cases [1]. On MRI, hemorrhagic lesions with heterogeneous signal intensity and irregular appearance around a DVA are usually diagnosed as CM or chronic hemorrhage due to CM [2,3]. The etiology of an acute intracerebral hemorrhage (ICH) is difficult to determine when both a DVA and a CM are nearby. As a result, acute ICH due to DVA is rarely diagnosed [4]. In this study, we retrospectively reviewed a series of ICH caused by DVA and compared them with ICH caused by CM.

2. Methods and patients

A retrospective review of all medical records from our institution with a diagnosis of acute spontaneous ICH between 1 May 2008 and 1 May 2013 was performed by two neurologists. The diagnosis of symptomatic intracranial hemorrhage was confirmed by a clinical history of acute neurological deterioration and radiological evidence of associated acute hemorrhage. Based on medical history and laboratory, radiographic, angiographic and/or pathological findings, patients diagnosed with DVA-induced ICH or CM-induced ICH were identified. Their imaging was further reviewed by a radiologist. The diagnosis of DVA was established by typical MRI appearance, which included stellate or linear vascular lesions converging onto a collecting vein with a caput medusae-like appearance after enhancement [1]. The hemorrhage was attributed to a DVA if it was located in the venous drainage of the DVA. CM consisted of hyperintense and hypointense lesions or had a core with reticulated signal intensity surrounded by a hypointense rim on conventional T2-weighted images [5]. When both a DVA and a CM were near an ICH, the etiology of the hematoma was difficult to determine. Therefore, ICH accompanied by both DVA and CM were excluded from this study. We also examined the perilesional signal intensity of the hematoma to exclude CM [6]. Finally, patients with DVA-induced hemorrhage were compared to those with CM-induced hemorrhage in terms of demographic features, clinical symptoms, neurological deficits, and radiological findings. The size of the hematoma was determined by measuring the maximum diameter.

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Table 1
Comparison of demographic, clinical, and radiological features in patients with ICH induced by DVA or CM

	DVA-induced ICH	CM-induced ICH	*p* value
Patients	10	42	–
Mean age (years)	31.6	37.3	0.252
Male:Female	1:1	4:3	1.000
Supratentorial ICH	2 (20%)	20 (48%)	0.161
Cortical	1 (10%)	13 (31%)	0.254
Subcortical	1 (10%)	7 (17%)	1.000
Infratentorial ICH	8 (80%)	22 (52%)	0.161
Brainstem	2 (20%)	20 (48%)	0.161
Cerebellum	6 (60%)	2 (5%)	0.000*
Mean size of hematoma (cm)	1.84	2.05	0.445
Main symptom			
Dizziness	7 (70%)	10 (24%)	0.009*
Headache	4 (40%)	14 (33%)	0.723
Epilepsy	0	4 (10%)	0.576
Neurological deficit			
Cranial nerve palsy	4 (40%)	19 (45.2%)	1.000
Limb weakness	0	7 (17%)	0.322
Ataxia/Gait disturbance	6 (60%)	1 (2%)	0.000*
NIHSS score	0.90 ± 0.88	2.07 ± 1.98	0.008*

* $p < 0.05$.
CM = cavernous malformation, DVA = developmental venous anomaly, ICH = intracerebral hemorrhage, NIHSS = National Institutes of Health Stroke Scale.

2.1. Statistical analysis

The Statistical Package for the Social Sciences version 18.0 software was used to perform all statistical comparisons (IBM, Armonk, NY, USA). Categorical variables were compared with Fisher's exact test, and continuous variables were compared with one-way analysis of variance. Differences with a p value of less than 0.05 were considered statistically significant.

3. Results

From 1 May 2008 to 1 May 2013, a total of 1706 patients with acute spontaneous ICH were admitted to our hospital. In 42 cases, the ICH was caused by CM, and in 10 cases, the ICH was caused by DVA. Five ICH accompanied by both DVA and CM were excluded from this study. All 52 patients underwent either CT angiography or digital subtraction angiography to rule out arteriovenous malformations (AVM) or aneurysms. All CM were diagnosed by classical MRI appearance, and in 37 cases, the diagnosis was further confirmed by pathological examination. CM-induced ICH accounted for 2.47% of all ICH during this period, while DVA-induced ICH accounted for only 0.59%.

In the 10 patients (five women, five men) who were diagnosed with DVA-induced ICH, the age ranged from 22 to 51 years (mean age 31.6 years). No subarachnoid hemorrhages were observed. The clinical symptoms included dizziness (n = 7), ataxia (n = 6), and headache (n = 4). The National Institutes of Health Stroke Scale (NIHSS) scores ranged from 0 to 3, with a mean score of ≈1. The hematoma was infratentorial in eight patients (brainstem, n = 2; cerebellum, n = 6) and supratentorial in two patients (frontal horn of the lateral ventricle, n = 1; frontal lobe, n = 1). The mean size of the hematoma was evaluated by measuring the maximum diameter (Table 1). After contrast-enhancement, 12 DVA were found, 10 of which were hemorrhagic and located very close to the corresponding hematomas; the other two asymptomatic DVA were located in the frontal lobe (n = 1) and corona radiate (n = 1). The classical fan-shaped caput medusae was apparent in all cases, with the multiple vessels appearing as linear, curvilinear, or dot-like structures depending on the orientation of the vessel with respect to the plane of imaging. No associated CM were found. In every case, a close relationship between the DVA and the hematoma

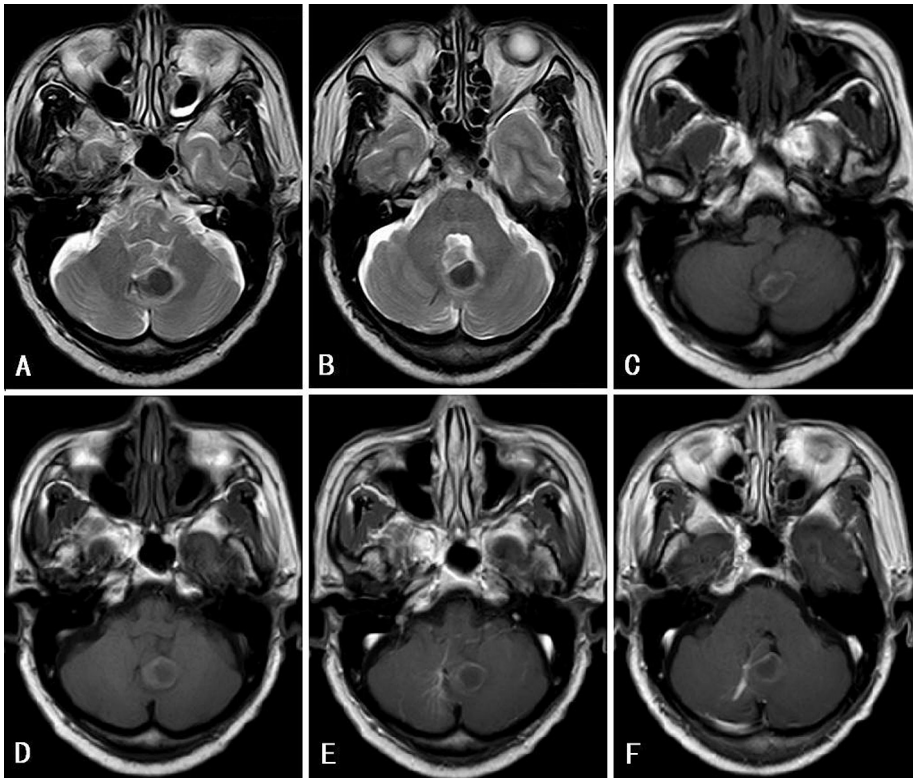

Fig. 1. Close relationship between the hematoma and the collecting vein of a developmental venous anomaly (DVA). (A, B) Axial T2-weighted MRI showing a hypointense lesion indicative of a hematoma and several linear flow voids adjacent to the hematoma. (C, D) Axial T1-weighted MRI showing the isointense hematoma, with flow void signal faintly visible. (E, F) Axial T1-weighted MRI with contrast enhancement clearly showing the caput-medusae-like appearance of the DVA. The hematoma is located within the territory of the venous drainage of the collecting vein.