

Fatal Hemorrhage in Cerebral Proliferative Angiopathy

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Summary

Cerebral proliferative angiopathy (CPA) is a rare vascular abnormality with several angiognomorphological features that are distinct from brain arteriovenous malformations (AVMs). The natural history of CPAs indicates a lower risk for hemorrhage compared to brain AVMs. A 62-year-old woman presented with gait instability and dysarthria. MRI and angiography revealed a diffuse vascular network involving the tectum and cerebellar vermis with intermingled brain parenchyma. This lesion had no dominant feeder, high-flow arteriovenous shunt, flow-related aneurysm or highly dilated veins on angiogram. These findings were consistent with a diagnosis of CPA. During follow-up, she developed progressive gait instability and eye movement abnormalities, but no remarkable change was detected on the repeated MRI and angiography. Nine years later, she died of mesencephalic hemorrhage originating from the CPA. To the best of our knowledge, this is the first description of a patient with CPA who died as a result of the initial hemorrhage. It is important to recognize that a part of CPAs is aggressive and can be more vulnerable to critical hemorrhage.

Introduction

Brain arteriovenous malformation (AVM) is a vascular abnormality consisting of the feeding arteries, nidus and draining veins¹. Based on the angioarchitecture, a rare entity called cerebral proliferative angiopathy (CPA) was suggested and distinguished from “classical” brain AVM². According to the previous report, the natural history of CPA differs from that of

brain AVMs in that patients with CPA have lower risk of hemorrhage than those with brain AVMs². Here, we describe a case of CPA in the tectum and cerebellar vermis which caused a lethal mesencephalic hemorrhage. To the best of our knowledge, this is the first case report of an aggressive CPA in which the first hemorrhagic event was fatal.

Case Report

A 62-year-old woman presented with a two-month history of unsteadiness and slightly slurred speech. Neurological examination was unremarkable except for mild truncal ataxia and dysarthria. MRI demonstrated fine flow voids in the right tectum and vermis, and a single large flow void that ran from the fourth ventricle to the vein of Galen cistern (Figure 1A). Gradient echo T2 star imaging showed no evidence of previous hemorrhage. Angiography (Figure 2A-C) revealed a large nidus supplied by the bilateral posterior inferior cerebellar and superior cerebellar arteries. No dominant feeder or high-flow arteriovenous shunt could be identified. Several moderately dilated veins drained from the nidus into the straight and bilateral transverse sinuses. Venous drainage from this angiopathy was seen in the capillary and early venous phase. The dominant draining vein near the junction of the straight sinus was slightly narrow. The veins of the unaffected cerebellar and cerebral hemispheres were tortuous, indicating venous congestion (Figure 1B). Based on these radiological findings, the patient was diagnosed with CPA. During follow-up, her gait disturbance with spasticity gradually deteriorated. She had been followed with MRI every year

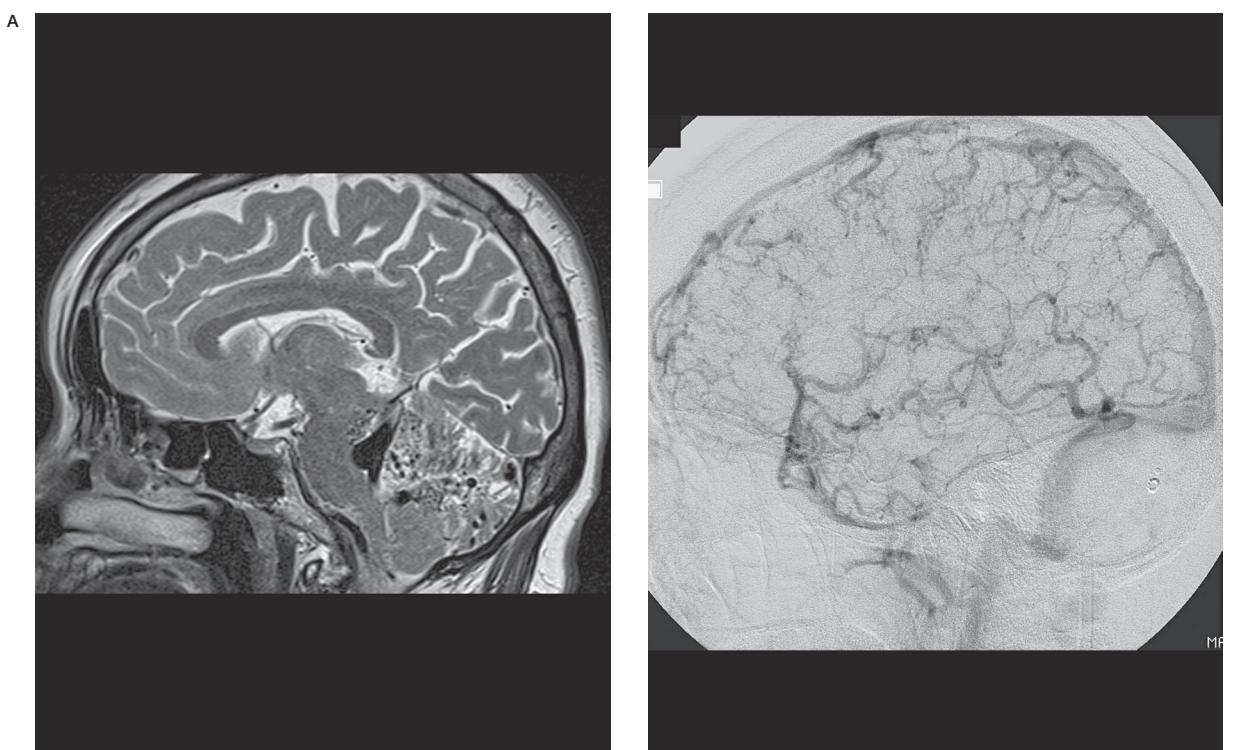


Figure 1 A) MRI sagittal T2-weighted image. MRI demonstrated fine flow voids in the right tectum and superior and inferior vermis. The nidus was not well demarcated. Normal brain parenchyma was interspersed between the abnormal vessels. B) Right internal carotid arteriogram, venous phase. Bilateral internal carotid arteriogram showed the sign of venous congestion of the nonaffected cerebral hemispheres. Most of the cerebral cortical veins were tortuous and venous drainage was delayed.

and catheter angiography was performed three years later (at 65 years of age) and seven years later (at 69 years of age). For these seven years, no marked interval changes were apparent on the serial MRI scans or angiography, but the patient was no longer able to walk by herself.

Nine years after her initial visit to the clinic (71 years old), she suddenly complained of a headache and fell into a comatose state with bilateral extensor plantar responses and episodic decerebrate posturing. The emergency CT showed a massive mesencephalic intraparenchymal hemorrhage extending to the cerebellar peduncle and the quadrigeminal plate with intraventricular and subarachnoid hematoma (Figure 3). During the transfer to our hospital by ambulance, she suffered respiratory arrest and died.

Discussion

Brain AVM is a vascular abnormality in which there are some dominant feeders, an arterialized draining vein due to arteriovenous

shunting, and a nidus without interposed brain tissue¹. According to Lasjaunias et al.², CPA is distinguished from brain AVM based on angiographical features such as a discrepancy between the large size of the nidus and the small shunting volume, the absence of large dominant feeding arteries or flow-related aneurysms, the presence of angioectasia within the nidus and the dural supply, and the slightly early and dilated venous drainage. On MRI, CPA is characterized by a diffuse vascular network with intermingled normal brain tissue. In our case, angiography and MRI findings were consistent with the features detailed above with the exception of the dural supply. The absence of the dural supply appeared to result from the location of the nidus, which was deep-seated and distant from the dura of the cranial convexity.

CPA is a rare entity comprising 3.4% of all brain AVMs and the infratentorial brain is affected in 22% of CPA². Because CPA is relatively rare, its natural course, especially of infratentorial CPA, is still unclear. Lasjaunias et

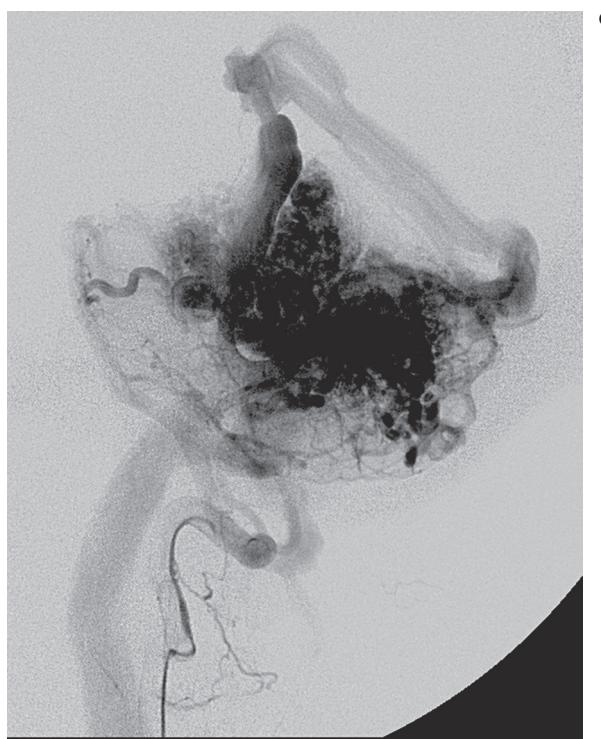
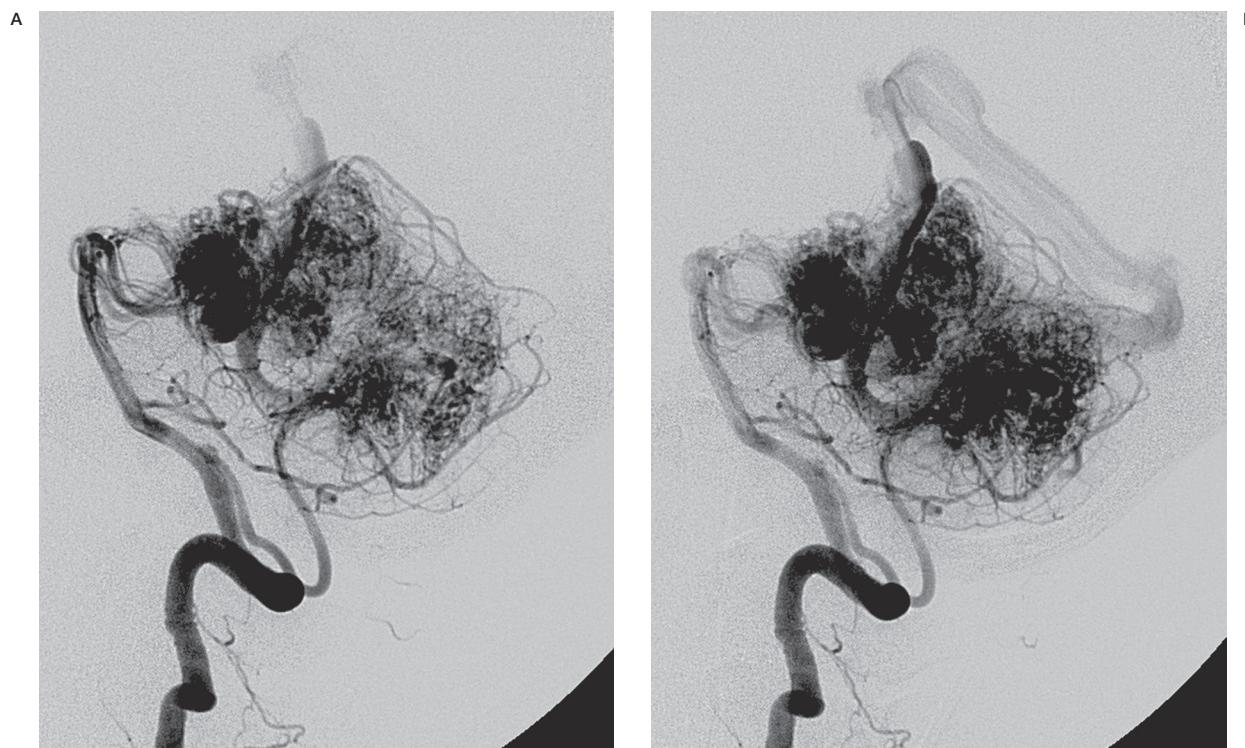


Figure 2 A-C) Left vertebral arteriogram demonstrated the large nidus with capillary ectasias. No dominant or dilated feeder can be identified. The venous drainage of the nidus was seen in the capillary and early venous phase, however no marked high-flow arteriovenous shunt could be identified. The veins of the nonaffected cerebellar hemisphere were tortuous and the contrast medium stagnated, which indicated venous congestion.



Figure 3 Nine years after her first visit, the patient suddenly went into a coma. CT revealed massive intraparenchymal hemorrhage in the mesencephalon with intraventricular and subarachnoid hematoma.

al. reported that the natural course of CPA was less aggressive than that of brain AVM². The most common symptoms of CPA were seizures and disabling headache. Other neurological deficits were present in 16% of patients. The authors reported that six patients (12%) out of 49 had hemorrhagic events and, of those six patients, four (67%) had recurrent hemorrhages. One patient died of a recurrent hemorrhage. Unfortunately, the annual rate of hemorrhage was not made clear. The initial clinical manifestations of our case were truncal ataxia and dysarthria. These symptoms deteriorated gradually over time. Although we could not find any interval changes on the follow-up radiographical examinations, we believe the ischemia around the nidus of the CPA gradually worsened, which resulted in the aggravation of her symptoms. There was the possibility that venous congestion also contributed to the progressive symptoms. Finally, she died as a result of the first hemorrhage she suffered in the mesencephalon and the tectal region. The severity of this hemorrhage can be explained by the anatomical location of the lesion.

Treatment for brain AVM includes microsurgery^{3,4}, radiosurgery⁴⁻⁶ and embolization^{4,6,7}, and is indicated on the basis of clinical classification^{8,9}. On the other hand, treatment for CPA

is limited in cases with hemorrhage or severe symptoms because it carries the risk of damaging the normal brain tissue intermingled in the nidus¹⁰. If the fragile part of the angiopathy could be identified, target embolization may be the most effective way to control the symptoms and reduce the risk of hemorrhage². In the present case, the hemorrhagic location seemed to be near the nidus and the draining vein because the hematoma occupied and expanded the forth ventricle and aqueduct. Indeed it is possible that frequent follow-up will detect some angiographical change in the angioarchitecture in some CPA cases, but, in spite of the repeated MRI and angiography, any changes in the size of the feeders, nidus, or the draining veins could not be identified for seven years in this particular case. Any intranidal aneurysm was not detected, either. Therefore, neither embolization nor radiosurgery was indicated.

Although the risk of hemorrhage in CPA is considered low at presentation, there is a potential risk of critical hemorrhage. The prognosis and natural history may depend on the location of the fragile component of the lesion. Therefore, the proper interpretation of angiographic features and analysis of the angioarchitecture are necessary for the management of this pathology.

References

- 1 Geibprasert S, Pongpech S, Jiarakongmun P, et al. Radiologic assessment of brain arteriovenous malformations: what clinicians need to know. *Radiographics*. 2010; 30: 483-501.
- 2 Lasjaunias PL, Landrieu P, Rodesch G, et al. Cerebral proliferative angiopathy: clinical and angiographic description of an entity different from cerebral AVMs. *Stroke*. 2008; 39: 878-885.
- 3 Hashimoto N, Nozaki K, Takagi Y, et al. Surgery of cerebral arteriovenous malformations. *Neurosurgery*. 2007; 61: 375-387.
- 4 van Beijnum J, van der Worp HB, Buis DR, et al. Treatment of brain arteriovenous malformations: a systematic review and meta-analysis. *JAMA*. 2011; 306: 2011-2019.
- 5 Pollock BE, Meyer FB. Radiosurgery for arteriovenous malformations. *J Neurosurg*. 2004; 101: 390-392.
- 6 Yashar P, Amar AP, Giannotta SL, et al. Cerebral arteriovenous malformations: issues of the interplay between stereotactic radiosurgery and endovascular surgical therapy. *World Neurosurg*. 2011; 75: 638-647.
- 7 Krings T, Hans FJ, Geibprasert S, et al. Partial “targeted” embolisation of brain arteriovenous malformations. *Eur Radiol*. 2010; 20: 2723-2731.
- 8 Spetzler RF, Martin NA. A proposed grading system for arteriovenous malformations. *J Neurosurg*. 1986; 65: 476-483.
- 9 Pollock BE, Flickinger JC. A proposed radiosurgery-based grading system for arteriovenous malformations. *J Neurosurg*. 2002; 96: 79-85.
- 10 Chin LS, Raffel C, Gonzalez-Gomez I, et al. Diffuse arteriovenous malformations: a clinical, radiological, and pathological description. *Neurosurgery*. 1992; 31: 863-868.

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