A rare case of cerebral proliferative angiopathy with bihemispheric morphology

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Cerebral proliferative angiopathy (CPA) is a rare type of cerebral vascular malformation, characterized by multiple, minimally enlarged arterial feeders supplying a lobar or hemispheric angiomatosis.^{1–3} Draining veins are dispersed throughout normal-appearing brain and demonstrate angiographic evidence of capillary angioectasia.^{1–4} We describe a case of bihemispheric, supratentorial CPA involvement. This case emphasizes the importance of identifying CPA as an entity separate from conventional arteriovenous malformations (AVM) or arteriovenous fistulae (AVF) as the natural history is dissimilar and treatment approaches vastly differ.^{2,4}

A 39-year-old woman presented to the emergency department with a history of recurrent headaches and nasal discharge. Examination revealed no focal neurologic deficits or cutaneous lesions. Funduscopic examination showed no retinal vascular malformations.

CT angiogram (CTA) head showed bilateral hemispheric vascular malformations with enlargement of feeding arteries and draining veins (figure 1A). MRI head confirmed

Figure 1 CT angiogram and MRI findings

(A) Axial CT angiogram image shows the diffuse nature of the bihemispheric cerebral vascular malformation without a discrete nidus. There is prominence of the supplying arteries and draining veins. (B) Axial T2-weighted MRI shows normal signal and morphology of the cerebral parenchyma dispersed between the abnormal vessels. (C) Coronal T2-weighted fat-saturated MRI shows the right nasal cephalocele.

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Figure 2 Cerebral catheter angiogram findings

(A), (B), and (C) Right and left internal carotid artery antero-posterior (AP) views show a diffuse network of bihemispheric enhancing vessels without a discrete nidus. (D) and (E) Right internal carotid artery AP and lateral view (delayed to venous phase) demonstrate a sinus pericranii. Although prominent, the extensive cerebral and transcranial venous drainage is not proportionately enlarged compared to the size of the malformation. (F) and (G) AP and lateral views of the left vertebral artery show mild enlargement of the arteries supplying the angiomas with no dominant feeding vessels. (H) Right external carotid artery lateral view shows 3 regions of transcranial supply from the right middle meningeal artery, marked with asterisks.

normal cerebral parenchyma dispersed within the angiomas (figure 1B). A right nasal cephalocele was noted (figure 1C). There were no acute infarcts or intracranial hemorrhages. Catheter cerebral angiogram demonstrated a diffuse network of bihemispheric vessels without a discrete nidus (figure 2, A–C). The supplying internal carotid and vertebral arteries were mildly enlarged (figure 2, A–C, F, and G). Additional transcranial feeding arteries originating from the right middle meningeal artery were noted (figure 2H). Despite extensive cerebral and transcranial venous drainage, no venous outflow obstruction or disproportionally enlarged draining veins were demonstrated (figure 2, D and E).

DISCUSSION

CPA is a rare type of vascular malformation. A large case series of 1,434 patients with arteriovenous malformation (AVM) showed a 3.4% incidence² and a smaller preceding AVM series of 180 patients showed a 6.7% incidence.⁴ In addition to ours, we identified one published case of a patient with bihemispheric supratentorial CPA involvement.⁵

Demographically, CPA differs from AVM, affecting young to middle-aged adult females.^{2,4} Patients present with headaches, seizures, or stroke-like episodes. Although the incidence of CPA-related hemorrhage is lower compared to conventional AVM (12% vs 42%), once a hemorrhage has occurred, the rebleed rate is much higher (67% in CPA vs 4% in AVM).²

It has been suggested that CPA as a vascular malformation is characterized by both cellular proliferation (as seen in hemangiomas) and endothelial turnover (similar to conventional AVMs).²

Angiographically, CPA is characterized by a diffuse network of densely enhancing vessels intermingled with normal brain parenchyma.^{2,4} There is absence of dominant feeding vessels. Instead, CPA is fed by multiple, minimally enlarged arteries that may demonstrate proximal stenosis.² Dural vessels characteristically also supply the malformation.² There is absence of a discrete nidus and flow-related aneurysms.^{2,4} Although the draining veins are mildly enlarged, there is overall discordance between the expected size of draining veins relative to the size of the malformation, with a lack of early draining veins.² There has been no documented evidence for CPA being secondary to venous occlusive disease.

Moyamoya disease and hereditary hemorrhagic telangiectasia (HHT) are entities to be considered in a differential diagnosis of CPA. Moyamoya disease is a proliferative arteriopathy characterized by extensive tiny basal ganglia perforator vessels and multiple transdural collateral vessels.⁶ However, in contrast to moyamoya disease, CPA does not manifest as occlusive disease of the supraclinoid internal carotid arteries.⁶ HHT, also referred to as Rendu-Osler-Weber syndrome, has an autosomal dominant inheritance, with gene mutations involving endoglin (ENG) or activin receptor-like kinase 1 (ALK1).⁷ HHT shows multiple vascular malformations that have the same angioarchitecture as sporadic AVM.⁷ However, individual cerebral lesions carry a more favorable prognosis compared to sporadic AVMs. HHT is characteristically associated with mucocutaneous telangiectasias and systemic AVMs involving the lungs, spine, liver, and gastrointestinal tract.⁷

Pathologically, CPA differs from conventional AVMs due to the presence of normal-appearing brain tissue and neurons interspersed between the angiomas, implying that aggressive embolization or excision of the vascular malformation would sacrifice the associated cerebral vascular territory.¹⁻⁴ As such, judicious conservative embolization for symptom management is advocated.² Therapeutic burr-hole placement may be useful in cases of severe CPA-related headaches.²

In our case a right nasal cephalocele was part of the presenting findings. Although this may be incidental, altered intracranial vascular/CSF flow dynamics may have predisposed our patient to skull base dehiscence.

It is important to recognize unruptured CPAs as distinct from AVMs because of the apparently lower risk of hemorrhage and risk to interspersed neural tissue. Because of the risk of rebleeding, intervention should be considered once a hemorrhage has occurred.

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