CASE REPORT

Infratentorial hemorrhagic cerebral proliferative angiopathy: A rare presentation of a rare disease

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ABSTRACT

Cerebral proliferative angiopathy (CPA) is a unique and rare vascular malformation with distinct clinico-radiological features. CPA is associated with low risk of hemorrhage as compared to classical cerebral arteriovenous malformations (AVMs). Infratentorial location of diffuse nidus is also an uncommon presentation. Infratentorial hemorrhagic presentation of CPA is a rare co-occurrence. Herein, we report a case of an elderly old male, who presented with acute onset severe headache, recurrent vomiting, vertigo and swaying toward left side while walking. Cerebellar signs in the form of scanning speech, nystagmus, impaired finger-to-nose incoordination test, dysdiadochokinesia, and limb ataxia on the left side were present. Magnetic resonance imaging (MRI) brain revealed diffuse vascular network with intermingled normal brain parenchyma and hemorrhage in the left cerebellum. Digital subtraction angiography (DSA) revealed diffuse, ill-defined, nidus in left posterior fossa involving the left posterior inferior cerebellar, anterior inferior cerebellar and posterior cerebral arterial territories. There were no definite arterial feeders. DSA showed scattered "puddling" appearance of contrast material in the widespread nidus and drained into the multiple ill-defined posterior fossa veins. The MRI brain and DSA findings were consistent with the diagnosis of the CPA. Treatment in our case was limited to supportive medical therapy because selective embolization was not feasible due to nonidentifiable arterial feeders. He was asymptomatic at 1-year follow-up. This case highlights a rare entity called CPAs, which have different clinical presentations, angiographic features, treatment options and patient outcome as compared to classical cerebral AVMs.

Key words: Cerebral proliferative angiopathy, arteriovenous malformations, intracerebral hemorrhage, infratentorial nidus

Introduction

The classical cerebral arteriovenous malformations (AVMs) are dilated tortuous vessel masses having direct connections between arteries and veins without intervening the capillary. They have definite nidus, draining veins and feeding arteries without intervening normal brain parenchyma in between nidus.[1,2] Based on angiographic features, a rare subgroup, different from classical brain AVMs, called cerebral proliferative angiopathy (CPA) was suggested by Lasjaunias *et al*.[3] It is characterized by diffuse, ill-defined nidus, multiple nondominant arterial feeders, small draining veins and capillary angiectasia. There is scattered puddling appearance of contrast material in diffuse nidus.[3] Magnetic resonance imaging (MRI) brain reveals diffuse vascular network with intermingled normal brain parenchyma. CPA is classified as a distinct subgroup of AVM because it significantly differs from classical AVMs in their epidemiology, clinical presentation, disease course, angiographic features and presumed pathophysiology. It has a lower risk of hemorrhage as compared to classical brain AVMs. Right hemisphere was the most common (45%) site for CPA nidus in Lasjaunias *et al*. case series. Twenty-two percent patients had the infratentorial location. Here, we describe a very rare case of infratentorial hemorrhagic CPA.

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Case Report

A 66-year-old male presented with severe headache, recurrent vomiting, vertigo and swaying toward left side while walking for 10 days. There was no history of seizure, loss of consciousness, trauma, hypertension, diabetes or illicit drug abuse. On neurological examination, cerebellar signs in the form of scanning speech, nystagmus, impaired finger-to-nose incoordination test, dysdiadochokinesia, and limb ataxia on the left side were present.

Hemogram, coagulation profile (platelets count, bleeding time, coagulation time, prothrombin time, INR), serum biochemistry including thyroid function tests were normal. Noncontrast computed tomography (CT) scan showed intraparenchymal hemorrhage in the left cerebellar hemisphere [Figure 1]. MRI of brain demonstrated a diffuse network of vascular channels and dilated veins with intermingled normal brain tissues between different vascular territories on the left cerebellar hemisphere, which extend to the left para-pontine, quadrigeminal and arachnoid cistern. A small intraparenchymal hemorrhage was present in posterior aspects of the left cerebellar hemisphere [Figure 2a and b]. CT angiography of brain vessels showed diffuse network of densely enhancing vascular channels in the left posterior fossa. There were no dominant arterial feeders [Figure 3a and b]. Cerebral digital subtraction angiography (DSA) revealed large, ill-defined, diffuse, nidus in the left posterior fossa. Diffuse nidus involved the left posterior inferior cerebellar, anterior inferior cerebellar and posterior cerebral arterial territories. There was no dominant arterial feeder identified. It had scattered "puddling" of contrast material in the widespread nidus and drained into the ill-defined, multiple posterior fossa veins to straight and left transverse sinus [Figure 4a-c]. MRI brain and DSA findings were consistent with the diagnosis of the CPA. The patient was managed with symptomatic medical therapy. He was asymptomatic at 1-year follow-up.

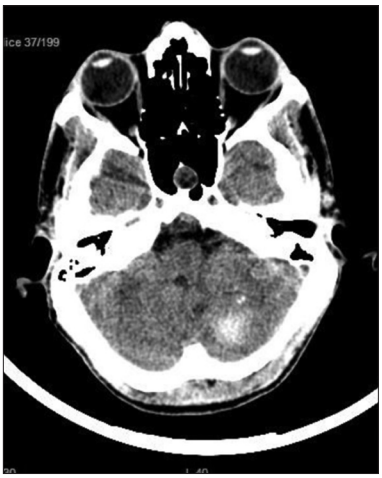

Figure 1: Noncontrast computed tomography scan showing intraparenchymal hemorrhage in the left cerebellar hemisphere

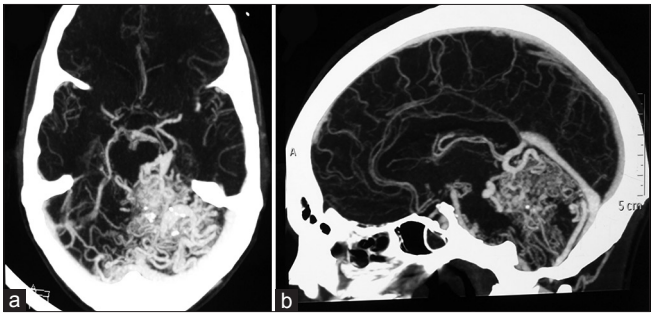

Figure 3: Computed tomography angiography of brain vessels showing diffuse network of densely enhancing vascular channels in the left posterior fossa. There was no dominant arterial feeders (a) axial, (b) sagittal

Discussion

Based on angiographic features, Lasjaunias *et al*. in 1989 first described a peculiar type of diffuse brain AVMs called CPA in 49 patients. CPA is a rare subgroup, encompassing 3.4% of all cerebral AVMs.[3] These abnormal vessels are due to anomalous embryonic development. It appears to be predominant in female (female: male 3:1) and age of presentation may vary from 10 to 65 years. The clinical features of CPA depend on the site of lesion, seizures being the most common (45%), followed by severe headache (41%). Other clinical presentations were

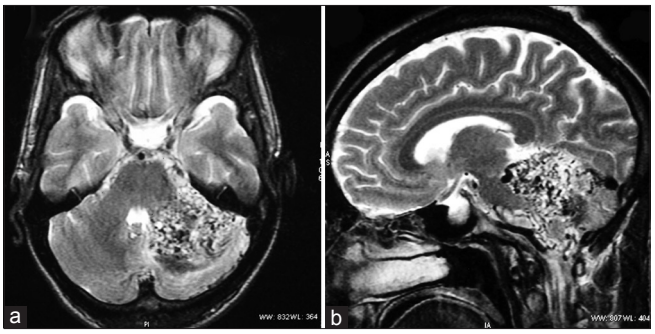

Figure 2: T2-weighted magnetic resonance imaging of brain (a) axial, (b) sagittal showing a diffuse network of vascular channels and dilated veins with intermingled normal brain tissues between different vascular territories on the left cerebellar hemisphere, which extend to the left para-pontine, quadrigeminal and arachnoid cistern. A small intraparenchymal hemorrhage is seen in posterior aspects of the left cerebellar hemisphere

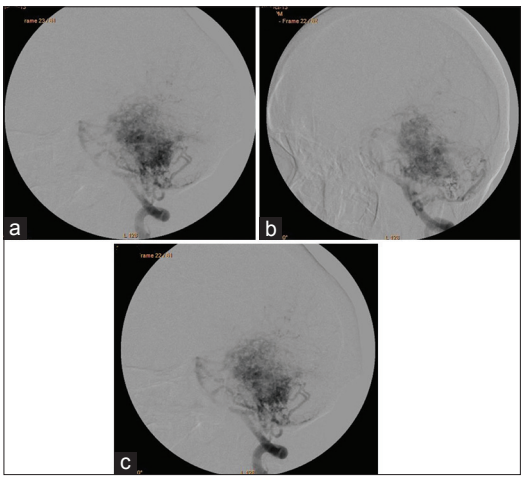

Figure 4: (a-c) Cerebral digital subtraction angiography reveal large, ill-defined, diffuse, nidus in the left posterior fossa. Diffuse nidus involved the left posterior inferior cerebellar, anterior inferior cerebellar and posterior cerebral arterial territories. There was no dominant arterial feeder identified. It had scattered "puddling" of contrast material in the widespread nidus and drained into the ill-defined, multiple posterior fossa veins to straight and left transverse sinus

transitory ischemic attacks, stroke-like symptoms, and focal neurological deficits. Twelve percent patient was presented with intracranial hemorrhage, out of which recurrent hemorrhage were observed in two-third of patients. That suggest a high chance of the recurrent bleed, when patients present with intracranial hemorrhage.

Right hemisphere was the most common (45%) site for CPA nidus in Lasjaunias *et al*. case series. Other common sites were temporal lobe (49%), frontal lobe (41%), parietal lobe (39%) and occipital lobe (24%). Twenty-two percent patients had the infratentorial location of nidus. There had been only a few case report of infratentorial CPA with hemorrhage[4] as observed in our patient, and there was no recurrence of hemorrhage at 1-year follow-up. Infratentorial hemorrhagic presentation of CPA is a rare co-occurrence in our patient.

Special attention should be given for posterior fossa vascular malformations, as these abnormal vascular channels have increased the amount of blood flow, which can cause significant steal phenomenon. Classical posterior fossa AVMs have definite feeding vessels, which are usually unilateral, and aneurysm formation is common due to high-flow feeders. Whereas, CPA has ill-defined, trivial arterial feeder prone for hemorrhage and aneurysm formation is very unlikely due to poor-flow feeders.

Classical cerebral AVMs have various treatment options, including surgical resection, radiosurgery, and embolization.[5,6] However, CPAs have intervening normal brain parenchyma between vascular channels, which makes above mentioned treatment procedure difficult because such patient have risk of developing permanent neurological deficit. Therefore, treatment options for CPA are limited.[7,8] Usually, symptomatic medical therapy are offered to such patient in the form of analgesics and antiepileptic drugs. The various neuro-interventions like microsurgery,[6] arterial endovascular embolization[9] and radiosurgery[10] have also been tried with variable success rates. Selective arterial embolization of feeders can improve severe persistent headache, and if fragile feeder can be identified, this procedure might be the most effective treatment for reducing the risk of hemorrhage. In our patient, fragile arterial feeders could not be identified, so target arterial embolization was not feasible.[3] Posterior fossa CPAs are difficult to manage, as critical vessels that originate in the vertebrobasilar system, ultimately perfuse both the vital structures of posterior fossa and CPA. Therefore, any neuro-intervention (microsurgery, radiosurgery, arterial endovascular embolization) may be fatal. Microsurgical dissection has better access and visualization of feeding vessels, therefore it may avoid the potential life endangering inadvertent brainstem infarction, which occur during endovascular procedures.

If a CPA is misdiagnosed as classic AVM, the results may be lethal, whenever; microsurgery or arterial embolization is undertaken. Therefore, our case highlights the need of proper interpretation of angiographic features of cerebral vascular malformations to identify and separate CPA entity from classical brain AVMs because correct diagnosis is important for process of decision making and outcome of the patients.

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