CASE REPORT

Diffuse proliferative cerebral angiopathy: a case report and literature review on a very rare and misdiagnosed entity

Sagar Panthi1,*, Nimesh Khanal2, Sajana Poudel1, Siddhartha Bhandari3, Pradeep Khatiwada1, Rochana Acharya1, Raksha Bhattacharjya4, Bharosha Bhattacharjya1 and Sandeep Khanal1

1Department of General Surgery, B. P. Koirala Institute of Health Sciences, Dharan, Province 1, Nepal, 2Department of General Surgery, Creasion Nepal, Kathmandu, Bagmati Province, Nepal, 3Department of General Surgery, Tribhuvan University Teaching Hospital, Kathmandu, Bagmati Province, Nepal and 4Department of Radiodiagnosis and Imaging, B. P. Koirala Institute of Health Sciences, Dharan, Province 1, Nepal

*Correspondence address. Department of General Surgery, B. P. Koirala Institute of Health Sciences, Dharan 56700, Nepal. Tel: +977-9867116661; E-mail: drsagarpanthi@gmail.com

Abstract

Diffuse proliferative cerebral angiopathy (DPCA) is an uncommon type of cerebral vascular malformation, mostly diagnosed in young females. It is characteristically different from other cerebral arteriovenous malformations and can be differentiated by its peculiar imaging findings. A nidus of normal brain parenchyma is present between the abnormal vascular channels. Therefore, it is crucial to diagnose it as a separate entity because unnecessary treatment of DPCA increases the risk of damage to the normal parenchyma leading to neurological deficits. Here we describe a case of a 60-year-old male who presented with severe neurological deficits and was later diagnosed with DPCA. He was managed conservatively with antiepileptics and almost completely recovered to normal within 2 weeks. A rare case of DPCA confused with other hemorrhagic disorders is discussed here. Rare cases are often overlooked. Correct diagnosis helps to prevent tragic consequences.

INTRODUCTION

Diffuse proliferative cerebral angiopathy (DPCA) is an uncommon type of cerebral vascular malformation, mostly diagnosed in young females [1, 2]. Patients present with headaches, epileptic features and progressive neurological deficits but rarely with a hemorrhage [2]. It is characteristically different from other cerebral arteriovenous malformations (AVMs) and can be differentiated by its peculiar imaging findings [2]. One critical finding is a nidus of normal brain parenchyma in between the abnormal vascular channels [1, 2]. Therefore, it is crucial to diagnose it as a separate entity because unnecessary treatment of DPCA increases the risk of damage to the normal parenchyma leading to neurological deficits [1, 2].

CASE REPORT

A 60-year-old male presented to our institute in the emergency department with a history of loss of consciousness for 20 minutes followed by aphasia, seizures and left-sided hemiparesis without any prior similar history. He was a known case of
AVMs are of several types—the most common being glomerular and the uncommon being fistulous type [4]. Based on the natural history and pathophysiology, AVMs types are cavernous malformations, venous malformation and capillary telangiectasia [5].

DPCA is common in young females (mean age ∼ 22 years) and is a diagnosis of exclusion [2]. Contrary to this, our patient was male presenting at 60 years of age which is quite more as per the available literature. The exact cause of DPCA is unknown; however, its diffuse character is confirmed by the presence of a trans-dural supply in remote locations (supra and infra-tentorial) suggesting an unpressed response to cerebral sub-ischemic manifestations [2]. DPCA is characterized by the presence of a nidus of normal brain parenchyma composed of multiple arteries as an angiogenetic response to cortical ischemia [1, 2, 4].

Patients with DPCA mostly present with seizures, headaches and progressive neurological symptoms similar to our patient presenting with loss of consciousness followed by aphasia, seizures and left-sided hemiparesis [2]. Hemorrhage occurs rarely; however, if bleeding occurs, then the risk of recurrence is higher. The risk of hemorrhage is negatively affected by the association of arterial stenoses with angiogenesis [2].

The natural history of proliferative cerebral angiopathies indicates a lower risk for hemorrhage compared with other AVMs [6]. It is necessary to diagnose DPCA as it identifies the presence of normal brain tissue intermingled with the vascular spaces, and damage to the structure with an intervention could lead to serious complications [2]. Thus, the natural history and management of each entity are different [4]. Furthermore, its treatment is challenging and best done at centers with expertise [1]. Findings that are consistent with the diagnosis of PCAs on angiography include the absence of dominant feeder, without high flow arteriovenous shunt or early draining veins, highly dilated veins or flow-related aneurysms differentiating it markedly from other AVMs [6].

Plain CT scan findings in our patient mimicking acute SAH could have misled to a wrong diagnosis inviting unnecessary neurosurgical intervention had it not been to the consultation from a radiologist for a CT cerebral angiography. Though DSA is the gold standard for the diagnosis of DPCA, CTA and magnetic resonance angiography (MRA) are quite accurate too in ruling out other AVMs [4, 6].
Classic AVMs require treatment after weighing the disease versus treatment-related risks, whereas DPCA is managed conservatively. A treatment would do more harm than cure a DPCA [4]. Treatment is reserved for those with intractable headaches and epilepsy. Treatment options are surgery, radiosurgery, large non-targeted embolization, targeted embolization, synangiogenesis and calvarial burr holes. Those with hemorrhage are treated with endovascular treatment [1]. Surgical treatment is not indicated unless areas of the angioarchitecture suggest zones of weakness or demonstrate obvious constraints to the eloquent brain. Headaches are treated with arterial embolization in non-eloquent areas without treatment of dura [2].

CONCLUSION

Havoc acknowledged is havoc prevented. Prior suspicion is critical to diagnosis of DPCA and to rule out other types of AVMs. In our case, the presentation was not acute, and surgery could be delayed, so all necessary investigations were done to correctly identify the disease. Thus, unnecessary morbidity or even mortality was prevented.

CONFLICT OF INTEREST STATEMENT

None declared.

FUNDING

None.

REFERENCES

1. Srivastava T, Mathur T, Mittal RS, Raghavendra BS, Jain R, Handa R. Cerebral proliferative angiopathy: a rare case with rare presentation and rarer angiographic features. EJVES Extra 2013;26:e27–9.
2. Lasjaunias PL, Landrieu P, Rodesch G, Alvarez H, Ozanne A, Holmin S, et al. Cerebral proliferative angiopathy: clinical and angiographic description of an entity different from cerebral AVMs. Stroke 2008;39:878–85.
3. Dória-Netto HL, Souza-Filho AM, Dória-Netto RH, Marques RA, Oliveira DA, Chaddad-Neto F, et al. Cerebral proliferative angiopathy. Arq Neuropsiquiatr 2010;68:300–2.
4. Geibprasert S, Pongspech S, Jiarakongmun P, Shroff MM, Armstrong DC, Krings T. Radiologic assessment of brain arteriovenous malformations: what clinicians need to know. Radiographics 2010;30:483–501.
5. Rohit PS. Diffuse proliferative cerebral angiopathy: a case report and review of the literature. J Radiol Case Report 2015;9:1.
6. Maekawa H, Tanaka M, Hadeishi H. Fatal hemorrhage in cerebral proliferative angiopathy. Interv Neuroradiol 2012;18:309–13.