CASE-BASED UPDATE

Epilepsy in a boy with a developmental venous anomaly—case-based update

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Received: 9 August 2014 / Accepted: 17 February 2015 © Springer-Verlag Berlin Heidelberg 2015

Abstract

Background Developmental venous anomalies (DVAs) are the most common intracranial vascular malformations of the brain. They are most commonly asymptomatic and often diagnosed incidentally.

Illustrative case We present a rare case of a 9-year-old boy with seizures caused by a subcortically located DVA posterior to the postcentral gyrus of the left brain hemisphere. MRI also showed an enlarged, microcystically altered pineal gland and an extracranial cavernous hemangioma. EEG showed left fronto-centro-parietal spikes that corresponded with the location of the DVA. The spikes were occasionally secondary generalized. The boy responded well to oxcarbazepine treatment, and no surgical intervention was required. Control MRI scans showed no progression of the DVA.

Conclusion Epilepsy may be caused by DVA. The treatment starts with anticonvulsants. The surgical treatment is the therapeutic choice only if seizures are resistant to conservative therapy because surgery might result in serious ischemic or hemorrhagic complications because DVAs are considered to be variations of normal transmedullary veins that are necessary for the drainage of white and gray matter.

Keywords Epilepsy · Developmental venous anomaly · Venous angioma · Pediatry

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Introduction

Developmental venous anomalies (DVA), formerly known as *venous angiomas*, are clusters of radially aligned veins that converge into a collecting vein, giving them a characteristic appearance on MRI and angiography, which can be described as *caput medusae*. The pathogenesis of this condition is not completely understood, although it is thought to be a disorder of normal embryogenetic processes. Pathohistologically, DVAs are characterized by mature venous walls with minimal smooth muscle and elastin and without arterial or capillary elements [1].

With a more frequent use of MRI, this anomaly has shown to be the most common intracranial vascular malformation, with an incidence between 0.7 and 2.5 % [1, 2], and is in two third of the cases present supratentorially [1, 3].

Clinical presentation

Since DVA is a benign vascular lesion, it is usually asymptomatic and diagnosed incidentally on brain MRI scans or in autopsy [4, 5]. Symptoms are often caused by the coexisting vascular malformations. Cavernous malformations (CMs) are present with DVAs in 8 to 33 % of cases, thus being the most common vascular malformation associated with DVA. Coexisting CMs are often the cause of intracranial hemorrhages (ICH) in patients with a DVA [1, 6].

When symptomatic, the most common symptoms of DVAs include headaches, seizures, and focal neurological deficits due to acute infarction or intracerebral hemorrhages attributed to DVA thrombosis [1, 4, 5]. It is important to have in mind, when classifying a DVA as symptomatic, that associated causes of the symptoms need to be excluded. Possible causes may include cortical dysplasia as a cause of epilepsy and

associated cavernomas as a cause of hemorrhage. Those seizures for which no other causes have been reported are thought to be related to reduced blood outflow from the DVA or to small scale neuronal migration deficits that are undetectable with neuro-radiological methods [2, 4].

Management of DVA

The treatment of symptomatic DVAs is most commonly symptomatic and includes a pharmacological approach to dealing with seizures. It is important to note that DVAs are considered to be variations of normal transmedullary veins that are necessary for the drainage of white and gray matter. Since DVAs are considered to be a variation of the normal venous system, invasive neurosurgical procedures or stereotactic radiosurgery resulting in excision or obliteration of the lesion may lead to serious ischemic or hemorrhagic complications [1, 7]. Because of that, neurosurgical treatment is not the first and the best option for the majority of these patients.

Illustrative case

A 9-year-old boy was admitted to our hospital following a seizure. In the morning, after acquiring little sleep caused by excitement because of the trip he was to attend that morning, while driving in the car, he suddenly complained of being hot, vertiginous, and experiencing a frontal headache. His parents said he was distant, unresponsive, disoriented, and looked to the side for 15 min. The patient experienced no vomiting or foaming of the mouth. He did not recall the drive to the hospital. One month prior to the incident, a similar episode occurred in school.

The family history is negative on epilepsy and other neurological diseases, but a few members from the mother's side of the family had arrhythmias. Perinatal history was uneventful. In the neonatal period, a brain ultrasound showed grades I–II intraventricular hemorrhage. The child's psychomotor development was normal, with mild articulation disorders.

Physical examination at the time of admission showed no pathology. In the neurological examination, the



Fig. 1 EEG showing fronto-centro-parietal spikes on the left hemisphere

Fig. 2 MRI showing DVA. Magnetic resonance of the brain showing DVA



patient was unstable while performing the Romberg's test. Electroencephalogram (EEG) showed left frontocentro-parietal spikes, which occasionally were secondary generalized (Fig. 1).

Brain magnetic resonance imaging (MRI) 3T showed numerous serpiginous hypointensive signals in the superficial cortex posterior to the left postcentral gyrus (Figs. 2, 3, and 4). The findings are consistent not only with an aggregation of blood vessels but also with traces of calcification and superficial hemosiderosis as a sign of previous hemorrhage. Discrete glial changes were present on the surrounding gyruses. The pineal gland was enlarged, dimensions 12×6 mm, microcystically altered, but with no compression to the mesencephalic aqueduct. An extracranial cavernous hemangioma in the left frontal region was also noticed.

Digital subtraction angiography (DSA) showed a developmental venous anomaly with hypertrophic cortical veins forming a *caput medusae* and calcifications. Conclusively, neuroimaging showed a developmental venous anomaly in the left centro-parietal part of the brain as well as an extracranial cavernous hemangioma in the left frontal region.

Transcranial color Doppler (TCD) of the circle of Willis showed an asymmetry with an increased mean flow velocity in the left anterior and posterior cerebral artery. Mixed scotomas



Fig. 3 DSA showing DVA. Digital subtraction angiography of the brain showing DVA



Fig. 4 DSA showing DVA. Digital subtraction angiography of the brain showing DVA

in the visual field were also observed. Visual evoked potentials were normal. Heart ultrasound and Holter 24-h EKG were normal.

In the follow-up, the boy experienced two more seizures with similar characteristics. He has been treated with oxcarbazepine 30 mg/kg. No seizures occurred in the last year. Control MRI scans after a year and 2 years showed no progression of the anomaly.

Discussion

The cases of epilepsy related to DVA were not often described in children. Our patient exhibited seizures which manifested with fronto-centro-parietal spikes on EEG. Focal neuroelectrical surges may be a result of underlying brain malformations. MRI and DSA showed a DVA which may be the cause of seizures because of the position consistent with the abnormal EEG result. The lesion may be associated with a gliotic scar or it may become hemosiderin-laden and may contribute to the etiology of epilepsy [1].

Although the DVAs are rarely symptomatic and are usually an incidental finding, when symptomatic, the most common symptoms are headaches, seizures, and hemorrhages [1]. No other changes, such as brain tumors or focal dysplasia, were detected in the centro-parietal region of our patient. However, a microcystically altered pineal gland, 12×6 mm, was noticed. Cysts in the pineal gland are rarely symptomatic when smaller than 10 mm [8], but those over 10 mm may result in symptoms which may include paroxysmal generalized seizures which manifest as a spike-wave complex of 3 Hz or more [9]. The diagnostical findings in our patient suggest that the DVA is the most probable cause of the seizures.

However, some authors disagree with the idea of DVAs as the reason for epilepsy [10, 11]. Striano et al. found that seizures occur only in a small percentage of patients with DVA. Those having epilepsy present with complex partial seizures, as it was the case with our patient.

DVAs are considered to be a variation of the normal venous system, so the surgical excision is, therefore, a risk factor for the development of serious postoperative complications, including venous infarction and cerebral edema, resulting from the brain being deprived of normal venous drainage [1, 6]. So far, our patient responded well to anticonvulsant treatment.

Conclusion

Epilepsy may be caused by developmental venous anomalies. The treatment of DVA-related epilepsy starts with anticonvulsants. The surgical treatment is the therapeutic choice only if seizures are resistant to conservative therapy because surgery might result in serious ischemic or hemorrhagic complications.

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