JVIN Journal of Vascular and Interventional Neurology
OFFICIAL JOURNAL OF THE ZEENAT QURESHI STROKE INSTITUTE

Complex Partial Epilepsy Associated with Temporal Lobe Developmental Venous Anomaly

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Abstract

Background—Developmental venous anomalies (DVA) are found incidentally but sometimes patients with these anomalies present with varying degrees of neurologic manifestations.

Objective—We report a patient with early onset complex partial epilepsy and associated DVA and discuss the natural history, neuroimaging and clinical characteristics, and management.

Case description—A 21-year-old man presented with a history of complex partial epilepsy with secondary generalization which started at the age of 4 years. An electroencephalogram (EEG) was performed which demonstrated spike and wave discharges predominantly in the left frontotemporal region. A magnetic resonance imaging (MRI) was performed which demonstrated a linear flow void suggestive of a DVA. The angiogram demonstrated DVA that connected with the left transverse venous sinus and an anastomotic vein between the straight sinus and the transverse venous sinus traversing the brain parenchyma. He was started on carbamezipine for the treatment of complex partial seizures.

Conclusions—Temporal lobe DVA may be associated with complex partial seizures and can be diagnosed by MRI and angiographic findings.

Keywords

Complex partial epilepsy; developmental venous anomaly

INTRODUCTION

Cerebral vascular malformations were classified by McCormick [1] in 1966 into the following groups: venous angioma, capillary telangiectasias, cavernous malformations, and arteriovenous malformations. Some cases of combination or hybrid malformations have been reported [2–5]. The phrase developmental venous anomaly (DVA) was proposed instead of venous angioma, by Lasjaunias et al., in 1986 because they are considered to be a variation of the normal venous drainage and formed during developmental stages in utero [6].

DVAs have been reported to occur in 2.5% to 3% of the generalized population and make up 60% of the central nervous system vascular malformations [7–9]. These malformations can be found in both adult and pediatric populations and is more common in men [10,11]. DVA is a congenital malformation of the venous drainage that occurs sporadically due to intrauterine ischemia, causing aberrant venous architecture to develop [12]. These might occasionally develop as a result of dominant inheritance of a gene mutation in the short arm of chromosome 9 [13]. These anomalies appear as radially arranged medullary veins (caput medusa) separated by normal brain parenchyma, which empty into a collector vein that drains into either superficial subcortical veins or deep pial veins [7].

These anomalies are generally described as benign, asymptomatic vascular malformations usually found coincidentally on neuroimaging [14]. Uncommonly symptomatic developmental venous anomaly may present with headaches, seizures, hemorrhage, and neurologic deficits [15]. The incidence of seizures associated with symptomatic DVAs ranges from 8% to 29% [16–18]. Although cases of existence of DVA and seizures have been reported, the correlation between the two

Vol. 8, No. 2, pp. 24–27. Published April, 2015.

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has not been firmly established [7,15,19]. Hippocampal sclerosis is commonly associated with temporal lobe epilepsy [18] and rarely with DVA [18].

We provide a description of a case of complex partial epilepsy and associated DVA and discuss the pathophysiology, neuroimaging, and clinical characteristics.

Case Report

A 21-year-old man presented with a history of complex partial epilepsy with secondary generalization and loss of consciousness. The seizures started at the age of 4 years. He was treated with Chinese herbal medication which reduced the frequency of these episodes to 1 or 2 seizure episodes annually. Because of infrequent occurrence, the patient discontinued the medication after which the frequency of the epileptic episodes increased 1 or 2 episodes every 3 months.

Patient was started on carbamezipine and phenytoin after which he had no further recurrence of seizures. Because of noncompliance, patient had intermittent episodes of complex partial seizures preceded by prodromal phase of visual scotomas in right visual field. Patient presented to the hospital with an episode of complex partial seizure with secondary generalization.

His examination showed no neurological deficits. An EEG was performed which demonstrated spike and wave discharges predominantly in the left frontotemporal region. A MRI was performed which demonstrated a hypointense linear lesion (flow void) suggestive of DVA and volume loss in the left temporal lobe (Fig. 1).

A cerebral angiogram was performed with selective contrast injection from catheter placed in left vertebral artery. Image acquisition and three-dimensional reconstructions were performed in the venographic phase. The angiogram demonstrated DVA that connected with the left transverse venous sinus and an anastomotic vein between the straight sinus and the transverse venous sinus traversing the brain parenchyma (Fig. 2).

The patient was restarted on carbamezipine for the treatment of complex partial seizures.

DISCUSSION

DVA is a variation of normal venous drainage considered to be formed during Padget's fourth to seventh stage of development [7]. These variations can either be in the form of dilated medullary veins that drain into a transcerebral collector vein [12] or intrinsic venous

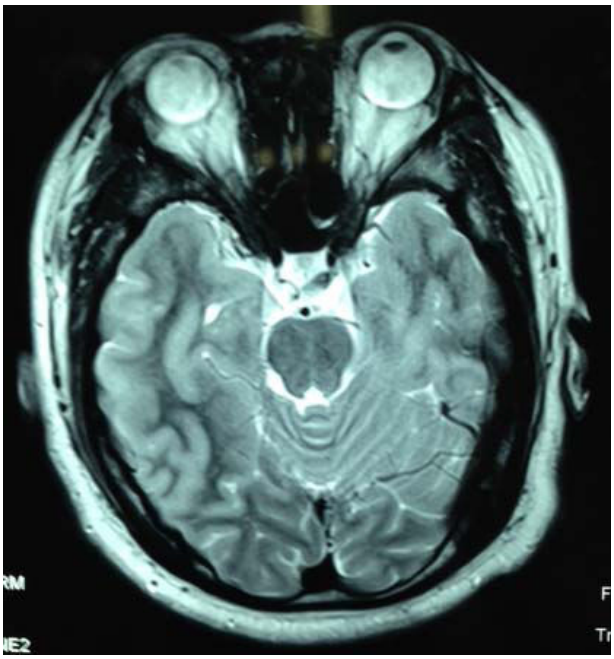

Figure 1. Linear flow void (arrows) suggestive of developmental venous anomaly in the left temporal lobe on T2-weighted image on MRI

anastomoses without the normal draining veins in the area [7]. DVAs are composed of thin-walled vessels dispersed in normal brain parenchyma, draining into a thick-walled large caliber vein, without an elastic lamina or smooth muscle layer [20,21]. DVAs lack any proliferative potential and do not have direct arteriovenous shunts [7]. DVAs can either drain into deep subependymal veins and the galenic system or drain into superficial cortical veins. The superficial pattern is present in about 70%, while the deep drainage pattern is present in 20% of the population [6,7,22]. The remaining 10% have a combination of the superficial and deep drainage [6,7,22]. DVAs are mostly supratentorial and are found most frequently in the frontal lobe (36% to 56%) followed by the parietal (12% to 24%) occipital (4%) and the temporal lobes (2% to 19%); the cerebellum (14% to 29%); the basal ganglia (6%); the thalamus and ventricles (11%) and the brainstem (less than 5%) [6,7,22].

The diagnosis of DVAs can be difficult and seldom possible on noncontrast CT scan unless there is an associated cavernous malformation [7,14]. On contrast-enhanced CT scan, the draining vein can be easily seen as a curvilinear or linear area of enhancement coursing the deep white matter and connecting to a deep or a cortical vein or to the dural venous sinus. Noncontrast MRI usually shows a flow void [7,14] and gadolinium con-

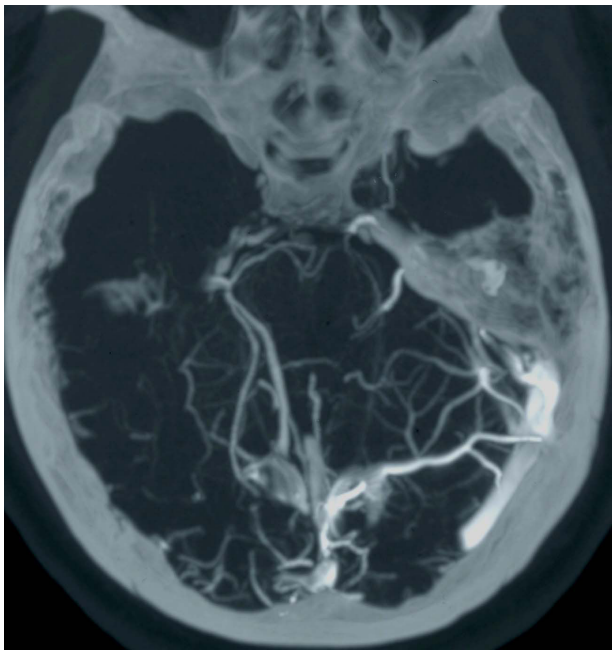

Figure 2. Development venous anomaly that connected with the left transverse venous sinus (arrow) and an anastomotic vein between the straight sinus and the transverse venous sinus (arrows) traversing the brain parenchyma

trast administration results in enhancement of the network of medullary veins and the collector vein due to the slow flow [9,14]. An angiogram classically shows a "caput medusa" appearance of the medullary veins during the early to middle venous phase [7,14].

The features of temporal lobe epilepsy commonly are simple partial or complex partial seizures with secondary generalization [23]. DVAs have been reported to be associated usually with generalized seizures, [16,24], but some patients have experienced partial seizures [16], complex partial seizures [24] or even Jacksonian march of motor seizures [25]. In our patient, the characteristics of seizures and spike and wave discharges in the left frontotemporal region on EEG indicate that the seizures were originating from the temporal lobe. No clear spatial correlation between the location of a DVA and the origin of seizures has been observed in previous reports [24,26]. Very few cases have been reported in which DVA was located in the same area as the EEG focus of the seizure [20]. In most of the cases, DVAs were located in a different region with respect to the focus of the seizure [18,26] or there was another lesion found that could be the cause of epilepsy [15]. It has been emphasized that a high incidence of seizures in patients with DVAs does merit some consideration [20]. It is also argued that the DVA is associated with abnormal neuronal migration and possible susceptibility to epileptogenesis [20]. However, recent literature suggests that DVA may be the cause of focal epilepsies in cases where no epileptogenic lesions can be detected [27]. Several mechanisms are postulated based on the following: (1) subclinical hemorrhage; more likely when a DVA is associated with a cavernous malformation [26] and (2) increased inflow or restricted outflow, resulting in intermittent cortical hyperemia and dysfunction creating an epileptic focus [7].

DVAs should be treated conservatively, and symptoms, such as seizures, should be managed medically [17]. Due to the fact that DVA may contribute to normal cerebral venous drainage, surgical obliteration of the anomaly can lead to venous thrombosis and occlusion causing venous ischemic and hemorrhagic complications [21,28].

CONCLUSION

DVAs are the most common cerebral venous malformations but are generally benign. Uncommonly, they can present with symptoms including seizures. These anomalies develop during embryonic development and may be a part of the functional venous drainage system of the brain. Therefore, these anomalies are managed conservatively and the seizures are treated with anticonvulsants medications.

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