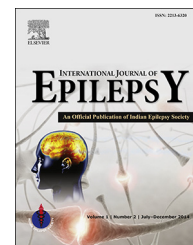


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Letter to the Editor

Vein of Galen malformation, a rare cause of epilepsy



ABSTRACT

Keywords:

Vein of Galen malformation
Median porencephalic vein
Neurological sign
Epilepsy
Brain imaging

Vein of Galen malformation (VOGM) is a rare congenital cerebral vascular malformation characterized by an aneurysmally dilated midline deep venous structure, fed by abnormal arteriovenous communication. Most patients develop severe congestive heart failure at neonatal period that is fatal if left untreated. Rarely, patients with low-flow fistula present with headache, seizure, or focal neurological sign at adulthood. A 28-year-old female with VOGM-related epilepsy was introduced in this brief report.

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Dear Editors,

The patient was a 28-year-old woman who presented with the first episode of a generalized seizure. At initial evaluation, she was normal mentally and her neurologic examination was unremarkable. There was no previous history of head injury, drug abuse, or other usual precipitants of a seizure attack. Routine lab tests were also within normal limits. So, for further evaluation, an unenhanced MRI was requested that showed a “vein of Galen malformation” (Fig. 1).

Vein of Galen malformation (VOGM) is a rare congenital vascular malformation characterized by an aneurysmally dilated midline deep cerebral venous structure, fed by abnormal arteriovenous communication. VOGM is a misnomer; this anomaly actually involves the median porencephalic vein of Markowski.¹ Most patients develop severe congestive heart failure at neonatal period that is fatal if left untreated. When there is a single fistula with a smaller shunt, patients present with macrocephaly or hydrocephalus and occasionally with mental retardation due to venous congestion and white matter injuries. Low-flow fistulas rarely present at adulthood with headache and epilepsy.^{1,2}

For assessment, MRI is the modality of choice. In classic cases, brain MRI shows markedly enlarged median porencephalic vein drains via the embryonic falcine sinus. Arterial feeders can be seen as flow voids along the wall of the vein. Periventricular white matter ischemic changes or calcification can also be detected.³



Fig. 1 – Coronal T2-weighted MR image showing abnormally dilated embryonic median porencephalic vein (white arrow) drains via the falcine sinus. Arterial feeder can be seen along the lateral wall of the vein (arrow head).

In this case, seizure was attributed to VOGM; carbamazepine was started and she is seizure free for more than one year.

Conflicts of interest

The author has none to declare.

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