Editorial
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Editors' Pick in May 2024

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Among the 13 papers published in the May issue of *Journal of Korean Neurosurgical Society* (*JKNS*) 2024, the following two papers, which deserve attention from readers, are selected by the editorial boards.

Update on the vein of Galen aneurysmal malformation : disease concept and genetics[2)]

Vein of Galen aneurysmal malformation (VGAM) is one of the important pediatric arteriovenous shunt diseases, especially among neonates and infants. It can be defined as direct arteriovenous fistulas (AVFs) between choroidal and/or quadrigeminal arteries and an overlying single median venous sac, which is the persistence of the embryonic median prosencephalic vein (MPV) of Markowski.

The MPV in human is an embryonic vein that appears as early as 32 days of gestation and disappears at 11th week, which is single midline vein, distinctly different from the permanent paired internal cerebral veins (ICVs). In normal embryogenesis, MPV regresses as paired ICVs form and annex the venous drainage of choroid plexus. Therefore, the ectatic vein in VGAM is actual residual MPV, not the vein of Galen per se.

The classification of VAGM is suggested in various ways based on the types of abnormal vascular connection. One of the points to be remembered is differentiation of VGAM from other conditions related to an enlarged vein of Galen.

It was found that RAS P21 protein activator 1 gene (*RASA1*) mutation was found in VGAM along with other vascular anomalies such as capillary malformation (CM), arteriovenous malformation (AVM). The gene *RASA1* is a neurofibromatosis type 1 (*NF1*) homolog and its product causes *RAS* activation. Damaging mutations in ephrin signaling genes, ephrin B2 (*EFNB2*) and ephrin receptor-B4 (*EPHB4*), which affect remodeling of vein from a capillary plexus into properly branched structures, were detected in VAGM probands. These genes were already reported in other Mendelian vascular diseases and it is plausible that VGAM may represent another phenotypic expansion of CM-AVM, hereditary telangiectasia (HHT)[4)]. Additional variants in *NOTCH* genes, *NOTCH3* and *NOTCH4*, were found in a series of VGAM. Further studies on the VGAM are required to deepen understanding of the disease and to identify potential druggable target.

Although VAGM is a rare disease entity, it is necessary to have a better understanding since it is challenging to manage.

Management of pediatric intracranial arteriovenous malformations[3)]

Central nervous system (CNS) AVM is the most common

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symptomatic intracranial vascular abnormality. It is known that the incidence of pediatric AVM is about 1/5000 children with no sex preponderance, and it represent about one sixth of AVMs in the general population. Although most AVMs are sporadic, familial AVMs may appear in hereditary hemorrhagic telangiectasia or mutations in *RASA1*. Mutations in familial or spontaneous AVMs share a common end pathway activating RAS, leading to poor vessel tube formation and dysregulated migration – helping to shed light on the pathophysiology of AVM.

The primary concern of pediatric AVM is spontaneous intracranial hemorrhage (ICH), results from the high-flow nature of the lesion. It is well known that bleeding from AVM not only causes 12–25% probability of fatality in each bleeding, but also increase the rebleeding risk after bleeding, which justifies aggressive approaches to treat the patients.

AVMs can be diagnosed on computed tomography angiography (CTA) and magnetic resonance imaging and angiography (MRI/MRA). However, once the AVM is identified, digital subtraction angiography (DSA) is to be followed to delineate detailed nidal anatomy. In addition to detailed anatomy, DSA can offer important predictive data on the risk of hemorrhage, including unfavorable factors such as outflow stenosis, smaller size and deep venous drainage.

It is acknowledged that aggressive approach towards the treatment of AVMs in the pediatric population is necessary, given the devastating outcome of bleeding and the cognizance that the longer lifespan of a child poses a higher overall risk over time. Treatment goal is total shutdown or removal of the AVM. Microsurgical resection confers immediate eradication of abnormal vessels causing hemorrhage, whereas the risk of surgery is to be considered, guided by various grading system. Radiosurgery and intervention also are useful next plans, and multimodality therapy may be especially important and referral to high-volume, experienced centers is encouraged.

Overall, most pediatric AVMs should be considered for treatment, including incidentally found, asymptomatic lesions. This recommendation is supported by data from the recent American Heart/Stroke Association guidelines[1]. This article gives the insights on overview of pediatric intracranial AVM and best treatment strategy for this challenging vascular lesion.

AUTHOR'S DECLARATION

Conflicts of interest

No other potential conflict of interest relevant to this article was reported.

Author contributions

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