

Vein of Galen Aneurysmal Malformation Presenting with Macrocephaly

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ABSTRACT

The vein of Galen aneurysmal malformation (VGAM) is a rare congenital intracranial vascular malformation found in children. Dilatation of the vein of Galen is a common feature of VGAM. This malformation is actually a vascular lesion that may present as different clinical pictures in childhood and is difficult to extract using classical surgical methods because of its locality and hemodynamic structure. Major complications of VGAM include heart failure, hydrocephaly, intracranial bleeding, and vascular steal phenomenon. Here the case of a 4-year-old girl who presented with macrocephaly and was diagnosed with VGAM is discussed in the light of literature.

Keywords: Vein of Galen malformation, macrocephaly, congenital

INTRODUCTION

The vein of Galen aneurysmal malformation (VGAM) is a rare congenital vascular malformation characterized by the shunting of the arterial flow into an enlarged cerebral vein of Galen. Although VGAM cases constitute only 1% of all cerebral vascular malformation cases, they comprise up to 30% of all pediatric vascular malformation cases. The arteries feeding VGAM are the posterior cerebral, choroidal, and posterior perforating arteries (1, 2). VGAM may present with different clinical pictures in childhood. Approximately 95% of newborns die of heart failure and the remaining 5% die of hydrocephaly, subarachnoid bleeding, or intraventricular bleeding. Older children present with different complaints, which most commonly includes a headache, followed by hydrocephaly, subarachnoid bleeding, and various neurological signs. An early surgical intervention is necessary to reduce the rate of these complications and prevent particularly severe cerebral injury (3-5).

In this case report, we present the case of a 4-year-old girl patient who presented with macrocephaly and was diagnosed with VGAM.

CASE PRESENTATION

A 4-year-old girl presented to our clinic with a headache and an enlarged head circumference. Her medical history revealed macrocephaly followed up since the age of 1. Her complaints had recently aggravated. A headache was localized in the frontotemporal region and was continuous. Upon a physical examination, her weight was evaluated to be 14 kg (<10th centile), height 93 cm (<3rd centile), and head circumference 52 cm (>97th centile). Her head appeared macrocephalic, and the frontal bossing was

present. Systolodiastolic murmur was auscultated all around the head, particularly in the left temporal region. She also had a systolic murmur of II/VI severity, best auscultated in the mesocardiac area. An echocardiography revealed a secundum type of atrial septal defect, 4 mm in size. A brain magnetic resonance imaging (MRI) demonstrated that the nidus and the vein of Galen were enlarged (Figure 1). On MR angiography, the internal cerebral vein and the vein of Galen were observed to be dilated and an arteriovenous malformation (AVM) was noted originating from the interpeduncular cistern extending into the quadrigeminal cistern and lateral ventricle, nourished by the posterior cerebral artery, draining into the internal cerebral vein and vein of Galen. On cerebral angiography, a grade III AVM nourished from the bilateral posterior choroidal and thalamo-perforating arteries was observed in the middle axis, and a surgical operation was planned. The Galenic vein system was markedly dilated (Figure 2). A color Doppler ultrasonography of the vertebral and carotid arteries was normal. Other laboratory examinations of the patient were also within the normal ranges. A written informed consent was obtained from the patient's parents.

DISCUSSION

The vein of Galen aneurysmal malformation is a congenital vascular malformation comprising 30% of all pediatric vascular anomalies and 1% of all pediatric congenital anomalies. It is believed to result from an insult to the cerebral vasculature at between 6 and 11 weeks of gestation after the development of the circle of Willis. Other venous anomalies commonly co-occur with VGAM, including anomalous dural sinuses, sinus stenoses, and an absence of the straight sinus (1, 2, 5). These anomalies com-

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Figure 1. Axial T2 weighted magnetic resonance imaging (MRI) demonstrating the arteriovenous malformation (AVM) of the nidus and an enlarged vein of Galen

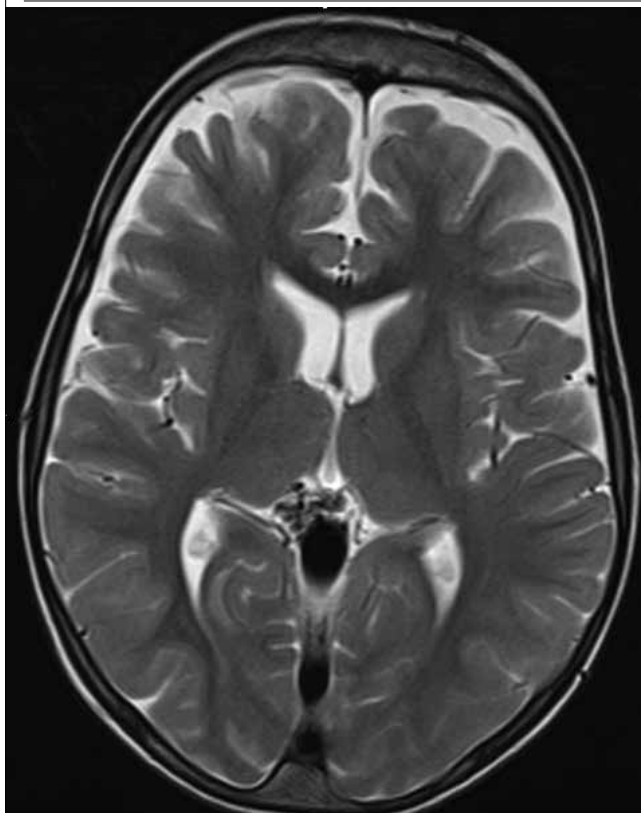
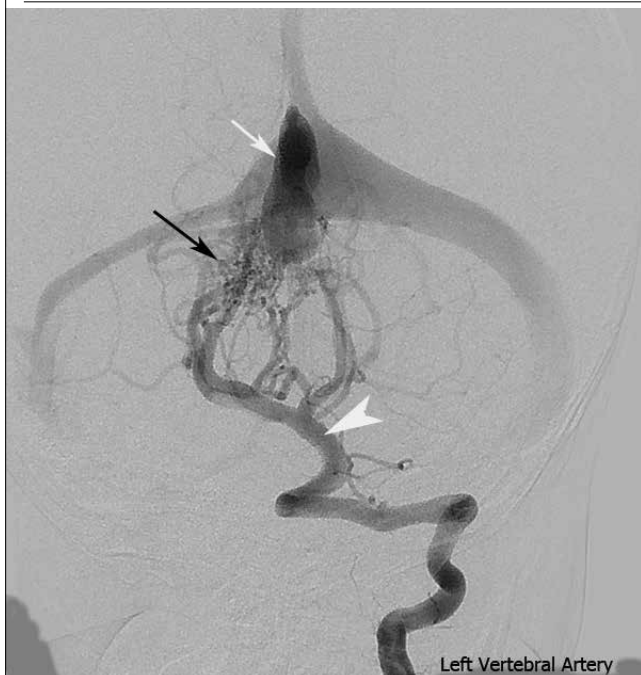


Figure 2. Left vertebral artery angiogram demonstrating AVM feeding artery (bacillary artery: arrowhead), nidus (black arrow), and draining veins (vein of Galen: white arrow)



monly present during the neonatal period, although they may also appear during early childhood. The major effects of VGAM include heart failure, hydrocephaly, intracranial bleeding, and vascular steal phenomenon (2, 6, 7).

In our patient, the main complaint upon presentation was a headache and an increased head circumference. Additionally, the systole-diastolic murmur was auscultated all around the head, particularly in the left temporal region. However, the physical examination and echocardiography did not demonstrate any finding of heart failure.

In their series of 59 cases, Lasjaunias et al. (8) classified VGAM into five different types, including parenchymatous AVM in 44%, choroidal AVM in 30%, mural AVM in 20%, vein of Galen varices in 7%, and dural AVM in 3% of cases, and reported that the pediatric population was particularly most sensitive to this shunt, regardless of the type (9). It has also been reported that the classification of this malformation is quite difficult in most cases.

In our patient, the cerebral angiography revealed a grade III AVM in the middle axis draining into the vein of Galen (approximately 2 cm in the longest diameter) (Figure 2). The case was considered to be complex, and surgery was planned.

Intracranial and vascular lesions might be treated using multimodalities, including surgery, chemotherapy, and conventional radiotherapy. However, complete excision might not be possible in all cases because of the anatomical localization of the lesion. Conventional radiotherapy and chemotherapy, as well as extended excision, may have adverse effects on the growth of child (10, 11).

Although the treatment of the aneurysm using endovascular methods is advantageous than that using surgery in several ways, it has certain drawbacks, including the use of a contrast material during the procedure and washing solution to prevent thrombosis, both of which adversely affect heart failure and require a close follow-up during anesthesia (12-14). VGAM can cause severe morbidity and mortality, particularly in neonates, but also in infants and older children. The mortality rate of VGAM is 9% and 50% in children and newborns, respectively, despite the administration of endovascular treatment (1, 2).

Endovascular surgery treatment was selected as the primary treatment in our case because of the localization of AVM. No complications developed after the surgery in our patient. After the discharge, the patient was followed up at the outpatient clinic.

Thus, VGAM must be considered in all children presenting with macrocephaly. Moreover, imaging studies must be performed in patients presenting with concomitant macrocephaly and headache. Endovascular surgery and various advanced surgical methods can be performed for the treatment of these cases.

Informed consent: Informed consent was obtained from parents of the patients who participated in this study.

Peer-review: Externally peer-reviewed.

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