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Secondary vein of Galen malformation with hydrocephalus: Treated with combined endovascular and endoscopic approach☆,☆☆,★

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ABSTRACT

Our goal is to describe a case of hydrocephalus in the setting of a Yasargil Type IV-C secondary vein of Galen malformation (VGM), treated with a combined endscopic and endovascular approach. We retrospectively reviewed the records of a patient admitted to the neurosurgical department hydrocephalus related to a secondary VGM.

A 47 year old male, presents with 8 months of progressive headaches and gait ataxia. Imaging displays a triventricular hydrocephalus, a tectal AVM, and an aneurysmal dilation of the great vein of Galen. Angiography displays a tectal AVM supplied by bilateral posterior choroidals with drainage into the vein of Galen. In addition there were direct high-flow arteriovenous fistulas into the vein of Galen via meningeal feeders. The diagnosis of a Yasargil Type IV-C secondary VGM was made.

The patient was treated via a combined endoscopic and endovascular approach. An endoscopic third ventriculostomy (ETV) was conducted for his hydrocephalus. He subsequently underwent a partial endovascular embolization of the left tentorial arterial feeder 1.5 months after his ETV.

Post-operatively his hydrocephalus resolved, and his complex secondary VGM completely obliterated after a single partial embolization.

Secondary VGMs with associated high-flow arteriovenous fistulas are rare. Our case provides a nice example a combined endoscopic and endovascular approach aimed at achieving cure of the hydrocephalus and the Yasargil Type IV-C secondary VGM. We were able to avoid the placement of shunt hardware via ETV treatment of the patient's hydrocephalus. Furthermore, our case displays the impact of partial embolization on control and potential cure of these complex vascular malformations.

Introduction

Tectal arteriovenous malformations (AVM) pose difficult treatment challenges. They can present through a variety of means: hydrocephalus, hemorrhage, and vascular steal syndromes. Treatment options include endovascular embolization and stereotactic radiosurgery. Microsurgical resection of these lesions is difficult given their location, proximity of critical structures, and deep venous drainage.

Rarely, tectal AVMs are associated with aneurysmal enlargement of the Galenic venous system, creating a secondary vein of Galen malformation (VGM) [1,2,4]. The clinical manifestations of VGMs depend on age, with congestive heart failure commonly affecting newborns and hydrocephalus (secondary to either aquaductal obstruction or venous hypertension) in older children/adults. The treatment of secondary VGMs involves endovascular, typically transarterial, embolization of the associated AVM or any fistulous connects to the vein of Galen [3]. In addition, associated hydrocephalus is usually managed via CSF diversion.

Within, we describe a rare case of a Yasargil Type IV-C [5] secondary VGM related to a tectal AVM, with both tentorial and middle meningeal artery direct arterio-venous shunts into the vein of Galen. The patient presented with symptoms related to hydrocephalus. We were able to successfully treat this patient with a combination of transarterial endovascular embolization of the malformation and an endoscopic third ventriculostomy (ETV) for the hydrocephalus.

Case report

A 47 year old male presents with a history of 8 months of progressive headaches, dizziness, and gait disturbances. Upon examination he is noted to display an ataxic gait. No other focal neurological findings

Abbreviations: VGM, vein of Galen malformation; AVM, arteriovenous malformation; ETV, endoscopic third ventriculostomy; MRI, magnetic resonance imaging; DSA, digital subtraction angiography.

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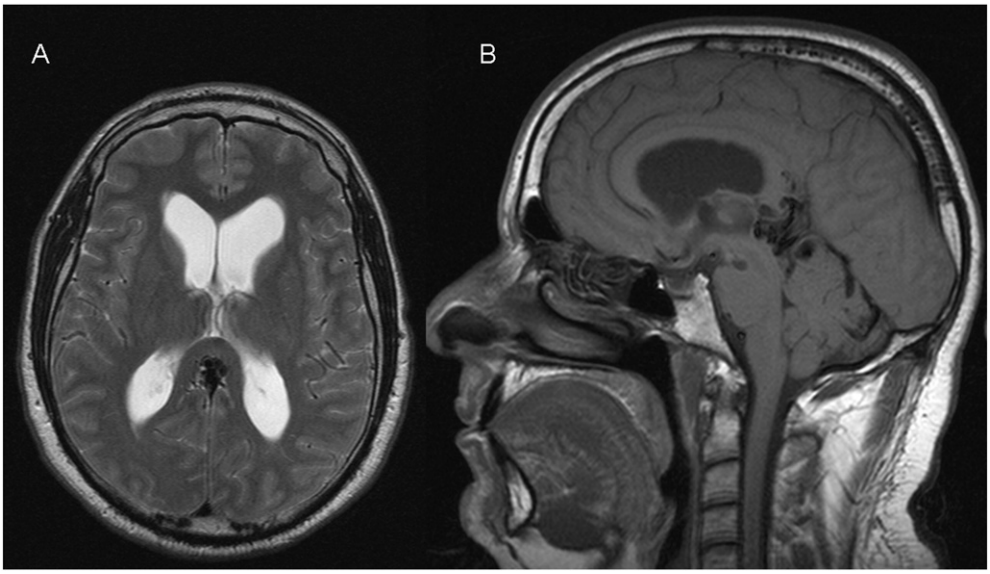

Fig. 1. MRI upon presentation. MRI = magnetic resonance imaging, DSA = digital subtraction angiography. Panel A: Axial T2 image displaying enlargement of the lateral ventricles, consistent with hydrocephalus. Large flow void can be seen in the area of the quadrigeminal cistern. Panel B: Sagittal T1 image displaying enlargement of the lateral and third ventricles. Arteriovenous malformation seen within the tectum, with dilated vein of Galen.

were demonstrated. Magnetic resonance imaging (MRI) of the brain (Fig. 1) displayed enlargement of the lateral and third ventricles, consistent with tri-ventricular hydrocephalus. In addition, there was a collection of flow voids noted in the area of the mesencephalic tectum/quadrigeminal cistern and a dilation of the great vein of Galen, all concerning for a tectal AVM.

Digital subtraction angiography (DSA) was conducted (Fig. 2) displaying a tectal AVM with arterial supply from the posterior choroidal arteries bilaterally. In addition, there were direct high-flow arterio-venous shunts into the vein of Galen via the left tentorial artery, and middle meningeal artery (bilaterally). The venous drainage consisted of the internal cerebral veins, and a large vein of Galen with aneurysmal dilation. The subsequent diagnosis was a Yasargil Type IV-C [5] secondary VGM from a tectal AVM with multiple direct arterio-venous shunts into the vein of Galen.

Given the patient's clinical presentation secondary to tri-ventricular hydrocephalus, it was elected to perform an ETV to provide CSF diversion and avoid a ventriculoperitoneal shunt. The ETV was performed without complication. The patient was then discharged home with immediate improvement in his headache symptoms, with the plan for endovascular treatment of his secondary VGM in the upcoming month.

The patient subsequently returned 1.5 months later for a planned staged embolization of his vascular malformation. The goal was to initially treat the high-flow shunt at the tentorial feeding artery. Histocryl embolic was administered directly into the left tentorial artery. Obliteration of this fistula was obtained. Injections of the internal carotid and vertebral arteries bilaterally demonstrated a significant reduction in the shunting into the vein of Galen, with some residual via the middle meningeal branches bilaterally. The procedure was stopped at this point, with plan to return in the future for staged embolization.

Post-operatively, MRI was conducted 3 months post-ETV and 1.5 months post-embolization. There was resolution of the hydrocephalus (Fig. 3). Similarly, a repeat DSA was performed 3 months after embolization, with the plan for a second embolization procedure. The DSA displayed complete resolution of both the tectal AVM, and absence of direct arterio-venous shunting into the vein of Galen via the previously untreated middle meningeal feeders. Thus the Type IV-C VGM was angiographically cured (Fig. 4).

The patient was subsequently followed over the next 2 years with MRI and 2 subsequent repeat angiograms. All continued to demonstrate complete resolution of the malformation and associated hydrocephalus.

Discussion

Secondary VGMs with associated high-flow arteriovenous fistulas are rare. Our case provides a nice example of a combined endoscopic and endovascular approach aimed at achieving cure of the hydrocephalus and the Yasargil Type IV-C secondary VGM.

A couple of important points can be gleaned from our case. We were able to avoid the placement of shunt hardware via ETV treatment of the patient's hydrocephalus. Furthermore, our case displays the impact of partial embolization on control and potential cure of these complex vascular malformations.

Despite the resolution of symptomatic hydrocephalus with the ETV, caution must be noted. The plan to trial an ETV was undertaken due to the presentation of clinically symptomatic tri-ventricular hydrocephalus with an adequately enlarged third ventricle. The underlying cause of hydrocephalus in these complex vascular lesions involving the vein of Galen is still unclear. Both aquaductal obstruction and venous hypertension leading to an absorptive defect are plausible explanations as to the etiology of hydrocephalus. Thus, there is the potential for resolution of hydrocephalus once the high flow fistulous component of the VGM is treated, though this may take time to occur. In our case, embolization of the secondary VGM may have contributed to the resolution of the hydrocephalus. However, given the progressive worsening of his symptoms, we decided that upfront surgical management of his hydrocephalus was warranted as we were worried about further clinical decline.

There are limitations to this study. First, this is a single case and in no way proves that this combined technique would be successful in all cases of complex secondary VGM. Second, partial embolization in certain circumstances can lead to dramatic changes in lesion hemodynamics and potentially increase the risk of hemorrhage and venous hypertension. Thus, whenever possible, complete embolization is preferred. Finally, our follow-up is limited to 2 years, thus the long-term angiographic results are unknown. Similarly, the long-term effect of the ETV on control of hydrocephalus in this case is limited as well.

Conclusions

Cases of secondary VGM related to tectal AVMs are rare. The associated hydrocephalus may be amenable to treatment via an ETV. Partial endovascular embolization of the AVM may lead to complete resolution of the lesion and the secondary VGM.