

Transarterial embolisation with Guglielmi detachable coils in an infant with a vein of Galen aneurysmal malformation

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Vein of Galen aneurysmal malformation is a rare entity in the paediatric population. However, it is being recognised with increasing frequency due to better diagnostic techniques. Neonates usually present with congestive heart failure, while in older infants and children it tends to manifest with seizures, hydrocephalus, intracerebral or subarachnoid haemorrhages. We present a case of ruptured vein of Galen aneurysmal malformation in a 3-month-old baby boy treated by transarterial embolisation using Guglielmi detachable coils.

Introduction

Vein of Galen aneurysmal malformations (VGAMs) are rare intracranial anomalies that make up 1% of all intracranial vascular malformations, and constitute 30% of all paediatric vascular malformations.¹ Although rare, VGAMs are being seen with increasing frequency due to better diagnostic techniques.

Case report

We present a case of a baby boy born at 35 weeks' gestational age via normal spontaneous delivery. Apgar scores were 8 at both 1 and 5 minutes. Antenatal ultrasound performed 5 days before birth showed a suspected intracranial aneurysm superior and posterior to the thalamus, which was confirmed to be vascular in nature by colour Doppler. His head circumference was 32.23 cm (within the normal range). Magnetic resonance imaging and magnetic resonance angiography (MRA) of the brain 3 days after birth showed a 2 cm x 1.9 cm enlarged central venous structure posterior to the thalami in the supracerebellar region, suggestive of a VGAM. At this time he was asymptomatic and had no clinical features of heart failure. At 3 months of age, he presented to the accident and emergency department with two episodes of generalised seizures and repeated vomiting. On physical examination, the baby was conscious and alert. Pupils were 3 mm equal and reactive. The anterior fontanelle was of normal tension with no dilated sutures nor dilated scalp veins. Computed tomography (CT) of the brain showed an intraventricular haemorrhage at the third, fourth and both lateral ventricles, together with hydrocephalus. A roundish soft tissue density was noted at the tentorium incisura (Fig 1a). Electroencephalography findings were normal. Digital subtraction angiography (DSA) showed a mural type VGAM with stricture at the falcine sinus. The VGAM was supplied by choroidal branches of both posterior cerebral arteries with venous drainage into falcine sinus (Figs 1b and 1c). Transarterial

Key words

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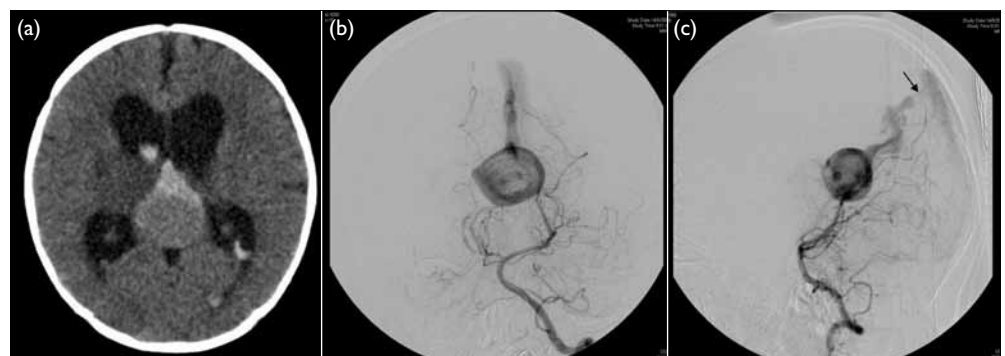


FIG 1. (a) Computed tomography of the brain (axial view) shows a vein of Galen aneurysmal malformation (VGAM) with intraventricular haemorrhage and hydrocephalus. (b) An angiogram shows a VGAM supplied by choroidal branches of both posterior cerebral arteries with venous drainage into falcine sinus (anteroposterior view). (c) Lateral view with stenosis at the falcine sinus (arrow) draining directly into the superior sagittal sinus with absence of straight sinus

使用電解可脫性微彈簧圈進行經動脈栓塞術治療一名患有Galen靜脈動脈瘤畸形的嬰兒

Galen靜脈動脈瘤畸形在小兒很罕見。不過，隨着診斷技術不斷進步，類似病例越來越多。新生兒患者通常會出現充血性心臟衰竭，而較年長嬰孩或兒童的症狀為癲癇發作、腦積水及腦內或蛛網膜下腔充血。本文報告使用電解可脫性微彈簧圈進行經動脈栓塞術治療患有Galen靜脈動脈瘤畸形的男嬰的病例。

embolisation was performed through packing of the aneurysm with 24 Guglielmi detachable coils (GDCs). The post-embolisation DSA showed complete obliteration of the malformation (Fig 2). No intra-operative complications occurred. Repeated CT brain



FIG 2. Post-embolisation angiogram shows complete obliteration of the malformation: (a) anteroposterior and (b) lateral views

showed complete resolution of the intraventricular haemorrhage. He was subsequently discharged in a stable condition without any neurological deficits.

Discussion

Vein of Galen aneurysmal malformation is a choroidal type of arteriovenous malformation involving the prosencephalic vein, a precursor of the vein of Galen that fails to develop because of the haemodynamic changes induced by the arteriovenous shunt. The ectatic venous structure seen in VGAMs actually arises from the median prosencephalic vein of Markowski rather than the vein of Galen itself.² The principal feeders of the malformation consist of two groups of vessels. The anterior group includes the anterior and middle cerebral, anterior and the posterolateral choroidal arteries, while the posterior group includes the posterior thalamoperforating, posteromedial choroidal, quadrigeminal and superior cerebellar arteries.^{1,2} Berenstein and Lasjaunias³ classified VGAMs into two types depending on the location of the fistula. Mural-type VGAMs have their communication within the wall of the median prosencephalic vein and are usually supplied by the collicular and posterior choroidal arteries, whereas choroidal-type VGAMs are characterised by multiple communications with the anterior end of the median prosencephalic vein and are supplied by the choroidal and pericallosal arteries, and branches of the thalamoperforators.^{3,4} It follows that in patients with suspected VGAMs, MRA provides greater detail of the feeding arteries than magnetic resonance venograms that focus on veins. In 1964, a clinical classification system of VGAMs that depended on the patient's age at presentation was proposed by Gold et al.⁵ These authorities indicated that neonates and infants had a higher tendency to present with heart failure. Whereas, in older children, compensated congestive heart failure, macrocephaly, hydrocephalus, headache, seizures, focal neurological deficits, developmental delay, as well as intracerebral and subarachnoid haemorrhages become dominant manifestations.

Persistent falcine sinuses (Figs 1b and 1c) are usually associated with anomalies such as VGAMs, arteriovenous malformations, absence of corpus callosum, osteogenesis imperfecta, and type II Chiari malformations. They serve as an alternate pathway to shunt blood from the deep venous system to the superficial system when the straight sinus is hypoplastic or atretic due to developmental abnormalities.

Heart failure in neonates is common, because the high-flow arteriovenous shunt causes increased venous return to the right atrium leading to high output cardiac failure; high pulmonary blood flow contributes to overload of the left cardiac chambers.

Right-to-left shunting occurs due to increased pulmonary arterial pressure, which prevents the normal closure of the ductus arteriosus.⁶ In our case, the baby had no evidence of heart failure at any time. Presumably, the stricture at the falcine sinus limited venous drainage thus eliminating high-flow arteriovenous shunting and associated phenomena.

Untreated VGAMs have a very poor prognosis.⁷ Johnston et al⁸ published a review of the literature in 1987 reporting a mortality rate of 77% in 92 untreated patients with VGAMs; neonates, in particular, fared worst and had the highest mortality rate of 96%. This was largely attributed to the high flow through the VGAM, which not only stole blood from surrounding brain parenchyma (leading to massive infarction), but also led to haemodynamic changes causing myocardial ischaemia and infarction. Early reduction in flow through the VGAM can reduce the steal phenomenon, and thereby prevent progressive cerebral ischaemia and worsening of congestive heart failure.⁷ Our patient presented with intraventricular rather than subarachnoid haemorrhage. This may have been because of ruptured vessels in the choroid plexus or germinal matrix owing to high pressures related to the VGAM. If left untreated, newborns with VGAMs almost uniformly endure a dismal outcome; surgery offers hope but outcomes are similarly disappointing with a neonatal mortality rate of about 79%.⁹ In two series that included five newborn patients, the death rate was 100%.^{8,10} Excluding neonates, the mortality improved to between 15 and 37%.^{8,10} With recent advances in endovascular treatment, mortality rates have dropped dramatically, down to 8% with transarterial embolisations.¹¹ Therefore, nowadays indications for surgery are largely limited to lesions inaccessible to endovascular therapy or where such treatments fail.

Hydrocephalus appears to be secondary to impaired resorption of cerebrospinal fluid (CSF) due to venous hypertension rather than aqueductal compression. Ventricular drainage of CSF by shunting may make cerebral venous hypertension worse, producing post-ischaemic epilepsy, neurological deficits and haemorrhage, and should therefore be avoided. According to a series described by Zerah et al,¹² only 33% of shunted patients had a favourable

outcome and more than 15% had significant mental retardation. On the other hand, 67% of non-shunted patients were free of neurological deficits or mental retardation; only 5% had significant mental retardation. Embolisation of the VGAM treats the underlying cause of the high venous pressure, leading to normalisation of the ventricles and thus is the treatment of choice.¹² Ventricular shunting should be reserved for patients who fail to improve despite adequate embolisation or have a hydrocephalus unrelated to the VGAM.¹³

In the past, VGAMs have been embolised transarterially with agents such as cyanoacrylate and detachable balloons. Other materials such as polyvinyl alcohol particles, silk sutures, or silicone beads have also been used.¹⁴ Possible fatal complications of endovascular embolisation include intracerebral haemorrhage due to venous hypertension and normal perfusion pressure breakthrough,¹⁵ perforation of the venous sac, ischaemic neurological deficits, and inadvertent pulmonary embolisation. Since the advent of GDCs in 1991, their use has become widespread due to easy repositioning and controlled detachment. They were also shown to be safe and effective in the obliteration of aneurysms.^{16,17} They offer an additional advantage over previous embolic methods in that the risk of inappropriate embolisation of the cerebral or pulmonary venous system is virtually non-existent. Furthermore, compared with balloons, it is believed that detachable coils are less traumatic and thus less liable to rupture an aneurysm.¹⁸

This report describes the effective use of GDCs for transarterial embolisation in the treatment of VGAMs. The result was complete obliteration of the VGAM without any complications and the patient subsequently recovered without any apparent sequelae.

Conclusion

Despite recent advances in embolisation techniques, the management of patients with VGAMs remains extremely challenging, as they are in an unusually high-risk group. Transarterial embolisation with GDCs is a safe and effective option for the treatment of VGAMs as shown by our case.

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