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Vein of Galen Aneurysms
Experience with Eleven Cases

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Summary

Eleven patients with so-called "vein of Galen aneurysms" are reported, six of whom presented with vein of Galen aneurysmal malformations (four with choroidal type and two with mural type malformations). The remaining five patients presented with vein of Galen aneurysmal dilatations secondarily due to an arteriovenous malformation in one patient, an arteriovenous fistula in another, dural arteriovenous fistulas in two patients, and a varix in another. Treatments for these patients were individualised with consideration given to the clinical manifestations and the angioarchitecture of their lesions. Endovascular intervention played a critical role in the treatment of these vein of Galen aneurysms.

Introduction

The entities known as "vein of Galen aneurysms" include two clinical findings, the vein of Galen aneurysmal malformation (VGAM) and vein of Galen aneurysmal dilatation (VGAD), that require different therapeutic approaches [1]. The VGAM is an embryologic remnant of the median vein of the prosencephalon [2] and can be further categorised as one of two types, choroidal or mural, according to its angioarchitecture [1]. With the choroidal VGAM, multiple bilateral arteriovenous fistulas (AVFs) are located in the velum interpositum cistern. With the mural type, a single AVF is usually located at the inferolateral aspect of the aneurysmal dilatation. In the VGAD, the great vein of Galen is embryologically normal, but becomes dilated secondary to a pial arteriovenous malformation (AVM), an AVF, a dural AVF, or a varix. We report our experience in treating eleven patients with vein of Galen aneurysms.

Methods

Eleven patients (nine males, two females; aged neonate to 58 years) comprised the study population, six of whom presented with VGAMs and five with VGADs. Their clinical characteristics are listed in table 1. Among the six patients with VGAMs, four presented with the choroidal type malformation and two with

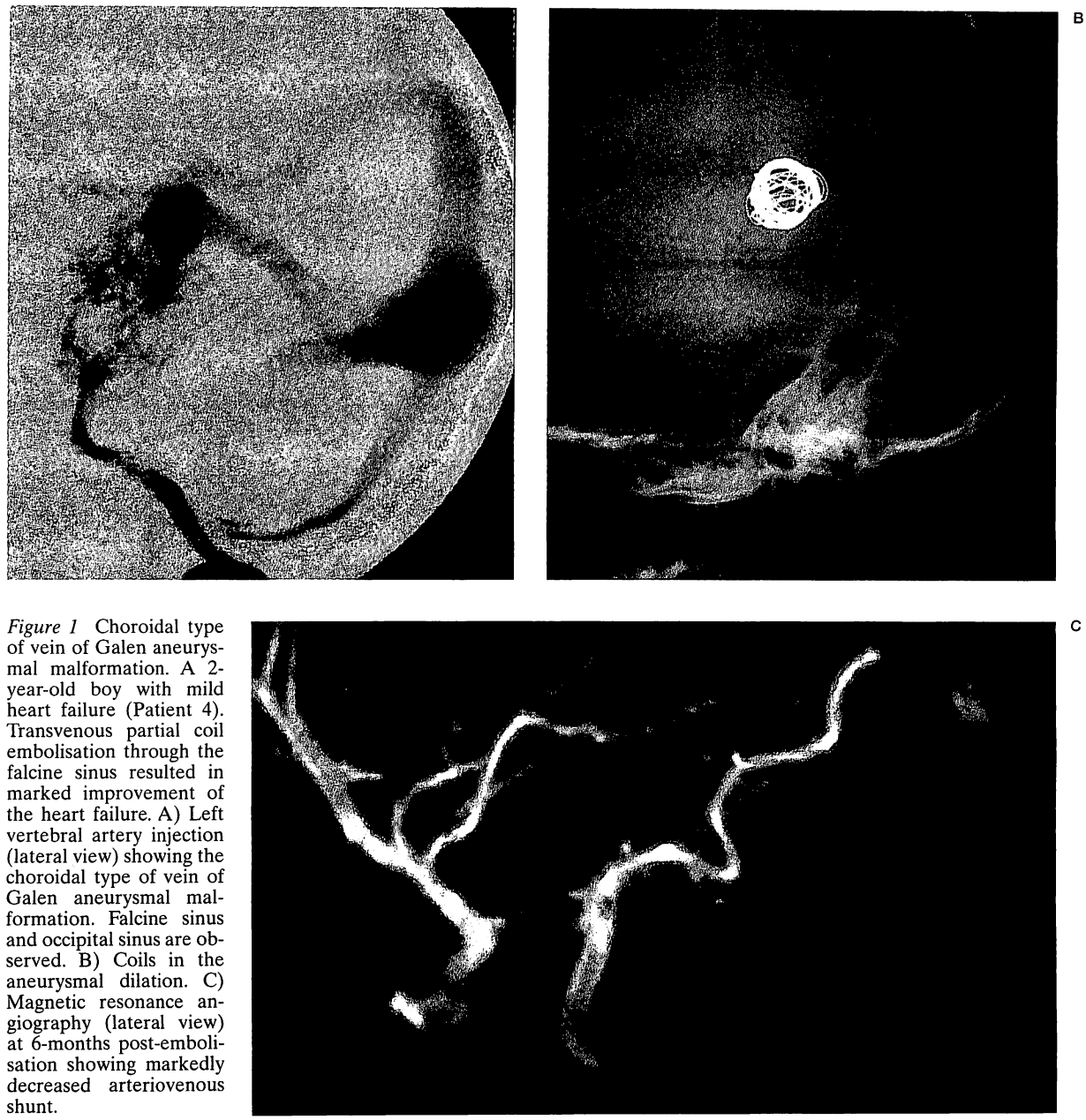

Figure 1 Choroidal type of vein of Galen aneurysmal malformation. A 2-year-old boy with mild heart failure (Patient 4). Transvenous partial coil embolisation through the falcine sinus resulted in marked improvement of the heart failure. A) Left vertebral artery injection (lateral view) showing the choroidal type of vein of Galen aneurysmal malformation. Falcine sinus and occipital sinus are observed. B) Coils in the aneurysmal dilation. C) Magnetic resonance angiography (lateral view) at 6-months post-embolisation showing markedly decreased arteriovenous shunt.

the mural type. Of the five patients with VGADs, one was caused by an AVM, another by an AVF, two by dural AVFs, and the last was the result of a varix.

Results

The four choroidal VGAMs were treated endovascularly. Two required urgent embolisation in the neonatal period for life-threatening congestive heart failure, but did not survive. Another neonatal patient with moderate heart failure died from a fatal perforation of the basilar artery during microcatheter navigation. A 2-year-old boy with mild heart failure was treated by transvenous partial coil occlusion of the aneurysmal dilatation, resulting in marked improvement of the heart failure (figure 1). The two patients with mural VGAMs were treated by direct clipping of the feeding artery near the arteriovenous shunts, resulting in clinical cure. Due to operative manipulation, unilateral oc-

cipital venous infarction occurred in one patient. At the time these two patients presented, endovascular surgery was not available.

One patient with a VGAD due to an AVM was initially treated by transarterial embolisation, which reduced the arteriovenous shunts but caused a mild hemicorporeal sensory disturbance. This patient was neurologically stable for six years, and underwent gamma knife surgery recently. One patient with a VGAD due to an AVF who presenting in status epileptics underwent open surgery. The operation was complicated by repeated postoperative haemorrhage and resulted in persistent vegetative state. One patient with a VGAD secondary to a dural AVF was treated first by transarterial embolisation and subsequently underwent gamma knife surgery. No rebleeding occurred for one year. Another patient with an incidentally discovered dural AVF refused any treatment. A neonate with a VGAD due to a varix presented with mild heart failure due to a persistent ductus arteriosus. Computed tomographic angiography on the first day of life showed the markedly dilated cerebral venous system, including the great vein of Galen. Ultrasound examination of this patient on the fourth day of life showed closure of ductus arteriosus after intravenous administration of indomethacin and spontaneous normalisation of the cerebral venous system. Computed tomographic angiography one month later demonstrated a normal dural venous system. Development of this patient was normal at the time of last follow-up (three years of age).

Discussion

Neonates with the choroidal type of VGAM typically develop severe congestive heart failure, while those with the mural type tend to present with hydrocephalus, mild heart failure, or seizures. Focal neurological deficits, headaches, subarachnoid haemorrhages, mental developmental delay are more frequently observed in older children with VGAMs[1,3]. Symptoms and signs of VGADs may include intracerebral haemorrhage, intraventricular haemorrhage, hydrocephalus, gaze palsy, and diplopia.

The treatment of a VGAM and VGAD differs, thus discrimination between the two is of vital importance. Angiographically, there are two important findings indicative of a VGAD: the presence of transmesencephalic vessels in the midbrain[4] and drainage to the deep cerebral venous system, such as the internal cerebral veins or the basal vein of Rosenthal. In the choroidal VGAM, an "arterial maze" between the dilated feeding arteries and the aneurysmal dilatation mimics the nidus of a pial AVM, and thus requires that these two entities be carefully discriminated. Secondarily dilated subependymal arteries and transcerebral arteries that converge toward the choroidal veins of the VGAM should be differentiated from the secondarily dilated subependymal arteries, transcerebral arteries, or above-mentioned transmesencephalic arteries found in VGADs. Due to the communication between the great vein of Galen and the normal deep venous system, it is contraindicated to treat the VGAD by transvenous occlusion of the vein of Galen. Normal venous return may flow into the Galenic system in the VGAD. In the VGAM, persistence of the falcine sinus, occipital sinus, marginal sinus, and steno-occlusive changes of the straight sinus and/or jugular veins are occasionally observed.

Indications for treatment of a neonatal VGAM depend on the patient's clinical status. Neonatal evaluation scores have been proposed to select candidates for treatment and determine appropriate timing[5]. The scoring system evaluates the neurological function (1-5 points), cardiac function (0-5), renal function (0-3), hepatic function including the coagulation system (0-3), and respiratory function (0-5), with a maximum score of 21. Therapeutic options are categorised according to scores as follows: no indication for treatment (<9), emergent embolisation (9-12), and delayed embolisation (>12). Intracerebral haematoma and cerebral infarction indicate serious brain damage, and are considered contraindications to therapy. The appearance of calcifications within the basal ganglia and the subcortex indicates parenchymal damage due to chronic venous ischaemia and requires emergent intervention.

Therapeutic options available for a VGAM include no treatment, open surgery, endovascular treatment, and stereotactic radiosurgery. Treatment of the given patient should be individualised with consideration of their age, the clinical manifestation of the lesion, and its angioarchitecture. Because of advances made with endovascular surgery, the prognosis of a

Patient no.	Classification	Age (year)	Gender	Initial symptoms	Treatment	Mode of embolisation	Complication	Outcome	Follow-up (year)
1	VGAM, choroidal	0 (neonate)	M	severe heart failure	embolisation	transarterial	-	death	-
2	VGAM, choroidal	0 (neonate)	M	moderate heart failure	embolisation	transarterial	vessel perforation	death	-
3	VGAM, choroidal	0 (neonate)	M	severe heart failure	embolisation	transarterial transvenous	subarachnoid haemorrhage	death	-
4	VGAM, choroidal	2.2	M	mild heart failure	embolisation	transvenous	-	good recovery	0.5
5	VGAM, mural	1.4	M	macrocephaly	clipping	-	occipital lobe infarction	good recovery	7
6	VGAM, mural	10	M	ventricular haemorrhage	clipping	-	-	good recovery	12
7	VGAD, AVM	36	F	diplopia, ptosis	embolisation gamma knife	transarterial	hemi-sensory disturbance	good recovery	6
8	VGAD, AVF	2.3	M	status epilepticus	surgery	-	postoperative haemorrhage	vegetative state	-
9	VGAD, dural AVF	50	M	ventricular haemorrhage	embolisation gamma knife	transarterial	-	good recovery	1
10	VGAD, dural AVF	58	F	incidental	-	-	-	good recovery	1.5
11	VGAD, varix	0 (neonate)	M	mild heart failure	-	-	-	good recovery	3

Table 1 **Clinical data of eleven patients with vein of Galen aneurysms**
Abbreviations: AVF, arteriovenous fistula; AVM, arteriovenous malformation; VGAD, vein of Galen aneurysmal dilatation; VGAM, vein of Galen aneurysmal malformation

patient with a VGAM has drastically improved, with this form of intervention now considered the treatment of choice due to its limited invasiveness [3,6-8]. However, the prognosis may largely depend upon the degree of arteriovenous shunting at the time of initial presentation. The VGAM diagnosed by ultrasound examination prenatally does not always require emergency treatment in the neonatal period. The goal of treatment is not an anatomical cure but normal growth. Stereotactic radiosurgery requires time to become effective, but it may be indicated in older children who do not require urgent intervention [9].

Endovascular treatments include transarterial, transvenous, and trans-torcular routes. In the neonatal period, a trans-umbilical route allows both transarterial and transvenous approaches [10]. Among these, it has been reported that transarterial embolisation provides the better outcome [7]. Transvenous and trans-torcular approaches are chosen as alternatives to transarterial should this approach not be feasible [6]. Embolic materials are selected with regard to the angioarchitecture, amount of arteriovenous shunting, route of approach, and experience of the operator. In general, liquid glue and coils are used in transarterial embolisation and coils in transvenous embolisation. If one session of embolisation is not enough, a subsequent intervention can be performed within a few days leaving the femoral vascular sheath in place.

Conclusions

Eleven patients with so-called "vein of Galen aneurysms" were reported. There were four choroidal and two mural VGAMs, and five VGADs. Treatments for these patients were individualised with consideration of the clinical manifestations and the angioarchitecture of the lesions. Endovascular treatment played an important role in the treatment of these vein of Galen aneurysms.

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