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Normal galenic drainage of the deep cerebral venous system n two cases of vein of Galen aneurysmal malformation

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Abstract *Introduction:* Vein of Galen aneurysmal malformations (VGAM) are assumed to be related to the persistence of the embryonic median prosencephalic vein of Markowski, which does not involute in cases of VGAM and becomes the venous collector of the shunt, characteristic of the malformation. The arterial feeders of VGAMs are all primitive meningeal arteries present during the embryonic period of the constitution of the malformation. It has also been assumed that the internal cerebral veins were absent in patients presenting with VGAM. There is no clear evidence indicating, however, that the deep venous structures cannot develop normally. *Case reports:* We report on two cases of VGAM in which superselective retrograde transvenous catheterization and MRI demonstrated that normal internal cerebral veins were draining into the aneurysmal vein of Galen. *Conclusions:* It is conceivable that, as in our patients, this drainage pattern is only angiographically detectable via selective retrograde transvenous drainage. The possibility of such normal deep galenic venous drainage must be considered in VGAM management, as it may imply occurrence of adverse effects when the malformation is occluded on the venous side.

Keywords Vein of Galen aneurysm · Embolization · Deep venous system

Introduction

Vein of Galen aneurysmal malformations (VGAM) are congenital arteriovenous malformations usually diagnosed during the perinatal period or early childhood [14]. The clinical presentation of a VGAM depends on morphologic factors such as the number and size of the arteriovenous shunts and the anatomy of the venous drainage pathways. In particular, the presence of stenotic lesions on the venous side represents a favorable prognostic factor that tends to limit the severity of the hemodynamic repercussions of the VGAM [14]. Typical clinical pictures include high-output congestive heart failure in neonates, or seizures, hydrocephalus, and developmental delay in infants and children rather than hemorrhage [10, 11]. Hydrocephalus may be caused by an impaired cerebrospinal fluid hydrodynamic due to increased intracranial venous pressure [4] or to a mass effect [2, 12] on the aqueduct or posterior third ventricle caused by the VGAM itself [1, 3, 15]. Diagnosis in adulthood is rare; it may be incidental or secondary to headache, seizure, and/or intracranial hemorrhage.

The morphologic hallmark of a VGAM is the persistence of a venous embryonic precursor, the median prosencephalic vein of Markowski [14], which normally participates in the formation of the vein of Galen. This embryonic vein does not involute in cases of VGAM, becoming instead the enlarged median venous collector that is characteristic of the malformation. VGAM treatment now relies almost exclusively on endovascular techniques, via either transarterial or transvenous approaches. The absence of normal venous drainage into the

varicose collector of the malformation is a commonly described anatomic feature of VGAM [6]. The assumption that the malformation does not participate in the venous drainage of normal cerebral structures has important implications for treatment planning, since it suggests that therapeutic obliteration of the venous component of the lesion can be safely performed. We report two cases of VGAM in which superselective retrograde transvenous catheterization and MR imaging demonstrated the presence of normal veins draining into or immediately posterior to the ectatic venous side of the lesion. The potential implications of these observations for the understanding and management of VGAM are discussed.

Case reports

Patient 1

A 40-year-old woman was investigated for pituitary dysfunction. Her past medical history was unremarkable. She had two healthy children born after uncomplicated pregnancies. Her physical examination was normal. MR imaging incidentally identified an enlarged vein of Galen draining into the straight sinus, with a moderate mass effect on the mesencephalon. The brain parenchyma was unremarkable and the cerebral ventricles were normal in size. MR imaging also suggested the presence of bilateral internal cerebral veins (ICV) terminating into the dilated vein of Galen. Digital subtraction angiography (DSA) confirmed the diagnosis of VGAM (mural type). An enlarged right lateral posterior choroidal artery was the sole angiographically detectable supply of the VGAM with a direct mural shunt. The venous drainage occurred directly into the straight sinus as well as towards the left transverse sinus via two tentorial veins. A stenosis was demonstrated at the junction between the vein of Galen and the straight sinus. The potential risks and benefits of endovascular management were reviewed with the patient, who chose to proceed with the treatment. The procedure was performed using a transfemoral arterial access. A microcatheter advanced into the posterior lateral choroidal artery could easily be passed into the venous side of the lesion through the mural arteriovenous shunt. The microcatheter was slightly pulled back on the arterial side of the VGAM and placed as close as possible to the shunt, which was occluded using mechanically-detachable platinum coils (Detach-18, Cook Europe, Copenhagen, Denmark). The early postprocedural angiogram demonstrated almost complete obliteration of the fistula, while the patient was still anticoagulated. Sequential MR imaging performed 3 days, 2 months, and 4 months after the embolization confirmed thrombosis of the lesion with progressive shrinkage of the pouch and resolution of the mass effect.

The physical examination of the patient was still normal 14 months later. However, follow-up angiography obtained concomitantly showed, besides complete occlusion of the initial varix, the presence of a new fistula supplied by the right and left posterior medial choroidal arteries. This fistula consisted of multiple microshunts linking a plexiform arteriolar network to the anterior and inferior aspects of the vein of Galen. The venous drainage occurred via the straight sinus. Endovascular treatment of this new shunt was attempted after 15 months, via both arterial and venous approaches. A 6-F guiding catheter (Envoy®, Cordis, Miami, FL, USA) was advanced into the left transverse sinus via a transfemoral venous access. A microcatheter (Rapid Transit, Cordis, Miami, USA) was then passed successively through the right lateral sinus, the straight sinus, and the vein of Galen, before finally being pushed across the supposed fistula. Superselective angiograms revealed that the shunt was draining first into multiple small venous structures located in the anterior and inferior aspects of the vein of Galen, which in turn communicated with the latter through a single small channel in which the microcatheter was placed. During the transvenous exploration of the ectatic vein of Galen, the microcatheter was also advanced into both ICVs, into a left tentorial vein, and into the right basal vein. Each of these vessels was documented by superselective angiography. Even if obliteration of the venous ectasia at the site of the shunt had spared these normal venous structures, the particular drainage of the shunt via an initial venous network that subsequently emptied into the vein of Galen via a single communicating channel would have left the actual fistulous connections untreated and put the patient at risk of intracranial hemorrhage by shunt rupture. Therapeutic obliteration of the venous side of the lesion was therefore not performed. A transarterial embolization was attempted by advancing a microcatheter into several choroidal branches of the left and right posterior cerebral arteries. This allowed visualizing the complex arteriolar plexus supplying the shunt, but the feeders were too small to be superselectively catheterized. The shunt could therefore not be occluded from the arterial side without taking a significant risk of brainstem stroke. It was decided to leave the residual lesion untreated. The patient was neurologically intact after 18 months of follow-up.

Patient 2

An 11-year-old girl was referred to our institution for macrocephaly and a history of mental retardation. Her medical past was otherwise unremarkable. Hydrocephalus sparing the fourth ventricle and a dilated vein of Galen were demonstrated by CT. A VGAM was confirmed by MR imaging, which also showed compression of the cerebral aqueduct by the venous ectasia. Multiple enlarged choroidal arteries were documented by MRA. DSA characterized the VGAM as being of a mixed mural and choroidal type, with bilateral posterior choroidal and right pericallosal arterial supply. The arteriovenous shunts were located in the anterior and superior aspects of the aneurysmal sac. The venous drainage occurred through the straight sinus and the superior sagittal sinus via an unusual falcine drainage (falcine sinus and “falcine loop” [14]). DSA also revealed proximal occlusion of the right pericallosal artery, as well as multiple stenoses along the right pericallosal artery and at the origin of the left middle cerebral artery. Feeders derived from the anterior and middle cerebral arteries were therefore tributaries of a complex collateral network that precluded direct transarterial access to the VGAM.

Endovascular treatment was performed under general anesthesia using 5-F and 6-F guiding catheters (Envoy, Cordis) placed via transfemoral arterial and venous accesses respectively. The tip of the 6-F guiding catheter was positioned in the left lateral sinus, and a 4-F catheter (Terumo®, Tokyo, Japan) was coaxially advanced into the venous ectasia via the straight sinus. Using a triple coaxial technique, a microcatheter (Rapid Transit®, Miami, FL, USA) was then navigated towards a large venous collector located at the anterior aspect of the dilated vein of Galen. This collector was draining both the mural and plexiform components of the shunt. Through this collector, it was possible to catheterize the posterior pericallosal artery, the posterior choroidal feeders, and both ICVs. All these vessels were superselectively documented.

Occlusion of the shunt in the anterior venous collector was obtained by using mechanically detachable platinum coils (Detach-18, Cook Europe, Copenhagen, Denmark). Additional transarterial embolization of a right posterior choroidal branch was performed using the same coils. Postprocedural DSA demonstrated almost complete occlusion of the fistula, with a minor persistent shunt fed by the left posterior choroidal artery. The patient woke up with an unchanged clinical status. She had a seizure 10 h later that

prompted an emergency CT revealing intraventricular and focal subarachnoid hemorrhages. A ventriculoperitoneal shunt was placed after documentation of progressing hydrocephalus. Follow-up DSA performed 4 months later demonstrated minor residual left posterior choroidal artery supply, with almost complete resolution of the venous ectasia. The venous drainage occurred through the straight sinus only, the falcine sinus being no longer opacified.

Discussion

The median prosencephalic vein of Markowski is the embryonic precursor of the vein of Galen (VG) [5, 14]. During normal development of the deep cerebral venous system, the median prosencephalic vein partially involutes after being joined by the newly formed ICVs. The portion of the median prosencephalic vein located cranially to the junction of the ICVs disappears, while its caudal end participates in the formation of the VG. This cranial regression does not take place in the presence of a VGAM. Instead, the development of multiple arteriovenous shunts between choroidal and/or collicular branches and the median prosencephalic vein results in the persistence and subsequent massive dilatation of the latter, which then assumes its characteristic posterior midline "aneurysmal" appearance. In this respect, VGAMs have to be distinguished from vein of Galen dilatations (VGAD), in which drainage of a neural or choroidal vascular malformation (Fig. 1) occurs through an enlarged but developmentally normal VG [8].

In cases of VGAM, the occurrence of normal venous drainage into the aneurysmal collector is debated. Raybaud et al. [14] detected an ICV connected to the vein of Galen in 6 out of 12 cases of VGAM with adequate angiographic data. These veins were always unilateral and nondilated. Five of these ICVs drained into the aneurysmal sac: 4 directly and 1 via an intermediate venous plexus. One ICV was opacified retrogradely. The basal vein was not seen connected to the venous sac in any of the patients in their series. Lasjaunias et al. [3, 7, 8, 9], on the other hand, stated that normal venous drainage never occurs through a VGAM collector. Normal, nondilated veins draining into the varicose sac of the malformation were observed in our two patients. These veins, which included both ICVs (Patients 1 and 2) and the right basal vein (Patient 1), were documented by superselective retrograde catheterization performed during transvenous exploration of the VGAM, but could also be retrospec-

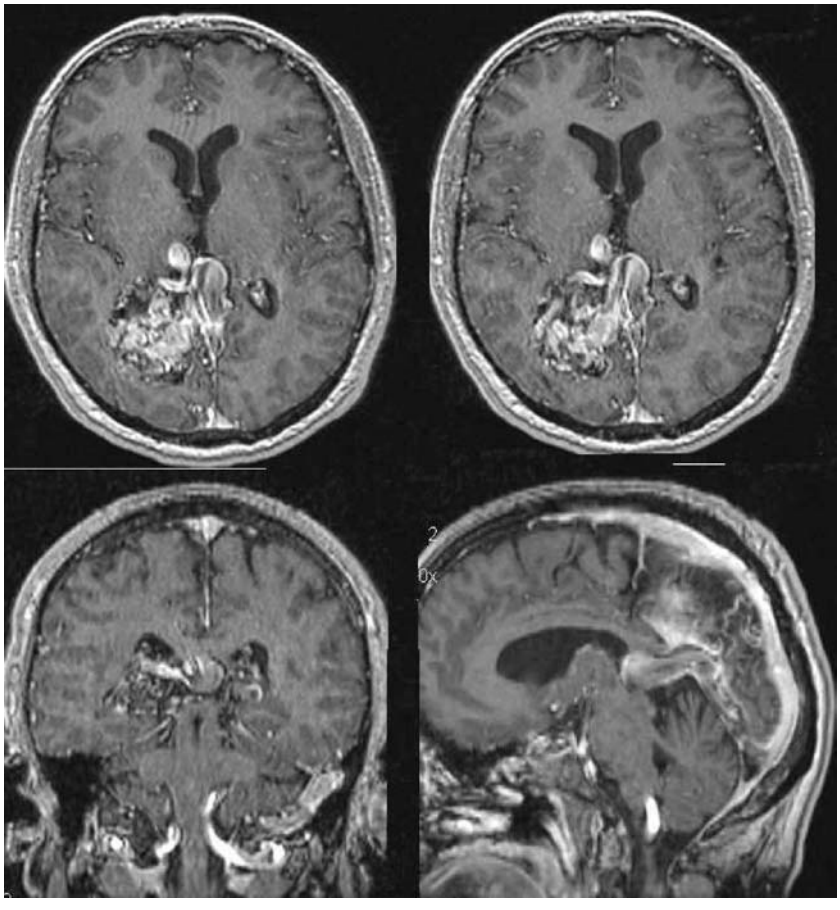


Fig. 1 Vein of Galen dilatations: MR examination of a patient presenting with an occipital arteriovenous malformation draining into an enlarged but developmentally normal vein of Galen

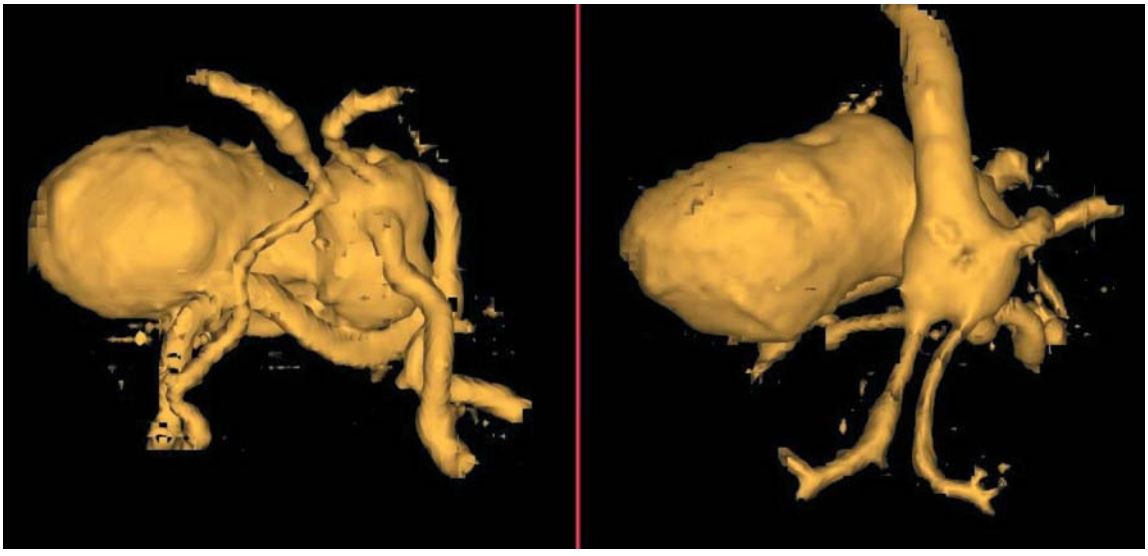


Fig. 2 3D reconstruction of Patient 1's pretreatment MR examination (gradient echo T1 WI post gadolinium). Internal cerebral veins and basal vein are located in front of the venous pouch. These normal veins were not seen on a regular angiogram

tively visualized on MR imaging studies obtained prior to DSA (Fig. 2). These veins were, however, not detectable on the venous phase of transarterial angiograms. This latter finding suggests that flow patterns and contrast distribution that occur in a VGAM may prevent visualization of normal veins draining into the venous collector. These normal veins may then only be detectable by superselective catheterization, as in our two patients. Such a hypothesis has important consequences for endovascular or surgical [13] therapy planning. It implies that therapeutic obliteration of the venous side of a VGAM may result in impairment of normal venous drainage, potentially leading to venous infarction and/or intracranial hemorrhage [10]. This would apply to both a transvenous approach or to a transarterial approach with passage of large quantities of embolic material to the venous side. When a transvenous approach is chosen, selective obliteration at the site(s) of the fistula(s) should be preferred to complete occlusion of the venous pouch, since it would lessen the risk of inadvertent normal drainage impairment. A schematic representation of the treatment options best suited to different VGAM configurations is provided in Figs. 3, 4, 5, 6, 7, 8, and 9.

In summary, our experience shows that drainage of normal venous structures can occur through the deep (galenic) venous system in cases of VGAM. These normal veins were not demonstrated on the venous phase of angiographic studies obtained via a transarterial approach, but were documented by superselective retrograde transvenous microcatheterization. Retrospectively, they could also be detected on MR imaging. This discrepancy between DSA and MR imaging may be attributable to preferential contrast agent distribution related to the VGAM pattern of flow. Pretreatment evaluation of a VGAM morphology should in our opinion include high-resolution MR imaging with gadolinium enhancement and MR angiography/venography focusing on the characterization of the normal cerebral venous drainage. Careful superselective retrograde microcatheterization

Fig. 3 Dilatation of a persistent prosencephalic vein due to the presence of direct or plexiform shunts located on the remnant itself. The posterior part of the prosencephalic vein is not directly involved in the arteriovenous shunts and normally participates in the formation of the vein of Galen. *ACA* anterior cerebral artery, *ICV* internal cerebral vein, *Chor A* choroid arteries, *A Chor A* anterior choroidal artery, *Colic A* collicular artery, *Aneur* aneurysm, *V of G* vein of Galen, *Falc S* falcine sinus, *SLS* superior longitudinal sinus, *StrS* straight sinus, *BV* basilar vein, *P Chor A* posterior choroidal artery, *MCA* middle cerebral artery, *P Com* posterior communicating artery, *PCA* posterior cerebral artery, *Car A* carotid artery, *Bas A* basilar artery, *S Cb A* superior cerebellar artery

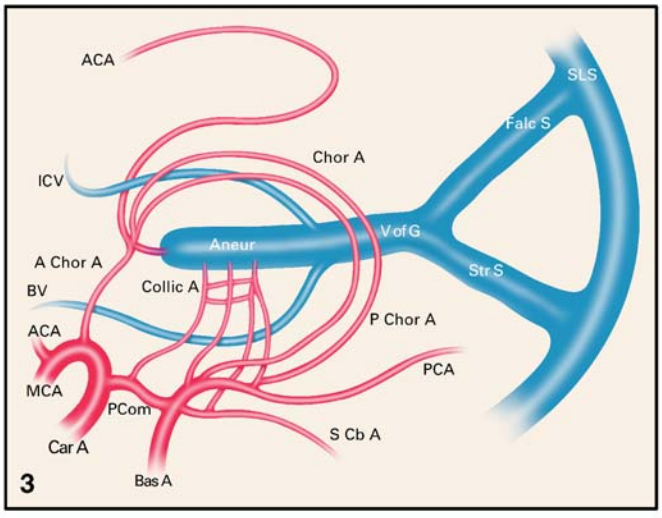
Fig. 4 Normal anatomy. The anterior part of the prosencephalic vein has disappeared, the posterior part contributed to the normal vein of Galen. *P Cb A* posterior cerebellar artery

Fig. 5 Schematic growth patterns of the vein of Galen (*arrows*) related to the increased flow from the shunt(s)

Fig. 6 Single direct shunt, simplest expression of the malformation. The treatment can be achieved by occluding the arterial feeder (with coils or glue)

Fig. 7 The anterior pouch is the true location of the shunt(s). The dilatation of the vein of Galen is a secondary hemodynamic consequence of the increased flow. Multiple direct shunts in an individualized anterior pouch. Treatment option: transvenous occlusion with coils, sparing the normal venous drainage

Fig. 8 Maximum dilatation of the vein of Galen region. The true shunt location cannot be determined on the basis of the external morphology of the vein of Galen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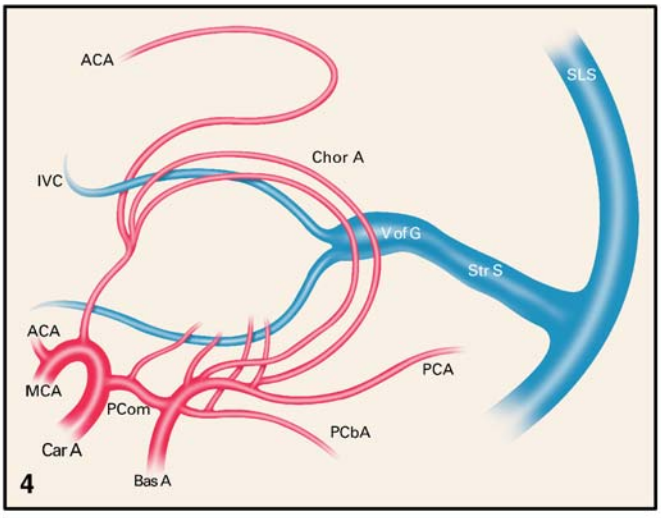
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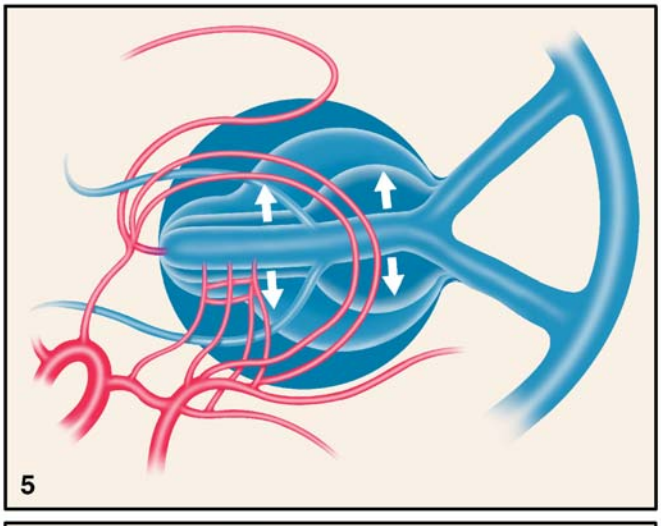
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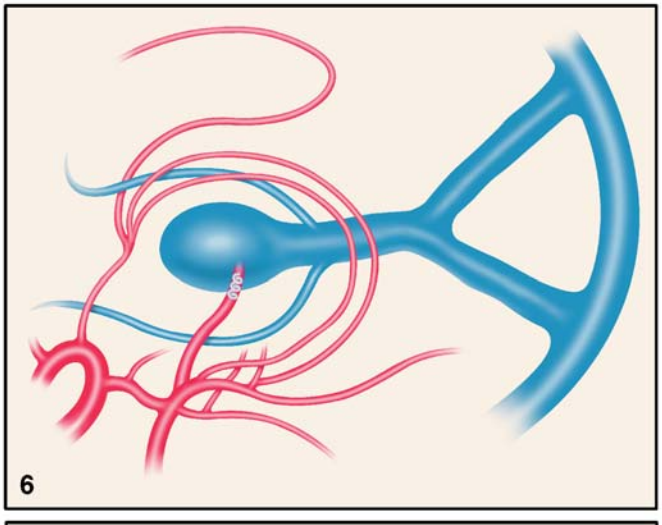
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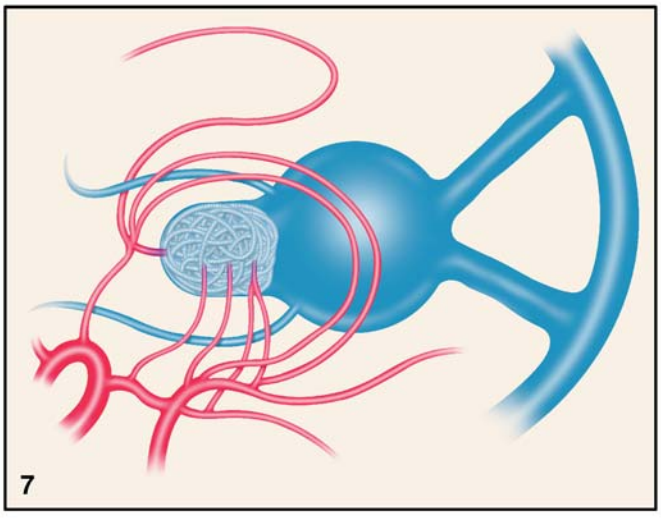
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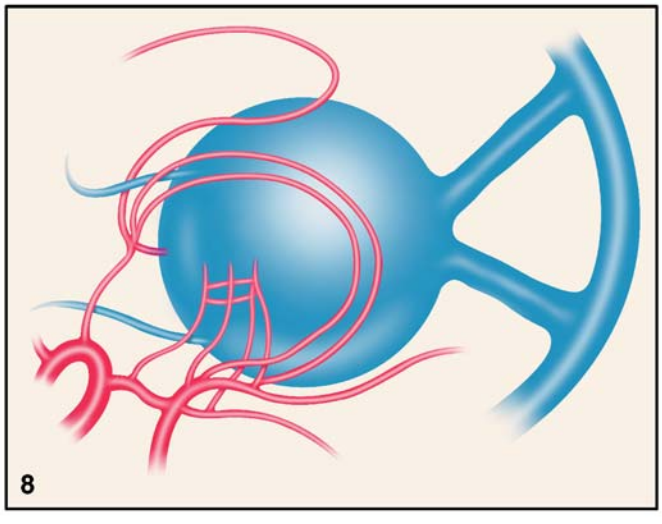
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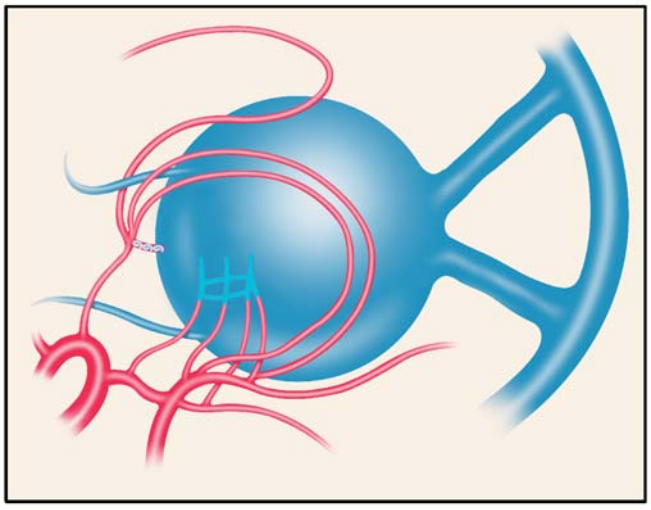

Fig. 9 Large lesion with no individualized anterior pouch. Treatment option: transarterial occlusion with glue in cases of plexus shunt or with coils in cases of large direct shunts. Transvenous occlusion of the pouch with coils would occlude the deep normal venous structures

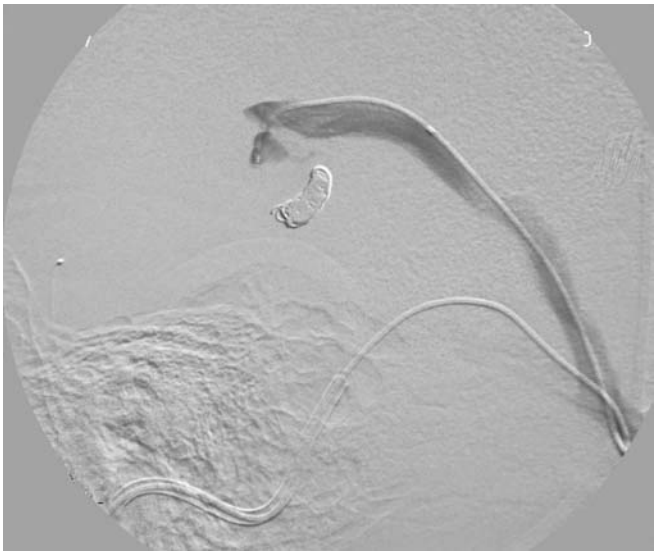

Fig. 10 Transvenous approach of Patient 1, 14 months after the first treatment. The small venous pouch is opacified by in situ contrast medium injection

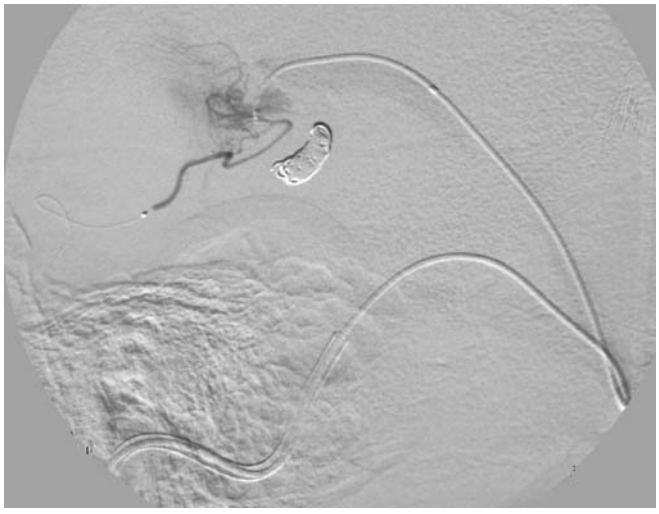

Fig. 11 Superselective injection into collicular arteries feeding the shunt. The venous microcatheter is in the same position in the venous pouch

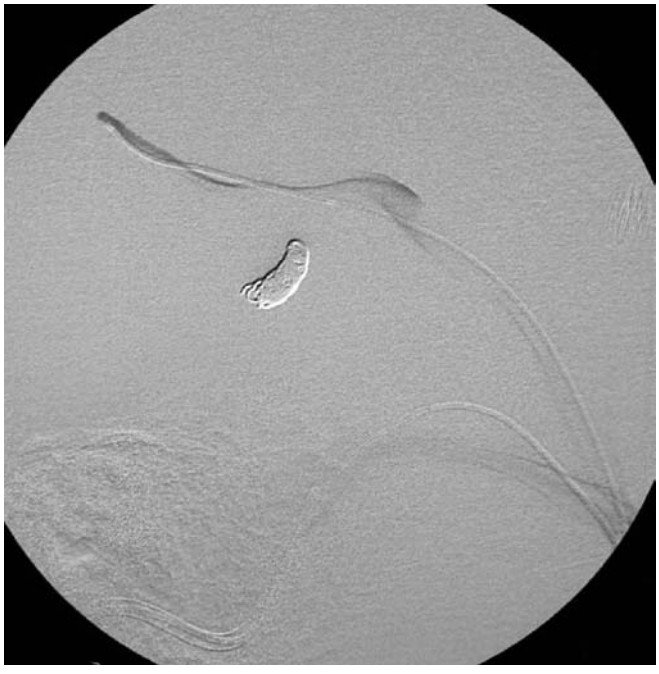

Fig. 12 Superselective retrograde injection into the internal cerebral vein demonstrates the normal anatomy of the deep venous system despite the vein of Galen malformation

may be used during transvenous VGAM therapy in order to help ascertain the absence of normal venous drainage (Figs. 10, 11, 12) in the portion of the venous pouch to be occluded. Our observations may be used to advocate selective treatment of the arteriovenous shunt(s), via either a transarterial or a transvenous approach, without downstream obliteration of the dilated but otherwise normal venous collector. This type of targeted occlusion can be achieved with the endovascular tools presently at our disposal.

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