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Unusual Manifestation of a Vein of Galen Malformation: Value of CT Angiography

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Summary: We describe a neonate with aneurysmal dilatation of the vein of Galen with arteriovenous fistulous sites located at the superior vermian vein. Helical CT angiography was useful for evaluating the anomalous vessels.

Index terms: Aneurysm, computed tomography; Computed tomography, three-dimensional; Vein of Galen

The vein of Galen aneurysm was described by Jaeger et al in 1937 (1). Since then, many investigators have discussed the embryology, anatomy, clinical presentation, and treatment of this anomaly (2). We report a case of a vein of Galen aneurysm presenting as an unusual manifestation, with arteriovenous fistulous sites located at the superior vermian vein. The lesion was analyzed by means of helical computed tomographic (CT) angiography in addition to conventional CT and conventional angiography.

Case Report

A 4310-g infant was born to a 34-year-old mother by cesarean section after a 40-week gestation. The pregnancy was uncomplicated. Apgar scores were 1/3/5 at 1, 5, and 10 minutes. After initial stabilization, the infant was transferred to our institution on the day of birth.

On admission, the head was macrocephalic and suggestive of hydrocephalus, which was confirmed by sonographic examination of the head. Skull percussion and auscultation revealed hyperresonance and cranial bruits. Neurologically, the infant was alert but with poor tone, generalized weakness, and bilaterally fisted hands. Cardiac sonography on the same day revealed a structurally normal heart with severe right ventricular enlargement, increased pulmonary artery pressure, patent foramen ovale, and patent ductus arteriosus. There was right-to-left shunting and a significant diastolic runoff with an aortic steal phenomenon toward the cervical arteries. Further

examination indicated hepatomegaly and pleural effusion. This high-output cardiac failure, together with the hydrocephalus and cranial bruits, was strongly suggestive of a cerebral arteriovenous malformation.

On day 2, the patient underwent a head CT study. An unenhanced CT scan of the head showed hydrocephalus and a large structure of homogeneous high density in the midline. Helical scanning was performed after intravenous injection of a total of 9 mL of contrast material via the dorsal metacarpal vein, with a collimator size of 1 mm, a table pitch of 1:1, and total area coverage of 60 mm. Axial reconstructed images at 1-mm intervals showed strong, uniform enhancement of the aneurysmally dilated vein of Galen as well as a markedly dilated vascular structure, suggesting the superior vermian vein with numerous abnormal vessels in the posterior fossa. The straight sinus was absent. The helical scanning data were transferred to a workstation to reconstruct CT angiograms with a surface-rendering technique and with a maximum intensity projection technique in various projections. CT angiograms showed direct fistulous communication between bilateral superior cerebellar arteries and a markedly dilated superior vermian vein, which emptied into the dilated vein of Galen (Fig 1A–D). The branches of the posterior cerebral arteries and the anterior inferior cerebellar arteries emptied into the dilated superior vermian vein with arterioarterial networks. All fistulas were at the various sites of the superior vermian vein and no arteries drained into the vein of Galen. Anterior or middle cerebral arteries did not contribute as feeders.

On day 3, conventional angiography was performed for transarterial embolization. Selective catheter placement could not be achieved owing to severe tortuosity of the carotid and vertebral arteries. The aortogram with the tip of the catheter at the aortic arch showed arteriovenous fistulas in the posterior fossa (Fig 1E and F). Detailed evaluation of arterial feeders and fistulous sites was difficult owing to the lack of selective catheterization.

Transtorcular embolizations of the arteriovenous fistulas were performed in two sessions, on day 4 and day 7. Despite the treatments, the patient continued to have high-

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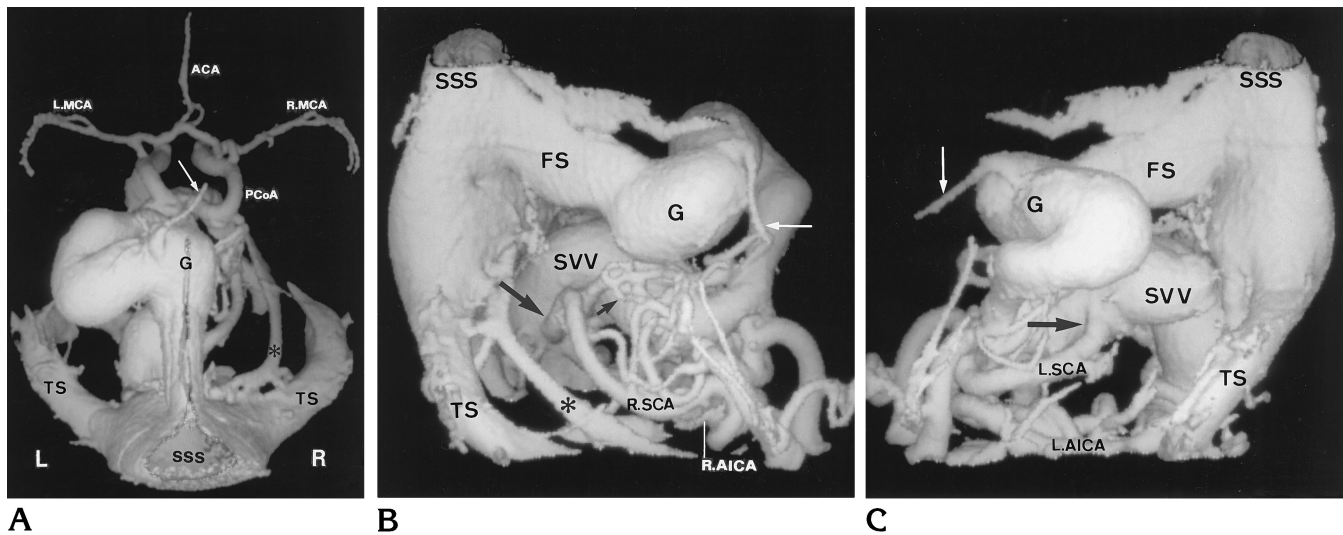
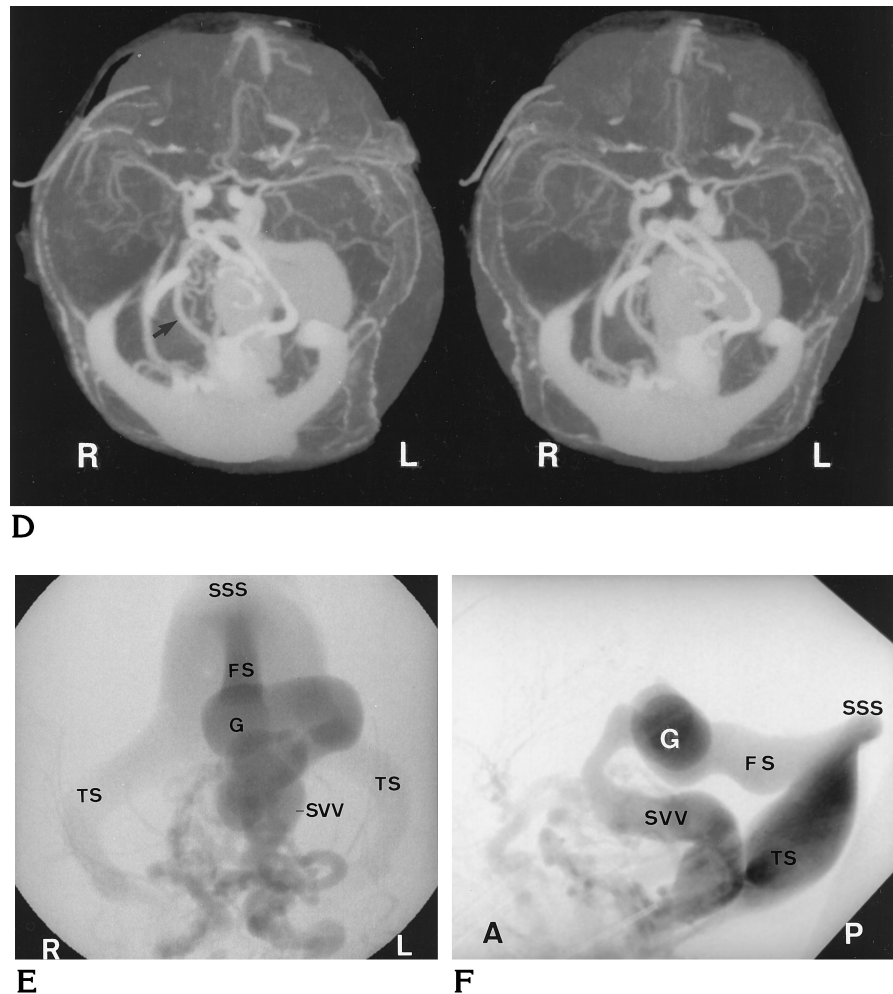


Fig 1. Infant with vein of Galen aneurysm with arteriovenous fistulous sites located at the superior vermician vein.

A–C, CT angiograms with a surface-rendering technique in top-down view (A), lateral view from the right (B), and from the left (C) show direct fistulous communications (*large black arrows*) between a markedly dilated superior vermician vein (SVV) and bilateral superior cerebellar arteries (R.SCA and L.SCA). The vein of Galen (G) is markedly dilated, emptying into the falcine sinus (FS), which drains into the superior sagittal sinus (SSS) and then into the transverse sinus (TS). Small branches of the posterior cerebral arteries with arterioarterial interconnections (*small black arrow*) empty into the superior portion of the dilated superior vermician vein. Tortuous anterior inferior cerebellar arteries (R.AICA and L.AICA), incompletely covered by helical scanning, seem to empty into the distal portion of the superior vermician vein bilaterally. Bilateral posterior communicating arteries (PCoA) are extremely dilated. ACA indicates anterior cerebral artery; R.MCA and L.MCA, right and left middle cerebral arteries, respectively; *white arrow*, internal cerebral vein; and *asterisk*, an anomalous vein connecting the transverse sinus and the inferior petrosal sinus.

D, A stereoscopic pair of CT angiograms obtained with maximum intensity projection technique show numerous feeding arteries in the posterior fossa (*arrow*) and marked dilatation of the superior vermician vein, the vein of Galen, and the torcular. A stereoscopic image from below can be viewed by a crossing-eye method.

Anteroposterior (E) and lateral (F) aortograms with the tip of the catheter at the aortic arch show arteriovenous fistulas in the posterior fossa with the markedly dilated superior vermician vein (SVV) and the vein of Galen (G). There is a prominent falcine sinus (FS) draining the vein of Galen into the superior sagittal sinus (SSS). Dilated torcular and transverse sinus (TS) are also visible.



output heart failure and died on day 8 of disseminated intravascular coagulopathy. Autopsy findings confirmed the arteriovenous fistulas seen radiologically. Posterior inferior and anterior inferior cerebellar arteries were observed communicating directly with the inferior portion of the dilated superior vermian vein, with multiple arterioarterial interconnections on either side. Markedly dilated superior cerebellar arteries were also noted to enter directly the middle portion of the superior vermian vein bilaterally. The posterior cerebral arteries were relatively small compared with the superior cerebellar arteries, although proximal branches of bilateral posterior cerebral arteries entered bilaterally into the superior portion of the dilated superior vermian vein, with arterioarterial interconnections with the superior cerebellar arteries.

Discussion

The term *vein of Galen aneurysm* includes various groups of anomalies with the common feature of aneurysmal dilatation of the vein of Galen. Berenstein and Lasjaunias (3) classified these anomalous arteriovenous shunts involving the vein of Galen into two groups according to their angioarchitectural differences: true vein of Galen aneurysmal malformations and vein of Galen aneurysmal dilatations. Their description of true vein of Galen aneurysmal malformations was based on the study of Raybaud et al (4). Arterial feeders supplying the malformations may include the anterior and posterior choroidal, anterior cerebral, anterior thalamoperforating, perimesencephalic, distal branches of the posterior and middle cerebral, and posterior thalamoperforating arteries. All cases have direct fistulous sites at the vein of Galen aneurysms. Vein of Galen aneurysmal dilatations are caused by parenchymal arteriovenous malformations or dural arteriovenous fistulas draining into a vein of Galen (5, 6).

In our case, direct arteriovenous shunts located in the posterior fossa communicated with a markedly dilated superior vermian vein, which then emptied into the aneurysmally dilated vein of Galen. Previously, a similar case of an arteriovenous fistula draining into the precentral cerebellar vein with dilatation of the vein of Galen was reported by Nelson et al (7) and was defined as a direct nongalenic pial arteriovenous fistula. In his investigation of venous developments in vascular malformations of the posterior fossa, Vidyasagar (8) did not discuss involvement of the superior vermian vein.

The arterioarterial interconnections in these vascular malformations as noted in our patient

have not been well described. Raybaud et al (4) mention that some cases had abnormal perivermian arterial networks, which arise from the posterior inferior cerebellar arteries, the superior cerebellar arteries, and perimesencephalic feeders, but they could not distinguish them further. These arterioarterial interconnections may represent persistence of the early fetal arterial pattern (9).

Magnetic resonance (MR) imaging and MR angiography are excellent noninvasive imaging techniques (10–12); however, they may be difficult to perform in a neonate in an unstable condition. Conventional angiography remains the standard tool for evaluating vein of Galen aneurysms, especially when interventional treatment is planned. However, it is often difficult to see the arterial feeders at the fistulous sites, because of rapid recirculation of contrast material through the fistula and/or wash-out by nonopacified blood (4). CT has been used for screening and evaluation of vein of Galen aneurysms, but it is insufficient for studying the details of the vessels (13). In this particular patient, CT angiography with a surface-rendering technique significantly aided the planning of embolization in regard to the overall anatomy of the arteriovenous fistulas.

In conclusion, in connection with a rare case of a vein of Galen aneurysm with arteriovenous fistulous sites located at the superior vermian vein, we found CT angiography to be an excellent adjunct for understanding the anatomic relationships of the vessels and for aiding presurgical planning.

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