Case Report

Pediatric Neurosurgery

Pediatr Neurosurg 2001;35:149–152

Received: April 18, 2001
Accepted: June 13, 2001

Rapid Normalization of Marked Dilatation of the Cerebral Duro-Venous System in a Newborn Infant Mimicking a Great Vein of Galen Varix

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Key Words
Cerebral venous system · Great vein of Galen · Heart failure · Neonate · Varix

Abstract
A newborn infant with a marked dilatation of the cerebral duro-venous system is presented. The patient was diagnosed as having a vein of Galen aneurysmal varix by a cranial ultrasound examination immediately following delivery. Computed tomographic angiography on the following day, however, showed a marked dilatation of the cerebral duro-venous system, including the great vein of Galen, superior sagittal sinus, torcular herophili and transverse sinuses. There were no arteriovenous fistulas at the vein of Galen. Dilatation of the duro-venous system and concomitant heart failure subsided rapidly after intravenous administration of indomethacin for the treatment of the patent ductus arteriosus on the fourth day of life. Dilatation of the duro-venous system in a newborn infant should be differentiated from any form of vein of Galen aneurysm.

Introduction

Vein of Galen aneurysm is a rare congenital vascular anomaly known as a challenging disease in the neonatal period due to associated severe congestive heart failure. It is classified into two groups: vein of Galen aneurysmal malformation (VGAM) and vein of Galen aneurysmal dilatation (VGAD) [1]. In VGAM, arteriovenous (AV) fistulas are located at the remnant median vein of the prosencephalon [2], and VGAM is further divided into two subgroups according to the locations of the AV fistulas. The choroidal type of VGAM usually has multiple AV fistulas in the velum interpositum cistern and the mural type of VGAM has a single or a few AV fistula(s) on the wall of the dilated aneurysm. VGAD involves a dilated great vein of Galen due to parenchymal AV malformations, dural AV fistulas or varices. Except for VGAD with varices, dilatation of the great vein of Galen occurs due to AV fistulas draining into the great vein of Galen and is frequently associated with steno-occlusive changes in the duro-venous system. To our knowledge, there has been no report of marked dilatation of the cerebral duro-venous system in the first few days of life mimicking a vein of Galen aneurysm upon ultrasound examination. We present such a case with special emphasis on the differential diagnosis from neonatal vein of Galen aneurysms.

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1016–2291/01/0353–0149$17.50/0

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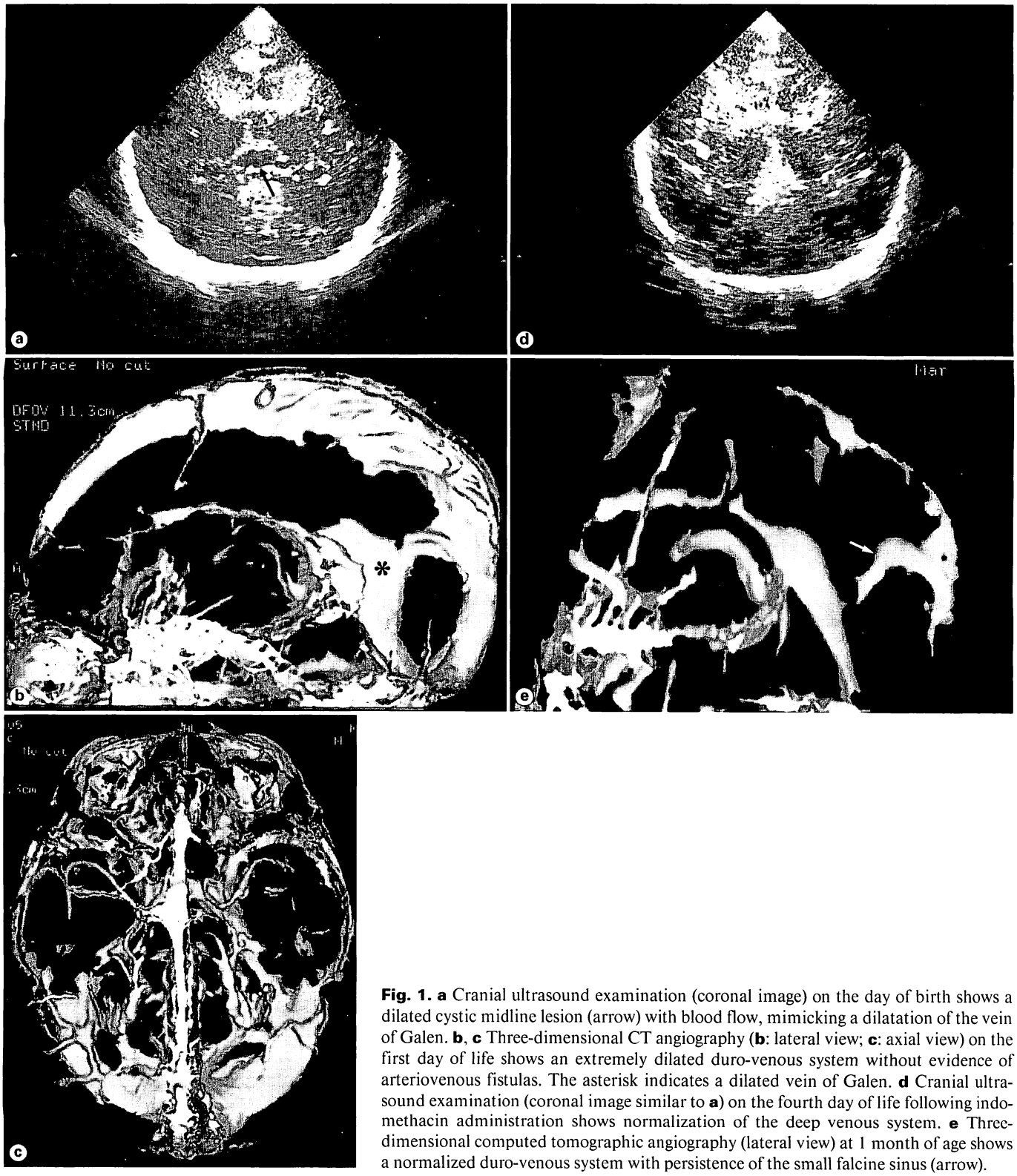

Fig. 1. a Cranial ultrasound examination (coronal image) on the day of birth shows a dilated cystic midline lesion (arrow) with blood flow, mimicking a dilatation of the vein of Galen. **b**, **c** Three-dimensional CT angiography (**b**: lateral view; **c**: axial view) on the first day of life shows an extremely dilated duro-venous system without evidence of arteriovenous fistulas. The asterisk indicates a dilated vein of Galen. **d** Cranial ultrasound examination (coronal image similar to **a**) on the fourth day of life following indomethacin administration shows normalization of the deep venous system. **e** Three-dimensional computed tomographic angiography (lateral view) at 1 month of age shows a normalized duro-venous system with persistence of the small falcine sinus (arrow).

Case Report

This male infant was born by spontaneous vaginal delivery at gestation of 36 weeks and 2 days as the third product of the healthy parents. Birth weight was 1,514 g, which was judged as small-for-dates. The pregnancy was uneventful except for mild genital bleeding during the 33rd week of gestation. Due to being small-for-dates, the patient was transferred to our hospital 1 h and 40 min after delivery. At admission, the patient was neurologically normal. Cranial bruits were not audible and the anterior fontanel was not tense. Cranial ultrasound examination showed a dilated cystic lesion on the midline with blood flow inside, which was interpreted as a great vein of Galen aneurysm (fig. 1a). Blood flow in the cystic lesion was slow on the color Doppler ultrasound examination. Cardiac ultrasound examination showed an enlarged heart, a persistent ductus arteriosus (to and fro motion) and mild mitral and tricuspid valve regurgitation. Chest X-ray showed an enlarged heart with a cardiothoracic ratio of 70%. Cranial computed tomographic (CT) angiography on the following day showed an extensively dilated cerebral duro-venous system, including the great vein of Galen, superior sagittal sinus and bilateral transverse sinuses (fig. 1b, c). The falcine sinus and the straight sinus were patent. Cerebral arterial structures seemed to be normal. Neither hydrocephalus nor AV fistulas were present. This patient was conservatively treated with intravenous catecholamine (dopamine and dobutamine).

On the third day of life, cardiac ultrasound examination showed persistent ductus arteriosus (diameter of 4.8 mm) with a left-to-right shunt, mild mitral and tricuspid valve regurgitation and a dilated superior vena cava (diameter of 6.7 mm). To close the ductus arteriosus, indomethacin (0.2 mg) was given intravenously three times at 12-hour intervals. Follow-up cranial ultrasound examination on the fourth day of life showed rapid normalization of the dilated duro-venous system (fig. 1d). Cardiac ultrasound examination showed closure of the ductus arteriosus. Cardiomegaly subsided spontaneously with a cardiothoracic ratio of 57% on the same day. Cranial CT angiography at 1 month of age showed a normal cerebral venous system except for a small falcine sinus (fig. 1e). Improvement of heart failure paralleled normalization of the dilatation of the cerebral duro-venous system. Magnetic resonance (MR) images obtained at 2 and 6 months and 1.5 and 3.5 years of age showed a normal cerebral venous system without a patent falcine sinus and no brain parenchymal damage or hydrocephalus. The patient's head and chest circumferences were within the normal range at the last follow-up examination at 3 years of age. Development was normal and the patient was neurologically normal throughout the follow-up period.

Discussion

Vein of Galen Varix and Dilatation of the Cerebral Duro-Venous System

Varix is one form of VGAD and it lacks AV fistulas. Lasjaunias [1] described varix of the vein of Galen and separated it into two types. The first type is a transient dilatation of the vein of Galen in neonates presenting as heart failure of another origin. This dilatation persists for a few days and disappears on follow-up ultrasound examination. The dilatation does not lead to any symptoms and its disappearance parallels the cardiovascular improvement. The second type occurs when venous drainage of the brain converges towards the deep venous system. It causes no specific symptoms and is often discovered in adulthood [3].

The former type of vein of Galen varix is quite similar (or identical) to our case, provided that the venous dilatation is not confined to the vein of Galen. In the neonatal period, only ultrasound examination is usually performed because other diagnostic imaging modalities are more invasive. As happened in our case, cranial ultrasound examination may fail to detect a dilatation of the peripherally located superior sagittal sinus and transverse sinuses. We believe that with careful observation, it may be possible to detect a dilated duro-venous system by cranial ultrasound examination. It is misleading to describe an ectatic venous system as a varix of the vein of Galen; instead, it should be termed a dilatation of the cerebral duro-venous system.

In the past 6 years, we have treated 776 newborn infants whose birth weight was less than 1,500 g. All had undergone cranial and cardiac ultrasound examinations on the day of birth as part of a routine workup. The patient presented here was the only one with a dilated cerebral duro-venous system, indicating an incidence of 0.13%.

Differential Diagnosis

In the neonatal period, cerebral ultrasound examination is a prerequisite for the detection of intracranial pathologies. A cystic lesion might be an arachnoid cyst, porencephaly or a vein of Galen aneurysm [4]. Vein of Galen aneurysm can be detected as a cystic lesion in the center of the brain, exhibiting rapid and/or turbulent blood flow within [5]. Arachnoid cysts and porencephaly are differentiated by the absence of blood flow. When the whole cerebral duro-venous system is dilated, peripherally located dural dilatation can easily be missed by this examination, as happened in our case. The presence of AV fistulas can be diagnosed by color Doppler sonography detecting high blood flow and turbulence in the neonatal period. In addition to ultrasound, CT and MR imaging are also less invasive diagnostic modalities than catheter angiography. CT and MR imaging can show hydrocephalus and brain parenchymal changes, such as infarction, hemorrhage or atrophy. CT is sensitive to calcification. In our case, there were no abnormal parenchymal changes, AV fistulas or enlarged feeding arteries. The presence of AV fistulas and dilated feeding arteries may

contribute to differentiating a dilated venous system from VGAM and VGAD.

Treatment

Indomethacin is used to close a patent ductus arteriosus in the early neonatal period. It is known that intravenous indomethacin causes a reduction in the cerebral blood flow velocity and increases cerebral arterial blood pressure, indicating cerebral vasoconstriction [6]. These effects last as long as 90 min after the administration of indomethacin. Although indomethacin was effective in closing the patent ductus arteriosus in our patient, its effect on the cerebral vasculature seemed to be temporary. Thus, it is less likely that indomethacin caused normalization of the cerebral venous sinus dilatation directly.

VGAM in neonates requires proper intervention with optimal timing, which is occasionally performed on an emergency basis [3]. Even when the diagnosis of a dilated duro-venous system is established in the early neonatal period, we believe that no treatment is required and that close follow-up with ultrasound examination is necessary. Our case with a 3-year follow-up suggests that a dilated duro-venous system in newborn infants without AV fistulas is benign with a good prognosis and requires no treatment. However, it is necessary to accumulate clinical data on similar cases to elucidate the nature of this clinical entity.

In conclusion, neonatal dilatation of the duro-venous system without AV fistulas should be differentiated from any form of vein of Galen aneurysms.

References

1 Lasjaunias P: Vein of Galen aneurysmal malformation; in Lasjaunias P (ed): Vascular Diseases in Neonates, Infants and Children: Interventional Neuroradiology Management. Berlin, Springer, 1997, pp 67–202.

2 Raybaud CA, Strother CM, Hald JK: Aneurysms of the vein of Galen: Embryonic considerations and anatomical features relating to the pathogenesis of the malformation. Neuroradiology 1989;31:109–128.

3 Lasjaunias P, Rodesch G, Terbrugge K, Pruvost P, Devictor D, Comoy J, Landrieu P: Vein of Galen aneurysmal malformations. Report of 36 cases managed between 1982 and 1988. Acta Neurochir (Wien) 1989;99:26–37.

4 Pilu G, Falco P, Perolo A, Sandri F, Cocchi G, Ancora G, Bovicelli L: Differential diagnosis and outcome of fetal intracranial hypoechoic lesions: Report of 21 cases. Ultrasound Obstet Gynecol 1997;9:229–236.

5 Tessler FN, Dion J, Vinuela F, Perrella RR, Duckwiler G, Hall T, Boechat MI, Grant EG: Cranial arteriovenous malformations in neonates: Color Doppler imaging with angiographic correlation. AJR Am J Roentgenol 1989;153:1027–1030.

6 Mardoum R, Bejar R, Merritt TA, Berry C: Controlled study of the effects of indomethacin on cerebral blood flow velocities in newborn infants. J Pediatr 1991;118:112–115.