Remote congenital cerebral arteriovenous fistulae associated with aortic coarctation

Case report


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A neonate presented with anatomically discrete cerebral arteriovenous fistulae located in the right sylvian fissure and the cerebellar vermis that were initially detected by prenatal ultrasonography. Following delivery of the baby by Caesarean section, both malformations were treated by surgical obliteration. These intracranial vascular lesions were associated with cardiac anomalies and a peri ductal coarctation of the aorta, which was treated with a left subclavian rotational arterial pedicle repair. Follow-up examination of the infant at age 13 months demonstrated an excellent clinical result with normalization of the circulation. The pathophysiology of this syndrome is discussed and the literature reviewed.

KEY WORDS • arteriovenous fistula • neonate • varix • aortic coarctation • prenatal ultrasound

ARTERIOVENOUS fistulae associated with giant varices are uncommon cerebrovascular malformations.3,27,28 The mode of presentation of these and other giant cerebrovascular malformations is related to the age of the patient. Cardiac failure, which may occur following the physiological changes in circulation after birth, is a frequent presentation in neonates with arteriovenous fistulae, often related to a poor prognosis.2,4,6-9,11,15,16,25,26 In the pediatric group, symptoms of raised intracranial pressure due to hydrocephalus, which may be associated with venous hypertension, are common.5,8,10 Other less frequent modes of presentation are mass effect or subarachnoid hemorrhage.1 Multiple cerebrovascular malformations are also uncommon and may coexist with inherited disorders such as Rendu-Osler-Weber syndrome or Wyburn-Mason syndrome.13,22,24,29 We report an isolated case of multiple intracranial arteriovenous fistula occurring in a neonate, associated with cardiac anomalies and peri ductal coarctation of the aorta.

Case Report

This 20-day-old baby boy was referred to our institution for evaluation of multiple congenital arteriovenous fistulae. The child was born by repeat lower-segment Caesarean section at 40 weeks' gestation, the third child of a 29-year-old Asian woman. Birth weight was 3715 gm. Prenatal ultrasonography performed at 35 weeks' gestation for assessment of uterine size and date mismatch, initially revealed cystic abnormalities in the right sylvian fissure and posterior fossa. Following delivery, Apgar scores were 7 and 8 at 1 and 5 minutes, respectively. General examination revealed that the baby was in good health with no obvious cardiac anomalies or signs of heart failure; however, a discrepancy between upper and lower limb blood pressures was noted. Neurological examination was normal, with a soft open anterior fontanel and overlapping sutures. Bruits were not audible on auscultation of the skull. Computerized tomography scans with contrast enhancement suggested the presence of multiple intracranial vascular malformations. Renal ultrasound studies were normal. Cardiac anomalies were demonstrated by echocardiography but a distinct coarctation was not seen. The infant was transferred to our institution.

Examination. Transfemoral cerebral angiography revealed two anatomically discrete arteriovenous fistulae, one located in the right sylvian fissure and the other
Right common carotid angiograms, lateral (A) and anteroposterior (B) views, showing an arteriovenous fistula located in the right sylvian fissure. Note the tortuous and elongated course of the enlarged internal carotid artery. There is backward and upward displacement of the middle cerebral artery (arrowheads), and a large anterior temporal branch (arrows) is shown feeding a large varix located within the proximal portion of the right middle sylvian fissure.

Close to the midline in the posterior fossa. The former consisted of a single arterial feeder arising from the anterior temporal branch of the right middle cerebral artery and filling a large varix with multiple draining veins. The right cervical carotid artery was large and tortuous, and the supraclinoid portion of the carotid artery had a high bifurcation and was displaced by the large varix (Fig. 1). The feeding branch of the right middle cerebral artery had a 3.6-mm lumen, and the venous pouch had a maximum diameter of 31 mm. The major drainage from the varix was via a large vein of Trolard and enlarged inferior temporal veins that ultimately entered the transverse sinus (Fig. 2). A smaller component of the venous drainage occurred via the cavernous sinus.

The posterior fossa complex involved two juxtaposed arteriovenous fistulae, each with a 2-mm diameter and a large midline varix (33 mm in maximum diameter) with multiple draining veins. The flow patterns observed in this varix in the early arterial phase suggested that the feeding arteries entered the venous pouch separately rather than via a common opening (Fig. 3A). The two arterial feeders arose from the intracranial portion of the right vertebral artery at the level of the foramen magnum and had a parallel course before separately joining the venous pouch. There was a major collateral supply to the rostral feeding artery in a retrograde fashion from the left vertebral artery. Similarly, the caudal feeding vessel received a minor retrograde collateral supply from the left vertebral artery. The venous drainage involved both posterior mesencephalic veins and a large right superior cerebellar vein. The latter drained into the ipsilateral sigmoid sinus through the right superior petrosal sinus while the posterior mesencephalic veins drained into the straight sinus (Fig. 3B).

An ultrasound examination of the intracranial contents with color-flow duplex evaluation demonstrated supra- and infratentorial vascular malformations. The former was a 2.6-cm mass, filled with swirling flowing blood in the right frontal region near the sylvian fissure. A vessel with an arterial waveform was demonstrated just posterior to this mass. A smaller right paramedian mass located in the posterior fossa was also filled with swirling blood.

Two-dimensional echocardiography was performed and revealed: 1) moderate coarctation of the aorta in the juxtradiuctal region, with a Doppler ultrasound gradient of 36 mm Hg; 2) paramembranous ventricular septal defect with left-to-right shunting; 3) enlarged brachiocephalic arteries and an enlarged innominate vein; 4) increased flow in the superior vena cava; and 5) mild tricuspid regurgitation. The left ventricular ejection fraction was 68%.

Operations. One week following cerebral angiography, endovascular occlusion of the anterior circulation fistula was attempted via a transfemoral approach under general anesthesia. Due to technical difficulties and the patient developing hypothermia and mild acidosis, the procedure was terminated. Anemia associated with blood loss during the attempted occlusion was corrected with transfusions of packed red blood cells and the baby made a satisfactory recovery. Two days later, a right frontotemporal craniotomy was performed. The single arterial feeder was identified at the entrance to the varix and clipped with a miniature Sugita clip. In order to ensure fistula thrombosis, the venous pouch was punctured with a No. 31 needle and approximately 8 m of 2-mm beryllium-copper wire was then placed in the varix using the Mullan technique.
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Postoperatively, there was no neurological deficit; however, congestive cardiac failure developed and was treated with digoxin and furosemide administration. On the first day after surgery, the baby developed a seizure disorder associated with apneic episodes. The seizures were characterized by episodes of tongue jerking and movement of the eyes up and to the left. Treatment included reintubation and mechanical ventilation and control of seizure activity with phenobarbital therapy. Laboratory investigations revealed a serum sodium level of 119 mM/liter; the hyponatremia was corrected by inducing a mild diuresis, fluid restriction, and use of hypertonic saline. In the following days, the baby made a satisfactory recovery and was discharged on the 10th postoperative day on a maintenance course of digoxin and phenobarbital.

At 3 months of age, surgical treatment of the posterior fossa fistula was performed through a right suboccipital craniectomy. Both feeding arteries were occluded with miniature Sugita clips, the venous pouch was punctured, and 8 m of beryllium-copper wire was introduced into the varix. There were no neurological complications postoperatively. Marked hypertension required the use of a combination of intravenous antihypertensive agents in the initial postoperative period. Propanolol was continued as maintenance oral antihypertensive therapy. On the 4th day postoperatively, cyanotic episodes occurred, which were attributed to sepsis and treated with broad spectrum antibiotic coverage provided by a third-generation cephalosporin. Marked clinical improvement followed. Escherichia coli, sensitive to the cephalosporin, was later isolated from blood cultures. Although not proven by culture, the primary source of infection was considered to be a central line site in the right groin. Antibiotic therapy was continued for an additional 21 days.

Because arterial hypertension persisted during the 14 days following suboccipital craniectomy despite adequate antihypertensive therapy, the status of the aortic coarctation was assessed with angiography and repeat two-dimensional echocardiography. Both studies showed that the coarctation had progressed; therefore, the periductal coarctation was repaired using a left subclavian rotational arterial pedicle.

Postoperative Course. Postoperatively, the patient made a rapid recovery without complication, and his blood pressure returned to normal. Follow-up echocardiography showed the widely patent coarctation repair. A sinus venous atrial septal defect with partial anomalous pulmonary venous connection to the superior vena cava was also detected. The infant was discharged with no maintenance medication.

Cerebral angiograms were not obtained in the immediate postoperative period. However, right vertebral and right carotid angiography 10 months following occlusion of the posterior fossa fistulae demonstrated that the supra- and infratentorial fistulae had been successfully occluded (Fig. 4). In addition, the deep venous drainage had remained patent following occlusion of both fistulae. In the anterior circulation, the superficial temporal venous drainage was not demonstrated, while the vein of Trolard remained patent although reduced in caliber. In the posterior fossa, the superior cerebellar vein was not demonstrated, while the deep venous drainage to the vein of Galen via the posterior mesencephalic veins remained patent. Physical examination at this time revealed normal progression of growth and appropriate achievement of developmental milestones. Neurological examination was significant only for slightly decreased prehension in the left upper extremity.
Discussion

Multiple cerebrovascular malformations occur infrequently, and multiple arteriovenous fistulae are rare. In our case, physical examination and family history did not suggest an association with inherited disorders such as Rendu-Osler-Weber or Wyburn-Mason syndrome, in which multiple cerebrovascular malformations may occur. Willinsky, et al. observed that, when multiple cerebral malformations occurred, they were located either in surface structures (as was seen in our case with both fistulae related to pial surfaces) or in deeper structures, but rarely in both sites. Multiple vascular malformations are most frequently diagnosed in adult life; however, in our case the lesions were initially detected in the prenatal period.

Diagnosis of Arteriovenous Fistulae

Prenatal diagnosis of central nervous system pathology has been improved by the routine use of ultrasound studies during pregnancy. Fetal midline vascular malformations, principally vein of Galen anomalies, can be accurately diagnosed with cerebral ultrasonography. The diagnosis of these vascular lesions is suggested by the detection of pulsations and associated intracranial pathology such as hydrocephalus. In the case presented, failure to correctly diagnose the intracranial pathology detected on prenatal ultrasound studies may be explained by the peripheral location of the lesions and the lack of detectable pulsations during the examination. Healthy preterm infants studied by Grant, et al., using cranial duplex sonography showed very weak signal intensities and significantly lower average velocities in all major cerebral arteries as compared with those in term infants. The authors also noted that these studies were often technically difficult to perform and concluded that the use of duplex sonography did not improve diagnosis when compared with real-time scanning, the method used in our case. Duplex sonography can, however, permit better evaluation of cerebrovascular flow than does nonpulse-gated, nondirected Doppler ultrasonography. While the color-flow duplex examination performed after birth correctly identified the location of both supra- and infratentorial vascular malformations in our patient, it did not provide specific information as to the exact nature of the lesions.

Radiological evaluation of arteriovenous fistulae includes cross-sectional imaging and angiography. The former provides evidence of hemorrhage and associated pathology such as hydrocephalus and thrombosis. Angiography is required to anatomically define the fistulae. The site of an arteriovenous fistula is suggested by the abrupt change that occurs in the caliber of the vessel, indicating the transition between artery and vein. Initially, the vein may be grossly dilated and form a varix. Additional information, such as the presence of collateral vessels, may be obtained using supraselective angiography and/or temporary balloon occlusion. If initial radiological investigations do not differentiate clearly between arteriovenous malformations and arteriovenous fistulae, it is vital that further studies be performed as the two diagnoses require different surgical treatment strategies.

Treatment of Arteriovenous Fistulae

The best results in treating arteriovenous malformations are obtained with complete excision, while the preferred treatment of arteriovenous fistulae is obliteration of the fistulae or interruption of all feeding vessels as close to the fistula as possible while leaving the venous drainage intact. The introduction of wire into the venous pouches was described by Mullan in the treatment of carotid-cavernous fistulae. This additional procedure was performed to maximize the chance of thrombosis if additional smaller arterial feeders were present that were not visualized angiographically. In retrospect, this may not have been necessary in our case, and the contribution of this maneuver to the promotion of thrombosis in the venous pouch cannot be determined. The presence of this wire limits radiological examination as further magnetic resonance imaging is prohibited. In addition, extensive thrombosis of the venous drainage beyond the venous pouch could have occurred, had the wire actively promoted thrombogenesis. In our case, both fistulae have been successfully obliterated and extensive thrombosis of draining veins, including cortical venous thrombosis and hydrocephalus, has not occurred.

Cardiac Considerations

Congestive heart failure, a frequent form of presentation in patients with arteriovenous fistulae in the neonatal period did not occur in our case until after the occlusion of the middle cerebral artery fistulae. The development of cardiac failure soon after birth is ascribed to changes in the neonatal circulation. According to Cumming, circulatory overload before birth is shared between the two cardiac ventricles, with the left ventricle totally supplying the arteriovenous fistula and the upper half of the body while the right ventricle supplies the placenta and lower half of the body. If the usual changes occur after birth, the entire circulation must be sustained by each ventricle, so that the burden on each ventricle is increased and cardiac failure can ensue. The development of medically controlled heart failure following the first craniotomy performed on our patient may be attributed to an increase in systemic resistance associated with occlusion of one of the fistulae in the face of pre-existent aortic coarctation. The severe hypertension observed after the occlusion of the second fistula, in a setting of controlled heart failure, may be attributed to a further increase in systemic vascular resistance. In addition, the hypertension may have had

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a central etiology, perhaps due to transient compression or distortion of the medulla by the enlarged thrombosed venous pouch. Although an increase in the size of the varix was not documented radiologically, enlargements of other isolated vascular structures, such as giant aneurysms, following thrombosis is well known.

Embryogenesis of Coarctation

The association of aortic coarctation with the arteriovenous fistulae seen in our case may also have a hemodynamic explanation. Cumming speculated that, in the developing fetus with an intracranial fistula, a reduction in descending aortic flow may predispose to the development of coarctation or hypoplasia of the isthmus of the aorta. Although the presence of pathological intracranial fistulæ may be an etiological factor in aortic coarctation, the hemodynamic changes associated with birth and obliteration of the fistulæ may be responsible for the rate at which changes develop in the affected segment of the aorta, which in our case occurred over a period of several months.

Conclusions

The interest of this case lies in the prenatal detection of intracranial abnormalities that ultimately led to the diagnosis of rare anatomically separate congenital cerebral arteriovenous fistulae. The fistulæ were successfully repaired surgically without the attendant complications of symptomatic cerebral vein thrombosis and hydrocephalus sometimes associated with repair of such malformations. The differential diagnosis of intracranial cystic structures detected on prenatal ultrasound examination should include a vascular malformation such as an arteriovenous fistula with an associated venous pouch. The importance of supraselective angiography for the accurate diagnosis and location of the fistulae is reflected in the successful outcome of this case, achieved by simple obliteration of the fistulae rather than extensive resection of both malformations.

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References


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