Giant aneurysm arising from a single arteriovenous fistula in a child

Case report

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A case is presented of a child with an arteriovenous fistula and a giant aneurysm located beside the brain stem under the right temporal lobe. It was successfully treated by clipping its feeding artery, a branch of the right posterior cerebral artery. The similarities to, and the differences from, aneurysms of the vein of Galen are discussed.

KEY WORDS — arteriovenous fistula □ arteriovenous malformation □ aneurysm □ varix □ vein of Galen

CAROTID-CAVERNOUS fistulas are usually seen in adults; however, fistulas from various cerebral arteries that drain into the vein of Galen and straight sinus are most common in children. Less frequent presentations of arteriovenous (AV) fistulas include abnormal communications between intracranial arteries and dural sinuses or fistulas at extracranial levels. We present an unusual case of a child with a single AV fistula from a branch of the right posterior cerebral artery (PCA) with a giant aneurysm between the arterial and venous components.

Case Report

This 4½-year-old boy was admitted in October, 1982. He was born by Caesarean section. His parents had had four other children, two of whom had died from unknown causes. Our patient was hospitalized for the first time at the age of 11 months with bronchitis. At that time his head circumference was found to be above normal limits. A bruit was audible over a wide open anterior fontanel, but no heart murmur could be detected. The rest of the neurological examination was unremarkable. Chest x-ray films showed enlargement of the heart, but there were no hemodynamic anomalies. A computerized tomography (CT) brain scan and a right carotid angiogram were reported as normal. Follow-up examination at the age of 16 months showed the head circumference to be in the 97th percentile.

Examination. At the present admission, a CT brain scan was abnormal and the patient was referred for a neurosurgical consultation. His psychomotor development was normal, and his head circumference was 53.3 cm (within normal range). A bruit was audible over the right temporal region. A systolic cardiac murmur was present, but there were no other signs of cardiac malfunction. The liver was enlarged. Blood pressure was 85/60 mm Hg.

Chest films and cardiac echoencephalograms revealed enlargement and hypertrophy of the heart. A CT scan showed an area of increased density in the right temporal region that enlarged and enhanced after intravenous administration of contrast medium. The ventricles were normal in size and shape, with no displacement. Blood and urine analyses, including a hematogram, were within normal limits. Bilateral carotid and right vertebral angiograms (Fig. 1) showed a giant aneurysm (2.6 cm in diameter), to the right of the brain stem, under the temporal lobe. It was fed from a branch of the P1 segment of the right PCA and drained through a dilated vein into the torcular Herophili. Abnormal vessels in the interpeduncular fossa were visualized at the venous phase.

Operation. On October 20, 1982, the child underwent right temporal craniotomy under general anesthesia and in a lateral recumbent position. After the bifurcation of the basilar artery was identified and the P1
FIG. 1. Preoperative right vertebral angiograms, lateral (upper) and anteroposterior (lower) views. Upper Left: A giant aneurysm is seen, with a feeding artery from the right posterior cerebral artery (PCA) and a vein draining toward the torcular Herophili. Abnormal vessels are seen at the interpeduncular fossa. Upper Right: Enlarged view illustrating the feeding artery (lower arrow) and the draining vein (upper arrows). Lower Left: The aneurysm is shown lateral to the right PCA. Lower Right: Enlarged vein illustrating the feeding artery (single arrow) and the draining vein (two arrows).

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The venous system without abnormal interconnecting vessels; that is, an AV fistula. This is a rare lesion. In adults the most common form is a carotid-cavernous fistula. The incidence of these lesions has been estimated as 8.6% of that of subarachnoid saccular aneurysms and 7.1% of that of cerebrovascular malformations. Most carotid-cavernous fistulas are post-traumatic in origin; this type is three times as common as the spontaneous variety. Dural malformations form another unusual group of AV fistula.

In the pediatric age group, Galenic system malformations are often referred to as aneurysms of the vein of Galen. They constitute abnormal communications between branches of the vertebral and/or carotid arterial systems and the vein of Galen which cause a secondary dilatation of the latter. The exact incidence of AVM’s among children is unknown, but the number of diagnosed and treated cases is fewer in the pediatric age group than in the adult. Aneurysms of the vein of Galen are even more uncommon. Amacher and Shillito categorized these lesions into four clinical groups based on the time of onset of symptoms. They included 22 of their cases discovered at infancy in Group 3; 16 of these patients presented with cranio-megalgy and cranial bruit, two with mild heart failure, and the remaining four with other symptoms; significant cardiomegaly was noted on seven chest films. The patients they classified in Group 4 came under medical care later because of headaches, episodes of exercise syncope, or subarachnoid hemorrhage. Management of aneurysms of the vein of Galen is difficult and has a high mortality risk.

It is important to differentiate AVM’s from AV fistulas since treatment should be different. For AVM’s the best results are obtained with complete excision; however, for AV fistulas the preferred treatment is interruption of all feeding arteries as close to the fistula as possible, leaving the venous drainage intact. Although our patient did not have an aneurysm of the vein of Galen, the case is similar because of age of presentation, symptomatology, and the fact that the lesion consisted of an AV shunt with a giant dilatation or aneurysm-like formation. In our case, however, the location was lateral instead of midline, the vein of Galen was not involved, and the single feeder emptied directly into the aneurysm, which in turn drained through a vein into the torcular; Galenic malformations are usually supplied by one or more arteries that divide into many branches before shunting into the vein of Galen.

Drake mentioned two cases described as “huge varices from a single AV fistula” in a series of 166 AVM’s. One was a 20-year-old patient complaining of headaches, whose varix arose from the left posterior communicating artery, filling the third ventricle, and draining into the vein of Rosenthal. The base of the varix was clipped, preserving the posterior communicating artery, and a good result was achieved. The other case was that of a 30-year-old patient with a varix that had...
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FIG. 2. Postoperative right vertebral angiograms performed on March 14, 1983, documenting the disappearance of the preoperative abnormalities. Upper: Lateral view, arterial phase (left) and venous phase (right). Lower: Anteroposterior view, arterial phase (left) and venous phase (right).

eroded through the skull. Occlusion of the huge feeding branch of the middle cerebral artery resulted in immediate collapse of the sac without any problems; however, the patient became comatose and died 24 hours later from a massive intracerebral hemorrhage unrelated to the varix but due to hemodynamic alterations.

Schijman and Monges reported a giant AV aneurysm of the posterior fossa in a 3-month-old infant. There was no cardiac failure. The lesion was thrombosed, did not fill on angiography, and produced intracranial hypertension. The aneurysmal sac, complete with an afferent artery and an efferent vein, was resected in one block. Three years after surgery the child’s progress was good, but there was moderate disability.

Our 4½-year-old patient was successfully treated by craniotomy and clipping of the arterial side of the fistula close to the aneurysm sac.

References


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