Case Reports and Technical Notes

Excision of Thrombosed Vein of Galen Aneurysm in an Infant

Case Report

B. K. A. Weir, M.D., P. B. R. Allen, M.D., and J. D. R. Miller, M.D.
Division of Neurosurgery, University of Alberta Hospital, and Department of Radiology,
University of Alberta Hospital, Edmonton, Alberta, Canada

Over 50 vein of Galen malformations have now been reported. A few cases have been successfully treated by ligation of feeding vessels. To our knowledge, only one case has survived excision of the malformation, and this was in a young adult. This is the first reported case of a completely thrombosed vein of Galen aneurysm in which total excision was successfully accomplished.

Case Report

This male child was born following a 39-week gestation and weighed 5 lbs 15 oz at birth. His development and head circumference were normal when seen in follow-up at age 7 weeks. At age 2 months he had developed some head control but he gradually lost this by age 4 months.

First Admission. On December 19, 1966, at the age of 4 months, the patient was admitted to hospital because of a 3-day illness characterized by irritability, failure to feed, and vomiting. The mother noted that the head appeared to be larger and the fontanel was bulging. Bilateral subdural taps gave no evidence of subdural hematomas. A ventriculogram performed in conjunction with a lumbar pneumoencephalogram showed an aqueductal stenosis. The ventricular cerebrospinal fluid had a protein of 25 mg %. Chest x-ray was normal. There was no evidence of cardiomegaly or cardiac failure. On December 28, a right ventriculostional shunt was carried out. The child ran a low-grade temperature prior to and for a week following the shunt. This cleared spontaneously. The child's symptoms disappeared following the operation and the fontanel became soft.

Second Admission. The child was readmitted 1 month later because of recurrent lethargy, vomiting and loss of appetite associated with increasing tension of the anterior fontanel and mild fever. The head circumference was 17 1/2 in., which was 1/2 in. less than at the first admission. The shunt was felt to be working normally, and the child was discharged improved without any specific therapy. Again at this admission there was no evidence of cardiac failure and no bruit.

Third Admission. On April 2, 1967, at the age of 7 months, the child was admitted with symptoms of drowsiness, vomiting, and a bulging fontanel. The head circumference was 17 3/4 in. There were no bruits. The child did not have head control, had never sat alone and had bilaterally increased knee jerks and sustained ankle clonus. The ventriculoatrial pump was found to be functioning well but plain skull films showed the development of a spherical calcification in the region of the posterior margin of the third ventricle. This had not been present on the films taken 3 months before. The child had a generalized seizure while in hospital and was placed on Dilantin. Ventriculography was carried out; ethiodan was injected into the right lateral ventricle (Fig. 1). The spherical calcification was 2.2 cm in diameter and compressed the aqueduct from above. The obstruction to the aqueduct was complete and the lateral and third ventricles were enlarged. A left carotid angiogram showed that both pericallosal arteries filled from the left and were displaced upward. The anterior choroidal, posterior communicating and posterior cerebral arteries were all filled (Fig. 2). In the venous phase there was excellent filling of the cortical veins as well as the inferior anastomotic vein, the superior sagittal

Received for publication February 5, 1968.
sinus, the transverse and sigmoid sinuses. Despite several injections, however, the internal cerebral vein, the vein of Galen and straight sinus were never satisfactorily shown, and it was felt there was some obstruction to the deep venous system. The calcified mass in the posterior end of the third ventricle was again seen and did not appear to be an arteriovenous malformation.

Operation. On April 25, 1967, when the child was 8 months old, an occipital craniotomy was carried out. A bioccipital bone flap was raised but the dura was only incised on the left side of the falx and along a line parallel to and just superior to the transverse sinus. One cortical vein passing into the superior sagittal sinus had to be coagulated and cut. Excellent exposure of the lesion was then obtained. The posterosuperior aspect of the mass was contiguous with the falx. Anteriorly and superiorly the corpus callosum was draped over it. Using blunt dissection the entire lesion was surrounded by cottonoid strips and cut off the falx after use of silver clips and coagulation. At its anterior pole the lesion was connected with a confluence of veins including the internal cerebral, basal and superior cerebellar. A few small arteries apparently entered the capsule.

Fig. 1. Positive contrast ventriculogram, anteroposterior (left) and lateral (right) views, showing the spherical mass with a calcified rim situated posteriorly in the midline, obstructing the aqueduct and compressing the suprapineal recess. The shunt tube is visualized, with some positive contrast material seen in the reservoir. The sutures are split.

Fig. 2. Cerebral arteriogram, arterial phase, anteroposterior (left) and lateral (right) views. The anterior cerebral arteries are in the midline but are bowed upwards considerably while the posterior occipital branches of the posterior cerebral artery are displaced slightly downwards and posteriorly. The middle cerebral artery is bowed outwards. There is residual ethiodan in the occipital horns. The arterial findings are compatible with a marked obstructive hydrocephalus.
Vein of Galen Aneurysm

These were clipped and divided and at this point the lesion, which was solid, was incised and the contents removed. It contained grayish-red and pale gray tough thrombus. The anterior pole of the lesion was then cut circumferentially and the veins entering it were clipped. The bulk of the aneurysm could thus be removed leaving a dry cavity. The dura was closed and the bone flap wired in place. No drains were used.

Postoperative Course. The child was awake, alert, and moving all extremities 2 hours after the termination of anesthesia. The pupils were equal and reactive. The following day, however, the patient went into status epilepticus and had a period of respiratory distress necessitating intubation. It was necessary to use a respirator until the third postoperative day; at this time a tracheostomy and gastrostomy was carried out. The child was generally spastic. The pupils had remained equal and reacting but there was a divergent strabismus. There was no response to painful stimulation. Generalized edema developed on the fifth postoperative day. One week postoperatively the child was making no spontaneous movements, and the eyes were kept shut. During this period there had been a persistent tachycardia of 180–190 per min.

During the next 2 weeks the clinical picture gradually changed. The spasticity lessened, and the child spontaneously opened his eyes and began to make purposeful movements. He did not follow objects or blink when threatened. The gastrostomy and tracheostomy tubes were removed on May 22. When discharged on June 2, 1967, he was alert and eating normally. Head circumference was 17 3/4 in. When seen at follow-up 8 months postoperatively the child had minimal head control and only appeared to follow very bright lights. His head was normal in size for his age. He was moving all extremities well.

Pathological Report. Grossly, the specimen consisted of a fibrous capsule more than 2 cm in diameter which was pale yellow in color. Attached to the inner surface were spherical masses of friable brownish-red clot. Section showed the capsule to consist of the wall of a venous aneurysm. There were numerous deposits of calcium throughout the different layers.

Discussion

All reports on vein of Galen malformations were reviewed by Gold, et al., in 1964.8 Thirty-four cases had been previously described, and they added to this list eight cases from the Columbia Presbyterian Medical Centre in New York. All eight of their patients died. They reviewed 19 cases in children, all of which had evidence of hydrocephalus. Seven of the 19 had convulsions. Eight of these 19 patients are known to have had an operation and four of them survived it; the ages at operation were 11 months, 21 months, 21 months, and 4 years. Three of the patients were retarded, and the data are inadequate to judge on the fourth. Lehman, et al.,9 reported the case of a 5-month-old child who died at operation. All of Gold's nine cases of vein of Galen malformations in newborn children resulted in death. Our patient appears to be the youngest to have survived operation. As in the other cases, it also seems that the child will be retarded.

Thrombi within vein of Galen malformations have been reported.7,13 In our case the entire malformation was thrombosed. This accounts for the fact that it is the only case in which angiography failed to reveal the presence of the malformation. Nonthrombosed vein of Galen malformations, however, may not be demonstrated when only unilateral carotid angiography is carried out.12

A fine shell of calcification in this location is virtually diagnostic of a vein of Galen malformation.10,14 The youngest age at which this calcification has previously been demonstrated was 15 years. None of the newborns or infants reviewed by Gold, et al., showed this radiologic picture.

This case provided direct proof that the mechanism of hydrocephalus is aqueductal occlusion because, following the removal of the aneurysm, the ethiodan which had been trapped in the ventricles was observed to pass down into the lumbar subarachnoid space. The head circumference has also remained normal after excision of the aneurysm. This confirms the early work of Bedford.2,3 He occluded the vein of Galen in nine Rhesus monkeys. Two died within 2 days and two others died suddenly during status epilepticus which came on after a normal postoperative period of 14 days. The
clinical course of these monkeys was reminiscent of our case in which an uncomplicated immediate postoperative period was interrupted by severe status epilepticus and respiratory distress.

**Summary**

The case of a vein of Galen malformation that we have presented is unique in that the aneurysm was completely occluded by thrombus, and there was no filling of the deep venous system during carotid angiography. The lesion was excised when the infant was 8 months old. The postoperative course was complicated by status epilepticus and respiratory distress, but the child has now improved to his preoperative condition.

**References**

12. RASMUSSEN, T. Personal communication, 1967.