Arteriovenous Malformation Involving the Inferior Sagittal Sinus in an Infant*

Case Report

Edward L. Seljeskog, M.D., Harry M. Rogers, M.D., and Lyle A. French, M.D., Ph.D.
Division of Neurosurgery, University of Minnesota Medical School, Minneapolis, Minnesota

Although there has been an increasing interest in the problem of intracranial venous aneurysms, case reports dealing exclusively with them have been relatively infrequent. In 1964, Gold, et al.,3 were able to collect only 34 cases from the literature and in addition reviewed eight of their own. For the most part these congenital intracranial venous aneurysms have demonstrated a predilection for the region of the vein of Galen and invariably they have been associated with an arteriovenous malformation that has involved branches of several of the major intracranial vessels. It is believed that the pathogenesis of these venous aneurysms is related to the shunting of arterial blood under high pressure through a maldeveloped capillary system. This basic developmental defect permits blood under arterial pressure to enter thin-walled veins which, under these circumstances, dilate and become aneurysmal in appearance. It is then not unreasonable to expect that any of the intracranial veins or venous sinuses could be involved.

In general, the surgical approach to aneurysms of the vein of Galen has been to attempt to occlude the arterial supply1,5,7,8,10 and thus "shrink" the aneurysm and at times relieve the patient's symptoms. Obviously, because of the location of the lesion, as well as the general nature of the problem, a direct surgical attack is fraught with a high degree of morbidity and mortality. Nevertheless, several case reports6,7,10 have indicated that these lesions should be excised surgically.

This case report is concerned with the successful surgical excision of a venous aneurysm of the inferior sagittal sinus, which in all probability developed secondary to a congenital midline arteriovenous malformation in the frontal region.

Case Report

This 5½-month-old male infant came to the University of Minnesota Hospitals with a 3-week history of progressive enlargement of the head, complicated for the last few days by a number of left-sided focal seizures. Prenatal and neonatal histories were not contributory except that the parents had noted that his development had seemed somewhat "slow" in comparison to that of his siblings.

Examination. The patient was a well-developed, somewhat lethargic child in no obvious distress. The head circumference was 46 cm. There was a prominent venous pattern in the scalp of the frontal region and the fontanel was firm. Auscultation revealed a moderately loud bruit, which was best heard in the left frontal area. There was no specific clinical evidence of focal deficit, but there was a moderate degree of developmental retardation.

From a clinical standpoint, it was believed that the most likely diagnosis was that of an arteriovenous malformation, possibly in association with an aneurysm of the great vein of Galen, although the distinct possibility of a cerebral neoplasm or other intracranial space-occupying lesion was also considered.

Skull roentgenograms revealed an early diastasis of the sutures, and a technetium-99 brain scan was consistent with an area of increased uptake in the midline region anteriorly (Fig. 1). A routine chest roentgenogram demonstrated cardiomegaly. With the suspicion of an arteriovenous anomaly more firmly established, a right retrograde brachial angiogram was performed under local anesthesia (Fig. 2). This showed the presence of a large frontal midline arteriovenous malformation, draining directly into a venous aneurysmal dilatation of the inferior sagittal sinus.

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there was also a moderate degree of dilatation and tortuosity of the transverse and straight sinuses, the vein of Galen and the posterior aspect of the inferior sagittal sinus. The superior sagittal sinus appeared normal. The malformation was fed principally from a dilated right anterior cerebral artery; the right middle and posterior cerebral arteries appeared to be uninvolved. A left percutaneous carotid angiogram demonstrated a significant contribution from several large branches of the left middle cerebral artery which coursed over the cerebral convexity (Fig. 3). The left anterior cerebral artery was poorly visualized partly because of technical problems and partly because of its configuration.

Considering the progressive symptomatology and the known propensity for these lesions in this age group to enlarge gradually and progressively, particularly after they have once become symptomatic, we concluded that surgical excision of the lesion was the only advisable form of therapy.

**Operation.** A standard coronal skin incision was made and a bifrontal craniotomy
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performed. The scalp and dura were extremely vascular. There were numerous anastomoses between the dural and scalp vessels. Upon opening the dura many large tortuous branches of the middle cerebral artery were seen coursing over the cerebral convexity toward the interhemispheric region. These abnormal vessels were doubly clipped and divided, eventually allowing exposure of the region of the malformation and aneurysm. Attention was then directed to the right frontal area. Again the dural vascular pattern was greatly enlarged. The right anterior cerebral artery was exposed and as expected this proved to be dilated and tortuous, entering directly into a mass of vessels adjacent to the aneurysm. This large feeder and several comparatively smaller arteries were clipped and divided, resulting in a prompt, "softening" of the aneurysm. Additional smaller feeding vessels from the left anterior cerebral artery were occluded and divided, sparing the main trunk of this vessel which, interestingly, was hypoplastic distal to the origin of the feeding vessel; this was probably related to a robbing of the normal circulation by the shunting branches. At this juncture we decided to excise the aneurysm in toto since we could not be entirely sure that there were no other small arterial feeders entering the aneurysm in areas concealed by the mass of aneurysmal dilatation, which at a later date, might enlarge and re-establish a significant arteriovenous fistula. Equally significant was the fact that the venous drainage from areas of the brain uninvolved by the malformation entered an entirely normal superior sagittal sinus or the inferior sagittal sinus posterior to the aneurysmal dilatation. Consequently the falx was divided anteriorly and the aneurysm excised, along with the adjacent mass of vessels forming the malformation. Hemos- tasis was obtained and the incision closed in a routine fashion.

Figure 4 shows the aneurysm, about the size of a chicken egg. It was located within the interhemispheric region superior to the corpus callosum, displacing both frontal lobes laterally. For the most part the associated arteriovenous malformation lay on the cortical surface, consequently it was not necessary to excise cortical tissue to obliterate the arterial supply. When the procedure was completed, the cortical surfaces appeared normal, with apparently normal blood flow.

Postoperative Course. The child did very well. He awakened immediately and was eating well within 24 hours. There was no demonstrable neurologic deficit. During the postoperative period, pronounced edema developed around the frontal part of the scalp and the eyes, and there was an appearance of a generalized ischemia of the frontal scalp flap. Undoubtedly this was related to altera-

Fig. 3. Left: Left percutaneous carotid angiogram showing several branches of the left middle cerebral artery entering directly into the malformation. The left anterior cerebral artery is hypoplastic. Right: Left percutaneous carotid angiogram demonstrating the vessels, arising from the middle cerebral artery complex, entering the aneurysmal inferior sagittal sinus.
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The excised malformation and venous aneurysm of the inferior sagittal sinus.

Fig. 4. The excised malformation and venous aneurysm of the inferior sagittal sinus.

tion of its abnormal vascularization by the operative procedure. This ischemic appearance gradually cleared as collateral channels were established. A second problem was that of recurrent subgaleal extradural effusion, necessitating a number of aspirations, again possibly related to alteration of vascular drainage of the area.

Postoperative angiography just before the patient was discharged showed complete removal of the malformation and aneurysm (Fig. 5). As anticipated there was no visualization of the right anterior cerebral artery, but collateral flow into the area, as judged from the serial films, appeared to be abundant. Similarly, flow into the left frontal region appeared to be quite adequate. There was an extracortical avascular area in the left frontal region suggestive of a subdural hematoma. Reopening of a small area of the incision under local anesthesia revealed this to be a collection of pale xanthrochromic fluid, partly in the extradural space and partly subdural in the interhemispheric area formerly occupied by the malformation and aneurysm.

The child has continued to do remarkably well. He has had no further seizures, although he has been maintained on anticonvulsant medication. His head circumference is now well within normal range for his age, and his neurological examination is entirely normal. He has made considerable gains in psychomotor function and presently appears to be developing normally.

Discussion

Although there have been a number of case reports\(^1\)\(^,\)\(^3\)\(^,\)\(^7\)\(^,\)\(^8\)\(^,\)\(^10\) of apparently successful obliteration or excision of arteriovenous

Fig. 5. Left: Postoperative right retrograde brachial angiogram. The malformation has been excised, the carotid artery is about normal in size and the middle cerebral artery circulation seems normal. An avascular area is seen bifrontally in the subdural spaces which proved to be the former site of the aneurysm and malformation. Right: Postoperative right retrograde brachial angiogram which demonstrates normal middle cerebral vasculature and absence of midline malformation.
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Malformations, associated with aneurysms of the vein of Galen, we believe this is the first description of a congenital venous aneurysm involving the inferior sagittal sinus, as well as the first report of its successful surgical excision. Reports of venous aneurysms involving various intracranial dural sinuses have been recorded; however, these usually have been related to post traumatic arteriovenous fistulas. There should be no reason to believe that such a congenital venous aneurysm could not readily develop in any intracranial dural sinus, providing that a significant arteriovenous shunt occurs in an appropriate stage of fetal development.

Despite the fact that this particular aneurysm did not involve the vein of Galen, the child's clinical presentation, for patients of this age group, was quite typical of such a lesion. Psychomotor retardation, seizures, increasing head circumference, prominent distention of scalp veins, cranial bruit and cardiomegaly, certainly are typical signs and symptoms of an intracranial arteriovenous fistula with resultant intracranial venous hypertension. In prior reports, describing aneurysms of the vein of Galen, hydrocephalus and intracranial hypertension have been mainly attributed to occlusion of the aqueduct of Sylvius by the aneurysm. It would seem that an equally plausible explanation concerning the etiology of hydrocephalus and increased intracranial pressure may be related to venous hypertension and impairment of cerebrospinal fluid reabsorption. This is especially apparent in this patient in whom there was only moderate dilatation of the vein of Galen. It is possible, however, that there was aqueductal occlusion and that both factors were important.

An air study most surely would have been of interest. Earlier reports have shown that small infants can tolerate a surgical procedure of this magnitude and in this regard this case was quite typical. We were particularly interested in the gains in psychomotor development, after removal of the malformation. This certainly supports the concept that these lesions rob normal brain tissue of vital blood flow. Although it was necessary to ligate a major intracranial vessel to deal effectively with this lesion, and there was also a hypoplastic counterpart contralaterally, collateral vascular flow at the time of surgery seemed quite adequate. This correlates well with the patient's clinical course and postoperative angiographic studies. Probably these vessels contributed little to the vascular supply of normal areas and collateralization probably had built up earlier, secondary to the siphoning effect of the shunt.

One practical point concerns associated cardiac problems in this condition. A number of earlier reports have dealt with the propensity for a preoperative high-output cardiac failure and as the fistula is occluded during surgery, congestive failure develops due to circulatory overloading. In this particular case cardiomegaly was well demonstrated, although frank cardiac failure did not occur. This may have been related to the gradual obliteration of the shunt during the course of the operative procedure, as individual feeding vessels were occluded. Furthermore, no blood transfusions were given during the operative procedure, in the hope that the gradual blood loss inherent in the procedure would help compensate for any possible cardiac overloading. Nevertheless careful and continuous oscillographic monitoring of the electrocardiogram, pulse rate and arterial blood pressure with minimal fluid replacement during surgery and during the postoperative period is mandatory. The monitoring of the central venous pressure is valuable as this is a most reliable method of determining the need for fluid replacement. This is particularly true of the pediatric patient when even a minimum overtransfusion can be hemodynamically catastrophic. In this clinic a small central venous polyethylene cannula is placed into the superior vena cava via a cephalic vein cut down, or into the inferior vena cava via a femoral approach. In adult patients the catheter can usually be inserted percutaneously through the subclavian vein. The catheter is left in place throughout the operative procedure and during the immediate postoperative period when careful monitoring of fluid and blood replacement is paramount. In infants the venous pressure should be maintained in the range of 0-4 cm of saline.

From the diagnostic standpoint it is interesting to note the patient's "positive" preoperative brain scan and its return to normal after surgery. This finding has been previously reported by Pool and may prove to
be a valuable diagnostic adjunct in problems of this sort. However, there is still need for complete, and at times selective, angiography when planning a surgical attack on these lesions.

Summary

We have described the successful diagnosis and excision of a congenital venous aneurysm involving the inferior sagittal sinus and its associated arteriovenous malformation. We have stressed the invariable relationship of these lesions to arteriovenous malformations, and have demonstrated the imperative need for adequate preoperative angiography to define the feeding arterial supply. The use of radioactive brain scanning as a diagnostic adjunct and the necessity of adequate preoperative cardiac evaluation and monitoring during surgery have been emphasized.

References