Thrombosed Aneurysm of the Vein of Galen

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Computed tomography in a 3½-year-old boy with ataxia, lethargy, fever, vomiting, and increasing irritability revealed moderate hydrocephalus and a blood density mass lesion lying superior and posterior to the 3rd ventricle. On several of the sections, the blood density was greater in the dependent than in the superior portion of the mass. The average attenuation number within the mass was 40 EMI units. The infusion of contrast agent demonstrated capsular enhancement (a target sign), which was also demonstrated by angiography. The diagnosis, which was confirmed at operation, was thrombosis of an aneurysm of the vein of Galen. The massive thrombus was resected, but the aneurysm was not excised. The patient's recovery was uneventful. (Neurosurgery, 7: 274–278, 1980)

Key words: Aneurysm, Angiography, Capsular enhancement, Computed tomography, Thrombosis, Vein of Galen, Venous malformation

INTRODUCTION

Thrombosis of an aneurysm (or varix) of the vein of Galen is most unusual; only five cases appear in the literature (7, 9, 11, 14, 17). Those five cases are summarized in this report and a sixth case is presented, including the computed tomographic, cerebral angiographic, and surgical findings.

CASE REPORT

This 3½-year-old boy was admitted to North Carolina Baptist Hospital with a 2-week history of lethargy, intermittent fever, vomiting, and progressive irritability. His gait had been ataxic for almost a year. He had had no cardiac or respiratory symptoms and no seizures. His mental status had been normal, and he had reached all of his developmental milestones normally.

Physical examination revealed an irritable child with normal strength and normal reflexes. His extraocular movements were intact and he was able to look up. His fundi were normal. His head circumference was normal at 50 cm; he had no cranial bruits and no nuchal rigidity. Spinal fluid examination revealed clear fluid with 189 white blood cells (of which 82% were polymorphonuclear leukocytes), a glucose content of 52 mg/100 ml, and a protein content of 107 mg/100 ml. Plain roent-genograms of the skull were normal with no evidence of intracranial calcification.

Computed tomography (CT) (Figs. 1 and 2) demonstrated hydrocephalus and a normal posterior fossa. At mid- and high ventricular levels, an oval mass of blood density (40 EMI units) was seen superior to the 3rd ventricle and extending posteriorly between the atria. The suggestion of a fluid level with higher absorption values in the dependent portion of the mass was present on two horizontal sections.

Bilateral internal carotid and left vertebral angiography was performed. The left vertebral angiogram (Figs. 3 and 4) demonstrated posterior displacement of the splenial vessels and anterior displacement of the medial and lateral posterior choroidal arteries, compatible with a mass in the splenium of the corpus callosum. The choroidal vessels were normal in size, with a beaded appearance. Carotid angiography (Fig. 5) failed to show opacification of the internal cerebral vein or the vein of Galen. No abnormal vessels were present, and a few opacified subependymal veins demonstrated hydrocephalus. The superficial venous system was patent and prominent. A striking

finding during the postvenous phase was contrast enhancement of an oval structure compatible with the ring seen on the CT scan (Figs. 4 and 6).

At operation, via an interhemispheric approach, the posterior corpus callosum was split. Within 3 mm a smooth, firm, greenish yellow capsule was encountered, which, when opened, proved to contain a lamellated hematoma of varying ages and some liquid hematoma. It was evident that this capsule was a thrombosed aneurysm of the vein of Galen. The mass effect of the aneurysm was reduced by resection of the thrombus, but the aneurysm itself was not excised.

The patient tolerated the operation well and his lethargy and irritability improved. One month after operation, his physical examination was normal and his gait was remarkably improved. Three months after operation, a CT scan revealed normal ventricular size and overall reduction in the size of the aneurysm.

DISCUSSION

Clinical presentation

Amacher and Shillito recognized four clinical syndromes caused by an aneurysm of the vein of Galen, the four depending on the patient's age (1). Neonates present with cardiac failure and a cranial bruit; their prognosis is poor. Infants present with mild cardiac failure early and craniomegaly later. In the 1st year of life, children present with hydrocephalus, cardiomegaly, and occasionally seizures. Older children and adults present with headaches, syncope, and occasionally subarachnoid hemorrhage. In nearly all of the last group of patients, routine skull roentgenograms demonstrate calcification around the aneurysm.

There is no typical syndrome for a thrombosed aneurysm of the vein of Galen, but all of the young children presented with hydrocephalus and progressive irritability and lethargy (Table 1).

Computed tomography

CT does not always allow one to make a histological diagnosis of a mass in the posterior portion of the 3rd ventricle, but it will clearly demonstrate the mass and its effects on the ventricular system. With aneurysms of the vein of Galen, the 3rd ventricle is pushed forward and down by a large, rounded, enhancing mass and the lateral ventricles are dilated (2, 6, 8,

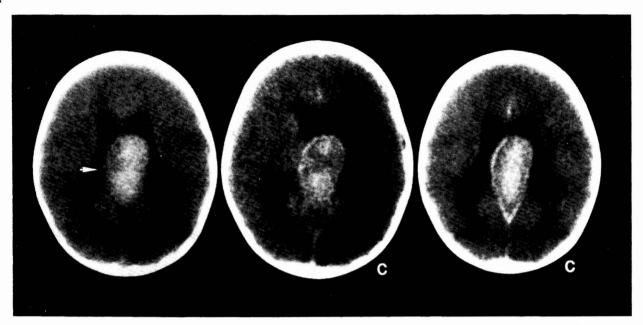


Fig. 1. Computed tomogram, axial projection, shows an interventricular blood density mass, with rim enhancement on the infused scan (C). Arrow indicates fluid level within the mass.

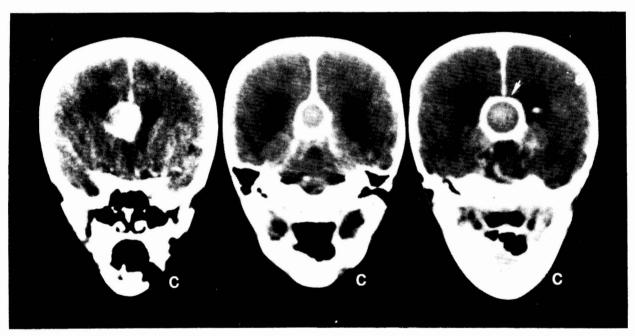


Fig. 2. Coronal computed tomogram after infusion shows flattening of the enhancing rim by the corpus callosum (*arrow*) and the interventricular extent of the mass.

13). No edema surrounds the mass. The circulating blood pool within the mass can be measured directly before contrast infusion; Berger et al. (2) and Macpherson et al. (12) both reported measurements of 18 to 20 EMI units. Spallone described a "triangular shaped aspect of the complex sinus rectus and the torcula Herophili" (16), which is characteristic of a malformation of the vein of Galen.

In our patient, the posterior 3rd ventricular mass and associated hydrocephalus were seen on CT scans, and several other features were noted as well:

1. Blood density was not uniform within the mass, although there was a dependent blood-fluid level. This stagnant blood pool within the aneurysm did not show contrast enhancement, as it would have had it been free circulating blood. In addition, the CT numbers averaged 40 EMI units—compatible with organizing thrombus—not the 18 to 20 EMI units normal for free-flowing blood.

- 2. On the coronal images, the upper portion of the aneurysm was flattened against the corpus callosum, which in turn was limited from above by the falx cerebri.
- 3. The periphery of the mass enhanced dramatically with contrast infusion. This effect was due to enhancement of vascular granulation tissue and was also seen clearly on cerebral angiograms, especially the anteroposterior projection.



FIG. 3. Left vertebral angiogram shows beading of the posterior choroidal arteries (*arrow*) and a mass effect on the posterior splenial artery.

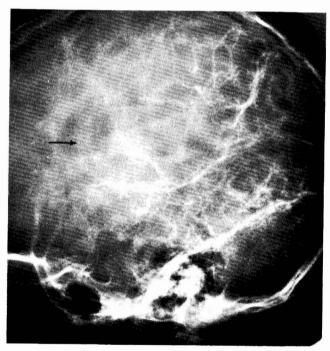


FIG. 4. Left vertebral angiogram, postvenous phase, shows an enhancing ring of granulation tissue around the mass (arrow).

The enhancing ring (or target sign), due to granulation tissue, that is seen on CT scans has been the subject of several reports from this institution. We have reported seeing a ring in scans of cerebral arterial aneurysms, in which the outer part of the circle was due to enhancement of granulation tissue in the wall of the aneurysm (15). Buonanno et al. have demonstrated abundant granulation tissue in the thrombus and inflammatory

changes in the walls of the thrombosed lateral sinus (3, 4). Laster et al. have demonstrated conclusively the enhancing effects of vascular granulation tissue in resolving intracerebral hematomas (10).

Macpherson et al. showed a target sign by CT scan 2 months after clipping the feeding vessels to an aneurysm of the vein of Galen and interpreted the finding as being due to blood circulating peripherally within the aneurysm around a central thrombus (12). We suggest that in their case the peripheral enhancement was due to vascular granulation tissue in the wall of the aneurysm rather than to free blood.

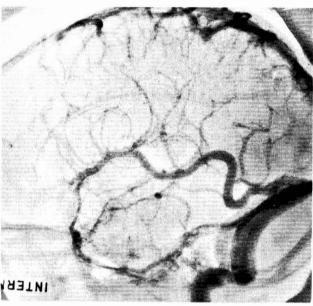


FIG. 5. Carotid arteriogram, venous phase, left internal carotid injection, shows lack of filling of the deep venous system and prominence of the superficial draining veins.

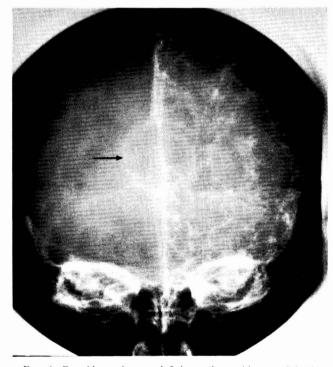


FIG. 6. Carotid arteriogram, left internal carotid artery injection, shows ring enhancement of the periphery of the mass (*arrow*).

TABLE 1
Summary of Cases of Thrombosed Aneurysm of the Vein of Galen

Author	Age	Sex	Symptoms and Signs*	Calcification on X-ray Film	Treatment*	Postoperative Course
Lazar (11)	11 mo	F	Drowsy, irritable; hydro- cephalus	+	VP shunt, aneurysm excised	Stormy
Heinz et al. (7)	25 wk	M	Irritable, lethargic, hemi- paresis; increased ICP, hydrocephalus	-	Aneurysm excised	Stormy
Jamieson (9)	15 yr	F	_	+	Aneurysm excised	_
Siqueira and Mur- ray (14)	30 yr	F	Progressive spasticity; hy- drocephalus	+†	Aneurysm excised	Stormy
Weir et al. (17)	8 yr	M	Drowsy, irritable, seizure; hydrocephalus	+	VA shunt, aneurysm ex- cised	Stormy
Six et al.	$3\frac{1}{2}$ yr	M	Drowsy, irritable; hydro- cephalus	-	Resection of thrombus	Uneventful

^{*} Abbreviations: ICP, intracranial pressure; VP, ventriculoperitoneal; VA, ventriculoatrial.

Cerebral angiography

Both carotid vessels must be studied to ensure filling of the deep venous system. The venous phase angiogram will show filling of the cortical veins, inferior anastomotic vein, superior sagittal sinus, and sigmoid sinus, but failure of the internal cerebral veins, vein of Galen, and straight sinus to fill (5, 17). On occasion, the inferior sagittal sinus and basilar vein may be carrying an increased venous load (7). The unusual appearance of the ring shadow of the capsule on angiograms in our patient has not been described previously.

Thrombosis

In four of the five previously reported cases of thrombosis of an aneurysm of the vein of Galen, rim calcification could be seen by plain roentgenography; three of these five patients were younger than 15 years. Siqueira and Murray reported a 14% incidence of rim calcification in 75 patients with aneurysms of the vein of Galen, and the majority of the patients with rim enhancement were older than 15 years (14). The higher incidence of rim calcification (3 of 5) in those reported patients with thrombosed aneurysms and their younger age are interesting, particularly because our 3½-year-old patient did not show rim calcification on plain roentgenograms or by CT.

Heinz et al. suggested that the aneurysm in their patient had thrombosed prenatally or during the birth process because the patient had no evidence of cardiac failure or respiratory difficulties (7). Lazar refuted that idea because of the presence of clot material of varying ages within the aneurysm in his patient (11). In our patient, there was no doubt that some of the clot was recent; in the noninfused CT scan there was blood density within the aneurysm with a distinct layering effect. At operation, we found an old, laminated, organized thrombus and a fresher liquid clot. Our patient also had no history of cardiac problems, as well as no cardiomegaly and no craniomegaly.

Management

We recommend that a thrombosed aneurysm of the vein of Galen should be treated by resection of the thrombus but not be removed. Simple reduction of the mass will decompress the aqueduct and relieve the hydrocephalus. Total removal of the malformation will cause unnecessary morbidity and mortality.

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REFERENCES

- Amacher, A. L. and Shillito, J., Jr. The syndromes and surgical treatment of aneurysms of the great vein of Galen. J. Neurosurg., 39: 89-98, 1973.
- Berger, P. E., Harwood-Nash, D. C., and Fitz, C. R. Computerized tomography: Abnormal intracerebral collections of blood in children. Neuroradiology, 11: 29–33, 1976.
- Buonanno, F. S., Moody, D. M., Ball, M. R., Cowan, R. J., Laster, D. W., and Ball, J. D. Radionuclide sinography: Diagnosis of lateral sinus thrombosis by dynamic and static brain imaging. Radiology, 130: 207-213, 1979.
- Buonanno, F. S., Moody, D. M., Ball, M. R., and Laster, D. W. Computed cranial tomographic findings in cerebral sinovenous occlusion. J. Comput. Assist. Tomogr., 2: 281–290, 1978.
- Gold, A. P., Ransohoff, J., and Carter, S. Vein of Galen malformation. Acta Neurol. Scand. [Suppl. 11], 40: 1–31, 1964.
- Harwood-Nash, D. C. and Breckbill, D. L. Computed tomography in children: A new diagnostic technique. J. Pediatr., 89: 343–357, 1976
- 7. Heinz, E. R., Schwartz, J. F., and Sears, R. A. Thrombosis in the vein of Galen malformation. Br. J. Radiol., 41: 424-428, 1968.
- 8. Iannucci, A. M., Buonanno, F., Rizzuto, N., Mazza, C., Vivenza, C., and Maschio, A. Arteriovenous aneurysm of the vein of Galen: A clinical, angiographic, CT scan and neuropathological study. J. Neurol. Sci., 40: 29–37, 1979.
- Jamieson, K. G. Excision of pineal tumors. J. Neurosurg., 35: 550– 553, 1971.
- Laster, D. W., Moody, D. M., and Ball, M. R. Resolving intracerebral hematoma: Alteration of the "ring sign" with steroids. A. J. R., 130: 935-939, 1978.
- Lazar, M. L. Vein of Galen aneurysm: Successful excision of a completely thrombosed aneurysm in an infant. Surg. Neurol., 2: 22-24, 1974.
- Macpherson, P., Teasdale, G. M., and Lindsay, K. W. Computed tomography in diagnosis and management of aneurysm of the vein of Galen. J. Neurol. Neurosurg. Psychiatry, 42: 786-789, 1979.
- Milhorat, T. H. Pediatric Neurosurgery. Philadelphia, F. A. Davis, 1978, pp. 318–322.
- Siqueira, E. B. and Murray, K. J. Calcified aneurysms of the vein of Galen: Report of a presumed case and review of the literature. Neurochirurgia (Stuttg.), 15: 106-112, 1972.
- Six. E. Use of the CT scan in the management of intracranial aneurysms. Presented at the 32nd Annual Meeting of the Neurosurgical Society of America, White Sulphur Springs, West Virginia, May 1979.
- Spállone, A. Computed tomography in aneurysms of the vein of Galen. J. Comput. Assist. Tomogr., 3: 779–782, 1979.
- Weir, B. K. A., Allen, P. B. R., and Miller, J. D. R. Excision of thrombosed vein of Galen aneurysm in an infant: Case report. J. Neurosurg., 29: 619–622, 1968.

[†] Calcification was first noted at age 5 years.

COMMENTS

This paper is of value because it calls attention to the unusual but very important entity of aneurysm of the vein of Galen and because it identifies a subset within the symptom spectrum that occurs with this lesion. The identification of the "target sign" on both CT and venous angiogram as a feature of thrombosed aneurysms of the vein of Galen is useful. I agree with the operative approach. No detectable deficit is produced in young children by division of the posterior part of the splenium, particularly when it has been splayed out by an underlying mass. Simple reduction in the size of the aneurysm in the circumstance presented here is the procedure of choice if the angiogram has indicated no active flow into the lesion. When an aneurysm of the vein of Galen is identified, full angiography must be carried out because the sac may fill from many sources, and a nearby angioma draining into the deep galenic system must be excluded before the operative approach

Aneurysms of the great vein of Galen are unusual, but very important. Except in the neonatal age group, their treatment may be accomplished with safety to eliminate mild, high output heart failure, hydrocephalus, and/or a posterior 3rd ventricular mass. When the aneurysm is thrombosed, resection will suffice. Aneurysm wall that can be removed easily should be, but where the aneurysm wall is adherent to the hypothalamus it should be left in place.

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Dr. Six and his colleagues are to be congratulated on their management of this difficult case and their excellent illustration of the neuroradiological findings.

I am not sure that one can categorically draw the conclusions that are referred to in their recommended form of management. There are several considerations that I would recommend to those who are concerned about managing a similar case. Many authors, including the present ones, have recorded the concomitance of obstructive hydrocephalus, which is usually related to compression of the aqueduct of Sylvius (3). I believe that, in addition to surgical management of the vein of Galen aneurysm, a preliminary surgical method should be utilized to treat the hydrocephalus. The form of therapy—ventriculostomy drainage for several of the perioperative days or a ventricular shunt-will depend on the circumstances. I agree with the authors that one may be able to afford an adequate permanent aqueduct decompression; however, one cannot be certain of this preoperatively nor assure it during the first few postoperative days. More importantly, there are significant technical considerations involved, mainly the preoperative stabilization of the increased intracranial pressure at more satisfactory lower levels and the intraoperative diminished requirement for firm retraction of the cerebral hemisphere. The latter offers a decreased risk of injury to the patient no matter which surgical approach is used.

The avenue of approach to the vein of Galen region has been the subject of intense neurosurgical interest for years. The present authors have utilized a parietal interhemispheric approach and correctly identified the need for sectioning a portion of the splenium. One must, of course, consider the consequences of any surgical corridor and weigh the risks and

advantages. There are, however, two other avenues to this region that could achieve the same result without risking injury to the splenium. The infratentorial supracerebellar and the supra- or transtentorial occipital approaches are currently advocated by a number of authors. Each has its own risks and advantages. One must also consider the advice about subtotal excision of the intra-aneurysmal clot. It would be difficult to judge not only how much clot remained but, more importantly, how much was critical to the successful relief of the aqueduct compression. I suggest that one consider the very careful excision of all of the intra-aneurysmal clot in these situations and then, for the sake of safety, also consider an aneurysmorrhaphy to try to ensure that the aneurysm will not recur or bleed. A simple technique of oversewing the opposing walls of the aneurysm may be used. Whether the walls of the aneurysm are amputated is an individual preference. Nevertheless, I do agree that an attempted total excision of the remaining aneurysm remnant directly adjacent to the tectal region is unwar-

A final point should be made concerning the occurrence of thrombus in these aneurysms. Although most of these aneurysms will not have any thrombus, the presence of clot of varying ages and amount is possible and has been reported (2, 5). Our patient, referred to in this article, is now 7½ years postoperative and continues to manifest mental retardation (as have a large portion of these patients preoperatively and postoperatively). In our patient there was clearly a multilayered thrombosis of varying ages; however, near the point of "origin" of the aneurysm the clot was very thin and seemed to be quite recent (4). This tends to reinforce my concerns about subtotal clot excision, recurrence, and postoperative bleeding-hence, the suggestion for oversewing the aneurysm. It is also worth restating that nonthrombosed aneurysms of the vein of Galen may not be demonstrated when only a unilateral carotid angiogram is done; at least a three-vessel (bilateral carotid and vertebral) angiogram should be considered because the primary or secondary feeding vessels can derive from a large number of immature, poorly differentiated vessels (1) or from the posterior cerebral, anterior and middle cerebral, or superior cerebellar arteries, among others (6).

Martin L. Lazar, M.D. Dallas, Texas

Cohen, M. M., Kristiansen, K., and Hval, E. Arteriovenous malformations of the great vein of Galen. Neurology (N. Y.), 4: 124–127, 1954.

Gibson, J. B., Taylor, A. R., and Richardson, A. E. Congenital arteriovenous fistula with an aneurysm of the great cerebral vein and hydrocephalus treated surgically. J. Neurol. Neurosurg. Psychiatry, 22: 224-228, 1959.

Lagos, J. C. Congenital aneurysms and arteriovenous malformations. In Congenital Malformations of the Brain and Skull, Part II, Handbook of Clinical Neurology, Vol. 31, edited by Vinken, P. J. and Bruyn, G. W., pp. 137-209, Amsterdam, Elsevier/North Holland Biomedical Press, 1977

^{4.} Lazar, M. L. See Lagos, J. C. (3), p. 197.

Russell, D. S. and Nevin, S. Aneurysm of the great vein of Galen causing internal hydrocephalus: Report of two cases. J. Pathol., 51: 375–383, 1940.

Yasargil, M. G., Antic, J., Laciga, M. D., Jain, K. K., and Boone, S. C. Arteriovenous malformations of vein of Galen: Microsurgical treatment. Surg. Neurol., 6: 195–200, 1976.