Spontaneous Thrombosis of Deep Cerebral Veins: A Complication of Arteriovenous Malformation

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Abstract:
An uncommon type of stroke in children is presented. An intracranial arteriovenous malformation in a 13-year-old boy spontaneously occluded about 22 months after surgical intervention. Precipitating factors, such as bacterial infections, could not be demonstrated in this patient, who had been attending school since the time of the craniotomy. The histological features of venous encephalomalacia in the galenic territory are contrasted with hemorrhagic encephalomalacia as seen after arterial occlusions: in the former, hemorrhages are more widespread and edema is more pronounced.

Additional Key Words
venous encephalomalacia
childhood stroke
hemorrhagic encephalomalacia
infarction

Introduction

Occlusion of intracranial vessels in children is rare, but when it occurs spontaneously, veins and sinuses rather than arteries are more apt to be involved. The vast majority of intracranial venous occlusions in children and adults have been demonstrated in association with bacterial infections, either systemic or intracranial.

We report an instance of spontaneous intracranial venous occlusion in a 13-year-old boy. Several vessels feeding into a large arteriovenous malformation, located in the anterior hypothalamus, had been surgically clipped approximately 22 months prior to death. Shortly before his death, the patient complained of severe headache and became unconscious while sitting in the classroom. Nonfilling of the venous component of the arteriovenous malformation and of the deep cerebral veins was demonstrated by angiography a few hours later. Phlebothrombosis of the same vessels, in the absence of an apparent precipitating factor, was demonstrated at autopsy.

The histological features of venous encephalomalacia in this case are contrasted with those occurring secondarily to arterial occlusion. The causes of nontraumatic intracranial venous occlusion are reviewed.

Case Report

An 11-year-old boy was first admitted to the University of Maryland Hospital on April 11, 1972, because of severe headaches and seizures. On admission, the blood pressure was 110/30, the pulse 78 per minute and regular, and respirations 16 per minute. There was marked nuchal rigidity. The patient was awake, alert and very irritable. Numerous, bilateral, subhyaloid hemorrhages and optic nerve papilledema were noted; retinal veins were engorged and pulseless. No motor deficit existed and the deep reflexes were hypoactive throughout. Pathological reflexes were not elicitable. Lumbar puncture yielded bloody spinal fluid with an opening pressure of 40 mm H2O. A curvilinear calcification in the left frontal area was seen on x-ray examination of the skull. Increased radionuclide uptake was noted in the left temporal area and an epileptogenic focus in the left temporal region was demonstrated by electroencephalography. Left carotid arteriography disclosed a vascular malformation (AVM) extending from the left anterior cerebral artery to the front of the left carotid siphon; the contrast medium drained primarily through enlarged basal veins of Rosenthal (figs. 1 and 2).

On April 28, 1972, the anterior communicating artery and several vessels leading into or draining the arteriovenous malformation were surgically clipped. Postoperatively, there was mild left hemiparesis, and bilateral angiography showed that the arteriovenous malformation filled from the left carotid artery only. There was decreased shunting into the deep cerebral veins (fig. 2).

Ten weeks later he was readmitted because of persistent high temperature (101°F), anorexia and drowsiness. After extensive work-up the conclusion was that these symptoms were secondary to anterior hypothalamic dysfunction.

At the end of 1973 he received several whole-blood transfusions and other medications because of pancytopenia of undetermined etiology. The patient improved sufficiently to return to school.
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FIGURE 1
Lateral arteriogram (April, 1972). The vascular malformation and its large draining veins are seen just above the planum sphenoidal with drainage flowing into the basal vein of Rosenthal, the vein of Galen and the straight sinus. Note that there is little or no filling of the cerebral hemispheric arteries on this injection; nonfilling is attributable to the "steal" by the vascular malformation. A calcified varix can be seen just above the contrast-filled veins.

FIGURE 2
Arteriogram (May 12, 1972): Lateral view showing multiple surgical clips and a decrease in the bulk of the vascular malformation. There is improved filling of the cerebral hemispheric arteries with decreased flow through the malformation.
On February 26, 1974, while sitting in the classroom he had severe headache, vomiting and grand-mal seizures. On admission to the hospital the blood pressure was 110/0, pulse 180 per minute and respirations 30 per minute. His temperature was 108°F. The patient was unconscious, while the pupils were equal and reacted weakly to light. Corneal reflexes and passive extraocular movements were normal. Sensory stimulation resulted in decorticate posturing. There was generalized hypeflexia. Laboratory examinations were noncontributory.

Twenty-one hours after admission the pupils were fixed and dilated. No corneal reflexes were elicited. Bilateral carotid angiography was performed. The right-sided study showed marked delay of the internal carotid artery circulation time, and the left-sided study disclosed slow filling of the carotid circulation. No contrast medium filled the arteriovenous malformation and the study was suggestive of dilated lateral ventricles (figs. 1 through 3). A ventricular needle was inserted into the right cerebral hemisphere and fresh blood was drained. The patient died approximately 36 hours after the last admission to the hospital.

**Autopsy Findings**

The heart weighed 260 gm (normal average weight for this age group: 140 gm) and showed biventricular hypertrophy. No significant abnormalities were demonstrated in the other thoracic or abdominal viscera. The brain weighed 1,550 gm, with the normal estimated weight for this age group being 1,360 gm. The cerebral hemispheres were asymmetrical, the left one being slightly more swollen. There was slight uncinate herniation bilaterally. The superior sagittal sinus was patent. There was mild, yellowish-orange pigmentation of the leptomeninges, which also adhered firmly to the dura at the base of both frontal lobes. Several surgical clips were found in place, occluding at least six branches attached to the main sac of arteriogram (February 27, 1974): The surgical clips are visible in place; there is no filling of the arteriovenous malformation. There is evidence of slow flow into the parenchymal vessels; elevation and straightening of the pericallosal artery is indicative of hydrocephalus. Downward displacement of the posterior communicating and the posterior cerebral arteries suggests uncal herniation.
the vascular malformation (one of the clips occluded the anterior communicating artery). Large collections of fresh blood were not present at the base of the brain. Several abnormal blood vessels coursed on the surface of the gyri rectus; some of these tortuous vascular channels were markedly dilated and partly calcified and some were occluded by fresh thrombi (fig. 4). Microscopically, these vascular channels showed irregular, thickened walls with frequent variations in structure of the elastic lamina and the smooth muscle layer (fig. 4b). The neighboring cerebral tissue displayed moderate gliosis and clusters of hemosiderin-laden macrophages. One of these abnormal vessels drained directly into the left basal vein of Rosenthal, which was markedly dilated (fig. 5). There were numerous recently formed thrombi in the basal veins, the internal cerebral veins, the great vein of Galen and the straight sinus (fig. 5b). These thrombi were composed of red blood cells and fibrin strands with occasional white blood cells and platelets. Extensive hemorrhagic encephalomalacia was seen in both thalamic nuclei, the hypothalamus, the ventral half of the corpus callosum, the head of both caudate nuclei, the left putamen and the paraventricular white matter of the left tempo-occipital region. Infratentorial tissues were not involved (figs. 5 and 6). A small amount, about 5 ml, of fresh blood was noted in the third and lateral cerebral ventricles. Microscopically, the lesion was characterized by the relative absence of blood in the arterial vessels and by the marked engorgement and dilatation of the veins and venules, which were surrounded by fresh hemorrhages and numerous, confluent areas of vacuolation (fig. 7).

Discussion
Nontraumatic cerebral vascular occlusions in children commonly affect veins and sinuses rather than arteries. Among the situations associated with intracranial venous occlusions in children, Banker found infectious processes (either localized or systemic), congenital cardiac anomalies and parturition. Cerebral venous thrombosis in children has been extensively studied and reported by Kalbag and Woolf, whose cases fell into the same categories described above. Neither of these reports mention instances of a spontaneous venous occlusion with an intracranial AVM. Perrett and Nishiooka reviewed the records of 543 patients with intracranial AVM, including five in whom the Galenic vein was involved. The instance reported herein is seemingly another uncommon cause of stroke in children, an entity that deserves the report of individual cases, as suggested recently by a national study group.

Encephalomalacia, or cerebral softening, is a descriptive term applied to lesions developing after circulatory disturbances. Those that result from occlusion of either arteries or veins are commonly designated infarction. Encephalomalacia with a large hemorrhagic component, usually in the form of gray matter petechiae, is attributed to arterial occlusions that are incomplete or temporary.

Hemorrhagic encephalomalacia secondary to venous occlusion (i.e., venous infarction) is a well-recognized but poorly illustrated lesion. The vast majority of instances previously reported occurred in association with leptomeningeal bacterial infections accompanied by phlebitis of the superficial cerebral veins or the dural sinuses. Aseptic occlusion in the deep cerebral venous territory is well illustrated by the present case. The distribution of the lesion corresponds to the draining territory of the internal cerebral veins, basal veins of Rosenthal and their main tributaries. The slight hemispheric asymmetry observed may be related to the presence of the arteriovenous malformation on the left side. Grossly, the lesion differs from hemorrhagic encephalomalacia of arterial origin in that the hemorrhages are confluent and profuse, and involve both gray and white matter structures. The histological features of encephalomalacia (infarction) of occlusive arterial origin have been contrasted with those features observed in the present case. The outstanding differences include a large number of perivenous hemorrhages and the presence of markedly dilated spaces in the neuropil. Both predominate in lesions of occlusive venous origin and are not demonstrable in arterial infarctions of a comparable age.

Although the mechanism for venous thrombosis in this case is unclear, we postulate that an initial episode of intraventricular hemorrhage was followed by markedly increased intraventricular pressure, slowing of the venous circulation, and finally by thrombosis.

The effects of isolated Galenic vein occlusion, ranging from four days to seven months, were studied in rhesus monkeys and dogs by Hammock et al., who reported minimal clinical or morphological consequences. These authors suggested that the effects of Galenic vein occlusion in humans might be difficult to assess, since the occlusion seldom occurs as an isolated event and its symptoms may be obscured by the underlying systemic or local diseases that affect the majority of these patients. The occlusion demonstrated in the present case included not only the vein of Galen but also the two veins of Rosenthal, the two internal cerebral veins, and their major tributaries. This probably accounts for the severity of the clinical deficit and the large size of the resulting hemorrhagic encephalomalacia.

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Coronal section of the cerebral hemispheres near the anterior communicating artery displaying a surgical clip in place. A large varix appears occluded near the midline. Histologically, the channels from the vascular malformation show arterial walls with irregular layers of smooth muscle and absence of elastic fibers; there is increased connective tissue in the adventitia. Note the absence of fresh subarachnoid hemorrhage (H & E, 6X).
Coronal section of the cerebral hemispheres through the splenium and the superior colliculi showing extensive hemorrhagic encephalomalacia partially involving the pulvinar and the adjacent structures on both sides. Basal veins of Rosenthal are engorged and occluded by fresh thrombi, the left one being particularly prominent.

Cross sections of the terminal portions of the veins of Rosenthal and the internal cerebral veins are seen shortly before their entry into the great vein of Galen, which was also completely thrombosed.

Coronal section of cerebral hemispheres through the posterior limb of the internal capsules demonstrates encephalomalacia in the territory normally drained by the deep venous cerebral system. A rubber catheter was placed within the left centrum semiovale. Note the diffuse, marked swelling of the cerebral hemispheres and the slight degree of bilateral uncal herniation.
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Histological appearance of the hemorrhagic encephalomalacia (previously illustrated in figure 6). Large distended veins surrounded by fresh hemorrhage and extensive vacuolation of the neuropil are seen in a sample from the right thalamic nuclei (H & E, 10X).

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References
4. Coppoletta JM, Wolbach SB: Body length and organ weights of infants and children. A study of the body length and normal weights of more important vital organs of the body between birth and twelve years of age. Amer J Path 9:55-70, 1933
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