Recurrent Cavernous Sinus Syndrome Complicating Supratentorial Arteriovenous Malformation: Report of a Case

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SUMMARY A 5-year-old girl with history of unexplained cerebral palsy and cranial asymmetry presented with an acute encephalopathy and a cavernous sinus syndrome. Radiologic studies disclosed an arteriovenous malformation draining into a dilated vein of Galen, arterialization of flow into the cavernous sinuses from runoff of the malformation, and non-filling of the cavernous sinuses on orbital venography. Initial remission of her symptoms was followed by recurrence three months later after an episode of mild head trauma. The relationship of AVM’s to cavernous sinus disease is discussed.

AMONG VASCULAR ANOMALIES involving the cavernous sinuses, spontaneous or traumatic carotid-cavernous fistulas are perhaps the best known. Less well-appreciated are arteriovenous malformations (AVM’s) which are either structurally contiguous with the sinuses, or communicate with them from distant locations. These lesions may be reflected clinically by cavernous sinus syndromes of varying extent, severity, and temporal evolution. However, these syndromes are distinctly uncommon with those AVM’s which occupy a supratentorial location. We report such a case which was further notable for (a) an unusual premorbid history and (b) recurrent clinical presentation.

Case Report

A 5-8/12-year old white girl was thought to be well until one year of age when she was noted to have internal turning of her left foot and poor weight bearing of her left leg. Exam revealed underdevelopment of the right side of the calvarium, diminished muscle bulk of the left leg, and left-sided hyperreflexia. The diagnosis was cerebral palsy, cause undetermined. She failed to keep appointments for prescribed laboratory studies, and was lost to neurological follow up. However her motor deficit did not progress, and she became independent by age 2½ years. Cognitive development proceeded normally.

She was in her usual state of health until one day prior to admission when she developed headache, anorexia and abdominal pain. The following day her symptoms were compounded by vomiting and irritability alternating with drowsiness. She was brought to Massachusetts General Hospital, where examination revealed an afebrile, normotensive girl who was lethargic but responsive to both voice and tactile stimuli. Skin and mucous membranes were dry; neck was supple; heart, lung and abdominal exams were normal. CBC and routine chemistries were unremarkable. Urinalysis showed a specific gravity of 1.027 and slight ketones. She was admitted with a tentative diagnosis of gastroenteritis and fluids were begun at twice maintenance.

On the following day she became difficult to arouse. Lumbar puncture yielded clear, colorless fluid, 2 lymphocytes/cmm, 0 rbc/cmm, protein = 19 mg/dl, glucose = 79 mg/dl. Opening pressure was not recorded. Neurologic consultation was then requested.

The child was found to be unresponsive to voice; tactile stimulation produced semipurposeful avoidance movements. Her vocalizations were mostly incomprehensible. A continuous, loud, high-pitched bruit was evident over both orbits and the entire cranium, with a right hemicranial predominance. Bilateral proptosis, chemosis, periorbital edema, and frontal venous engorgement were present. Intraocular pressures, measured with a Schiotz tonometer, were elevated bilaterally. Discs were sharp, and spontaneous venous pulsations were present. Extraocular movements were roving, and the oculocephalic response was intact. Pupils were equal and reactive and corneal reflexes were present. There was no localized paresis or asymmetry of withdrawal from noxious stimulation. The longstanding cranial asymmetry, left-sided hyperreflexia, and reduction in left leg muscle bulk were verified. The left plantar response was extensor and the right was flexor.

EEG showed diffuse slowing, no paroxysmal activity, and invariance to auditory, photic, and painful stimuli. Non-enhanced CT scan (fig. 1A) showed a large fusiform structure of pooled-blood density in the region of the quadrigeminal cistern and straight sinus. A serpiginous lesion of similar density in the right occipital region, with surrounding areas of tissue loss, was noted. Contrast CT (fig. 1B) revealed enhance-
Nonenhanced CT scan shows lesion of pooled-blood density in the regions of the quadrigeminal cistern, straight sinus, and right occipital lobe. Note adjacent areas of tissue loss.

FIGURE 1A: Nonenhanced CT scan shows lesion of pooled-blood density in the regions of the quadrigeminal cistern, straight sinus, and right occipital lobe. Note adjacent areas of tissue loss.

ment of the abnormal structures consistent with a large right occipital AVM draining into a dilated vein of Galen. No subarachnoid blood was apparent. The patient was begun on steroids, phenytoin, and, for suspicion of septic cavernous sinus thrombosis, antibiotics.

Over the ensuing hours her lethargy increased. Vocalizations became entirely incoherent; spontaneous movements were purposeless, although withdrawal to pain was preserved. A right gaze preference with incomplete ocular excursions to the left on doll's head maneuver evolved. Corneal reflexes became diminished; deep tendon reflexes were exaggerated on the right as well as the left.

Right and left internal carotid, right external carotid, and left vertebral angiograms were next performed. The AVM was supplied by multiple branches of the inferior division of the right middle cerebral artery, both pericallosal arteries, and both posterior cerebral arteries. In addition, there was supply from the external carotid circulation via a posterior branch of the right middle meningeal artery. Rapid fistulous shunting of blood into the vein of Galen — straight sinus system was noted (fig. 2). Sagittal, sigmoid, and transverse sinuses were patent. There was early filling of the cavernous sinuses from runoff of the AVM via the petrosal sinuses and accessory venous collaterals. Filling of the left cavernous sinus was incomplete compared to the right.

This study was followed by orbital venography (fig. 3). Contrast was injected into a frontal vein with simultaneous compression of the supra-orbital, angular, and facial veins. There was no spontaneous filling of either the superior ophthalmic veins or the cavernous sinuses.

Over the succeeding two days the patient began to respond appropriately to verbal stimuli. Extraocular movements became full, intraocular pressures declined, and the periorbital swelling and chemosis diminished. By her sixth hospital day, all vocalizations were comprehensible, movements were fully purposeful, all of her eye findings had resolved, and the cranial bruit was barely discernible. Cultures proved negative and antibiotics were discontinued. She was discharged on the eleventh hospital day with no exacerbation of her pre-existing motor deficit, and no overt change in her cognitive abilities.

Three months post-discharge, she was readmitted after an accidental blow to the left occiput precipitated a nearly identical combination of mental status changes and ocular findings. CT scan showed no interval change. Complete resolution followed several days of conservative therapy.

Discussion

Arteriovenous malformations have long been recognized as the most common vascular disorder of the CNS in childhood. While as many as one half are asymptomatic, the presenting features of the remainder are diverse, ranging from headache, behavioral abnormalities, and an audible bruit (of which the child...
herself may complain), to heart failure (in the very young), intellectual deterioration, and hemorrhagic stroke. This case presents another, less common initial feature, that of unexplained cerebral palsy. The location of the lesion contralateral to the limb involvement is topographically consistent, and the association of cranial asymmetry with AVM’s has previously been recognized. Although calvarial enlargement is more commonly found ipsilateral to the lesion, the magnitude of tissue loss adjacent to the AVM explains the pattern in this patient. Such tissue changes are thought to be due to the pulsatile effect of the lesion, vascular steal, or prior unrecognized hemorrhage. The last is not uncommon; eleven percent of Stein’s series of 55 patients had evidence of silent rupture. The morphology of this AVM, with its secondary dilatation of the vein of Galen, must be distinguished from a close relative, the primary vein of Galen aneurysm. The two lesions have different clinical characteristics (hydrocephalus and congestive heart failure are more common in the latter), and therapeutic approaches (the AVM, not the dilated vein, is the target of therapy in the former).

Although head trauma clearly precipitated the patient’s second episode, the cause of her first decline is unknown. Radiologic and CSF examinations did not disclose hemorrhage, and clinical and EEG observations did not reveal seizure activity. Vigorous hydration may have played at least a permissive role. The fulminant clinical picture would suggest that hemodynamic changes had occurred within the AVM and surrounding tissues. The vessels within these lesions are known to be structurally abnormal, showing thinning and hypoplasia on the one hand, and hyalinized thickening on the other. This suggests that their autoregulatory function may be impaired. Several workers, including Nornes and Grip, have validated this concept using Doppler techniques and pressure recordings.

Abnormal hemodynamics in this patient were reflected by the clinical appearance of a cavernous sinus syndrome, with the concomitant radiographic demonstration of (1) reversal of flow in the petrosal sinuses due to runoff from the AVM, resulting in (2) arterialization of flow into the cavernous sinuses, and (3) resistance to normal facial and orbital venous drainage into these structures. Furthermore, because of incomplete filling of the left cavernous sinus, partial thrombosis on that side was probable.

The association of venous sinus disease with AVM raises the possibility that the former may be the cause, rather than the consequence, of the latter. Houser et al reviewed 14 cases of AVM’s involving the dural venous sinuses, most of which showed evidence of sinus thrombosis. In two patients, occlusion of the transverse or sigmoid sinuses occurred before the de-
development of the AVM, and in all of the cases there was lack of sinus dilatation. They concluded that these AVM's represented acquired lesions due to neovascularization after sinus thrombosis.

A number of features in our case, however, indicate that the AVM was primary. First, clinical signs referable to the hemispheric lesion antedated those referable to the cavernous sinus by many years. Second, the topography of the AVM, with generous runoff into the vein of Galen and straight sinus system, is more in keeping with a congenital than an acquired lesion. Third, the tissue loss associated with the AVM reflected a long-standing process, whereas the peri-cavernous tissues were radiographically normal. Fourth, unlike the two cases cited above, the locus of sinus involvement was distant from the AVM. Finally, the cavernous sinus syndrome was both transient and recurrent, with an intervening period during which no symptoms or signs of cavernous sinus disease were manifest.

Several clinical points are emphasized by this case. First, evaluation of all patients with unexplained cerebral palsy should include auscultation for bruits; if a bruit is not heard, exercising the patient before repeat auscultation may serve to uncover it.

Second, the presence of cranial asymmetry in a child with abnormal neurologic signs, however apparently static, must raise the possibility of AVM. Although these lesions are usually ipsilateral to the side of enlargement, such is not always the case, as illustrated here. Third, cavernous sinus syndromes, particularly when appearing in a non-infectious context, should suggest AVM along with the more typical possibilities, such as carotid-cavernous fistulas. Finally, AVM's may cause sudden neurologic decline in the absence of subarachnoid hemorrhage and overt seizure activity. Changes in local hemodynamics, whether precipitated by head trauma or other less obvious causes, may be responsible.

References
Recurrent cavernous sinus syndrome complicating supratentorial arteriovenous malformation: report of a case.
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