Alterations in ventricular size and intracranial pressure caused by sagittal sinus pathology in man

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Lumbar cerebrospinal fluid (CSF) pressure and ventricular size were determined in six patients with impairment of cerebral venous outflow caused by either sagittal sinus thrombosis or arteriovenous shunting into the sagittal sinus. None of the patients had enlargement of the ventricular system, but all six had elevated CSF pressure (mean, 30 mm Hg). At least two mechanisms sufficient to prevent ventricular enlargement and significant hydrocephalus are suggested by these cases: 1) intracranial pressure elevations that occur as a result of cerebral venous outflow impediment maintain a positive pressure gradient between the intracranial CSF and the sagittal sinus contents, thereby increasing bulk CSF outflow; 2) in adults, increased function of alternative CSF outflow pathways occurs secondary to sagittal sinus thrombosis across the arachnoid villi of other intracranial vascular structures and in the spine. These mechanisms may have general importance in the generation of hydrocephalus caused by other disease states in adults but not in children.

KEY WORDS □ intracranial pressure □ hydrocephalus □ sagittal sinus □ arachnoid villi

VENTRICULAR enlargement following cerebral venous outflow obstruction has not been frequently observed in clinical practice or animal investigations. Early reports of iatrogenically altered venous outflow in man, such as deliberate ligation of the internal jugular vein for cancer or war wounds, resulted in death or severe neurological deficits. However, more recent reports relate unilateral or bilateral ligation of the jugular vein to chronic states of clinical intracranial hypertension, nonfatal neurological complications, or to the absence of symptoms. These authors did not specifically address the issue of ventricular enlargement or intracranial hypertension.

In his report on hydrocephalus and the dural venous sinuses in man, KINAL concluded that the clinical entities that affect the dural sinuses may result in hydrocephalus in a small percentage of cases, although often producing states of chronic intracranial hypertension.

A deliberate attempt to produce intracranial hypertension and hydrocephalus in 10 dogs by occlusion of the straight sinus or vein of Galen was attempted by Dandy and Blackfan. Hydrocephalus occurred in only one animal. Many authors subsequently verified this work by failing to produce hydrocephalus in dog and cat following experimental venous outflow blockage. However, observed well defined hydrocephalus in 74% of dogs in which cerebral venous drainage was obstructed by ligation of cervical veins and the major anastomotic channels to the spinal venous system in these animals. They concluded that chronic increased intracranial venous pressure could produce communicating hydrocephalus in man.

We present six patients in whom ventricular size remained normal in spite of venous outflow impairment caused by sagittal sinus pathology. The intracranial pressure (ICP) was carefully measured in each case and found to be elevated.

Two mechanisms considered sufficient to prevent hydrocephalus following sagittal sinus pathology in man were suggested by the findings in our patients. We discuss below these mechanisms as well as the...
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TABLE 1
Clinical findings in six patients

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Duration of Symptoms</th>
<th>Symptoms</th>
<th>Neurological Examination</th>
<th>Lumbar CSF* Pressure</th>
<th>Laboratory Data</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>55, M</td>
<td>6 mos</td>
<td>headache, visual impairment, mild confusional state</td>
<td>papilledema</td>
<td>&gt; 30 mm Hg</td>
<td>normal</td>
</tr>
<tr>
<td>2</td>
<td>22, M</td>
<td>1 yr</td>
<td>headache, diplopia, vomiting, visual impairment</td>
<td>papilledema, sixth nerve deficit</td>
<td>&gt; 35 mm Hg</td>
<td>thrombocytosis (700,000 platelets/cu mm)</td>
</tr>
<tr>
<td>3</td>
<td>20, M</td>
<td>hours</td>
<td>seizures, dysphasia, right hemiparesis, prompt resolution of symptoms</td>
<td>papilledema</td>
<td>&gt; 25 mm Hg</td>
<td>hyperfibrinogenemia</td>
</tr>
<tr>
<td>4</td>
<td>8, M</td>
<td>3 mos</td>
<td>headache</td>
<td>papilledema</td>
<td>&gt; 25 mm Hg</td>
<td>normal</td>
</tr>
<tr>
<td>5</td>
<td>45, M</td>
<td>5 mos</td>
<td>headache, diplopia, visual impairment</td>
<td>papilledema with retinal hemorrhage</td>
<td>&gt; 20 mm Hg</td>
<td>normal</td>
</tr>
<tr>
<td>6</td>
<td>40, F</td>
<td>3 days</td>
<td>visual impairment, loss of vision</td>
<td>remarkable papilledema</td>
<td>50 mm Hg, plateau waves</td>
<td>normal</td>
</tr>
</tbody>
</table>

*CSF = cerebrospinal fluid.

issue of the patient's age in the production of hydrocephalus following cerebral venous outflow impairment.

Clinical Material and Methods

Case 1

This 55-year-old man presented with a 6-month history of headache, mild confusional state, and decreased visual acuity. Neurological examination revealed marked papilledema. All laboratory studies were normal. The lumbar CSF pressure was monitored with a lumbar subarachnoid catheter, and was consistently above 30 mm Hg (Table 1). Angiography revealed an arteriovenous malformation (AVM) draining the torcular herophili and posterior third of the superior sagittal sinus (Fig. 1). Ventricular enlargement was not evident either in the angiogram or in computerized tomography (CT) of the head.

Case 2

This 22-year-old man presented with a 1-year history of headache, vomiting, diplopia, and visual impairment. Neurological examination revealed marked papilledema and sixth nerve palsy. Laboratory studies were consistent with thrombocytosis (700,000 platelets/cu mm). The lumbar CSF pressure was over 35 mm Hg (Table 1). Massive thrombosis of the superior sagittal sinus and normal ventricular size were two features present on the angiogram (Fig. 2).

Case 3

This 20-year-old man was promptly admitted after the onset of Jacksonian seizures followed by dysphasia and right hemiparesis. These neurological symptoms promptly cleared, but the papilledema persisted.

FIG. 1. Case 1. Angiogram showing a hypertrophic occipital artery, a prominent posterior branch of the superficial temporal artery, and tentorial vessels from the presellar portion of the internal carotid artery feeding an arteriovenous malformation, which drains into the torcular herophili and posterior third of the superior sagittal sinus.
Flc. 2. Case 2. Angiogram showing massive thrombosis of the superior sagittal sinus. There is a flow inversion in the frontal and parietal ascending veins draining into the cavernous sinus.

Laboratory studies demonstrated hyperfibrinogenemia. The lumbar CSF pressure was 20 mm Hg (Table 1). Lack of filling of the superior sagittal sinus and normal ventricular size were seen on the patient's angiogram (Fig. 3).

Case 4
This 8-year-old boy presented with a 3-month history of headache. Neurological examination revealed papilledema. Laboratory studies were normal. The lumbar CSF pressure was about 20 mm Hg (Table 1). Angiography showed abnormal dilatation of the superior sagittal sinus, torcular, and vein of Galen.

Case 5
This 45-year-old man developed headache, diplopia, and visual acuity impairment 5 months after head...
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trauma. Neurological examination revealed papilledema with retinal hemorrhages. All laboratory data were normal. The ICP was above 20 mm Hg. A lumbar infusion study (0.76 ml/min) had to be discontinued because of prompt onset of abnormally elevated ICP (Table 1). Cranial tomography revealed a depressed fracture at the vertex, and angiography was consistent with partial thrombosis of the superior sagittal sinus (Fig. 4). Normal ventricular size was verified by the angiogram and by CT scan.

Case 6

This 40-year-old woman presented with a 3-day history of rapidly progressive diminution of visual acuity. She was taking oral contraceptives. Neurological examination revealed marked papilledema. Laboratory data were normal. The lumbar CSF pressure was over 50 mm Hg with plateau waves (Table 1). Cerebral angiography was consistent with thrombosis of the posterior two-thirds of the superior sagittal sinus (Fig. 5). Ventricular size was normal as seen on the angiogram and CT scan.

Summary of Cases

The average age of our patients was 31 years. There were no patients younger than 8 years old. The average time from onset of symptoms to presentation at the hospital was over 4 months. All patients presented with headache and papilledema, and four (66%) had decreased visual acuity. Besides decreased visual acuity and the sixth nerve palsy in Case 2, neurological examination revealed no other major neurological deficits. Case 1 presented with a mild confusional state, but none of the other patients had an alteration of mental status. Elevated lumbar CSF pressure was a feature in each of the patients; the mean pressure was 30.83 mm Hg on admission to the hospital. None of the patients had evidence of ventricular enlargement. Electroencephalograms (EEG's) were normal in this series of patients. Table 1 summarizes the clinical findings in all six patients. The outcome was favorable in all but one of the patients; a severe visual acuity deficit has persisted in Case 6.

Discussion

In the patients presented here, either obstructive processes or abnormal shunting of blood altered venous pressure in the sagittal sinus. Most of the experimental and clinical literature clearly indicates that an increase in venous pressure alone does not result in hydrocephalus in adults, despite the presence of intracranial hypertension. We believe that mechanisms able to prevent ventricular enlargement in patients with cerebral venous outflow impairment have been suggested by our cases and, further, that these mechanisms may have general importance with respect to the formation of hydrocephalus caused by other disease states.

Elevated Intracranial Pressure

There are three intracranial components — brain tissue, blood, and CSF — that could be implicated in the production of intracranial hypertension if pathologically altered. Alteration of brain tissue is probably the least likely basis for elevated CSF pressure in our patients. Venous hypertension of sufficient magnitude could elevate hydrostatic capillary pressure, thereby producing an increase in net capillary filtration and progressive brain edema. In all but one of our cases, gross alteration of the mental status was absent, and the EEG was not consistent with significant cerebral edema. Moreover, typical findings of diffuse cerebral edema on CT scans, such as generalized, irregular, low-density zones in the parenchyma and compression of the ventricular and cisternal spaces, were absent.

It appears likely that alterations of intracranial blood and CSF volume are causative with respect to the elevated CSF pressures documented in our cases. The phenomenon of CSF absorption across the arachnoid villi is schematically described by the following classic relationship: absorption (CSF-bulk flow) = pressure (CSF-sagittal sinus/resistance (across arachnoid villi)).

In our patients, the only significant variable is the CSF-sagittal sinus pressure gradient, provided that resistance to CSF flow through the arachnoid villi is unaffected. Johnston and Paterson13,14 proposed that a reduction of CSF absorption can produce an increase in CSF intracranial volume, leading to an accumulation of excess CSF in the subarachnoid space. If CSF production does not fluctuate, CSF pressure can rise until its value overcomes the sagittal sinus pressure, thus restoring a positive CSF-sagittal sinus pressure gradient.

Apart from CSF, another source of excess intracranial volume capable of generating intracranial hypertension may be the blood in engorged cortical veins. After an increase in sagittal sinus pressure, an increase in total intracranial blood volume may occur because of impaired venous flow into the sagittal sinus.

It has been well documented in man and animal experimentation that despite gross elevations in both the intracranial and intrasinusoidal pressures, a positive CSF-sagittal sinus pressure gradient is almost always maintained.4,11,16,20,21,23,24,20,32 Lamas, et al.,18 reported a patient with raised ICP secondary to a dural AVM and direct shunting of blood in the transverse-sigmoid sinus. By recording simultaneously the sagittal sinus pressure and ICP, they found the values of the former to be considerably above normal but constantly lower than those of the latter. In effect, the preservation of
Alternative Cerebrospinal Fluid Pathways

It is difficult to understand how the maintenance of the CSF-sagittal sinus pressure gradient could effectively prevent hydrocephalus in patients with intracranial hypertension secondary to massive thrombosis of the superior sagittal sinus. We have presented four patients with complete functional exclusion of the sagittal sinus, who survived, and in whom ventricular enlargement was not present. In these cases, alternative CSF pathways through venous collateral vessels may have played a major role in preventing hydrocephalus. The CSF drainage routes from intracranial sources to the scalp veins, from the cavernous sinus to the external facial veins, from the sphenoparietal sinus or the pterygoid sinus, as well as from posterior fossa vascular structures to the basilar vertebral plexus could all become important.

All of these collateral pathways exclude the areas commonly believed to be the most important in absorbing CSF, that is, superior sagittal sinus and adjacent veins. We wish to emphasize the importance that these alternative routes of CSF absorption may assume when normal pathways of CSF drainage are occluded.

The quantitative capacity of the spinal arachnoid villi and of other alternative pathways of CSF reabsorption is unknown. However, we suggest that the absence of hydrocephalus in cases of complete functional exclusion of the sagittal sinus and adjacent cortical veins may be a possible argument in favor of their real effectiveness in assuring an adequate CSF reabsorption in such pathological situations.

Patients' Age

It is important in our opinion to stress the age of the patient, because many cases have been reported of infantile hydrocephalus following increased central and/or cephalic venous pressure. Haar and Miller reviewed the literature on patients with dural sinus hypertension of various etiologies in which ICP and ventricular size had been reported. They separated the reported cases into two groups, the first with hydrocephalus and the second with normal or small ventricles. These two groups differed in that most patients with hydrocephalus were less than 18 months old, while in the patients without ventricular enlargement the age varied from 3 to 48 years. Obrador, et al., in their review of 96 patients with AVM's or fistulas shunting arterial blood into the transverse-sigmoid sinus, found hydrocephalus in only 4% of patients, all infants, whereas headache, papilledema, and other symptoms due to increased ICP were noted in many more patients. An interesting explanation has been proposed by Young. He reported a case of an infant with a large dural AVM and communicating hydrocephalus, in which cisternographic findings indicated a grossly impaired CSF absorption. The author supposed that such an increase in intracranial venous pressure by massive shunting of arterial blood in the presence of a distensible skull does not result in a proportional increase in ICP as it would in the rigid skull of an adult. Thus, the elevation of ICP necessary to compensate for elevated sagittal sinus pressure cannot occur in infants, and hydrocephalus develops.

Another important difference between infant and adult CSF reabsorption is evident in the different features of the arachnoid granulations. They are macroscopically absent at birth, only existing as microscopic arachnoid villi. The villi are insufficiently developed until 18 months of age. By the age of 4 years, they are widely distributed in the dural venous channels and continue to increase after this age. In 1971, Gilles and Davidson reported two cases of communicating hydrocephalus associated with rudimentary and sparse or absent arachnoid villi in children. We suggest that, in early pediatric age groups, clinical situations that result in an increased cephalic venous pressure may overload the capacity of the arachnoid villi more easily than in adults, and hydrocephalus will develop.

Importance of Monitoring Intracranial Pressure

If the causal factor of elevated cerebral venous outflow pressure cannot be remedied promptly, the ICP must be carefully and continuously monitored. Values of ICP sufficiently high to significantly reduce cerebral perfusion pressure or produce neurological dysfunction must be treated. We believe that treatment of intracranial hypertension by repeated lumbar puncture without continuous monitoring of ICP should be undertaken with care, since lumbar CSF pressure may not accurately reflect ICP. The most advantageous method of treating patients with venous outflow obstruction and elevated ICP may be with intraventricular cannulation, since ICP values can be more accurately determined with an intraventricular cannula than with lumbar CSF pressure measurements.
Moreover, judicious treatment of intracranial hypertension when necessary by ventricular drainage can be accomplished with this technique.

References

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This work was supported in part by NINCDS Grant NS12587 and Teacher Investigator Award 5KO7 NS00346 to Dr. Greenberg.

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