Communicating hydrocephalus associated with deficient dysplastic parasagittal arachnoidal granulations

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Marked diminution of arachnoidal granulations with abnormality of those remaining is presented as a cause of communicating hydrocephalus in two children. Although this underlying abnormality has been alluded to in the past, it has not been adequately documented. The authors believe their observations constitute sufficient evidence to warrant consideration of this entity as one of the causes of communicating hydrocephalus.

KEY WORDS Pacchionian granulation · hydrocephalus · cerebrospinal fluid flow

ALTHOUGH obstruction plays the major etiological role in hydrocephalus, overproduction and malabsorption of spinal fluid are frequently mentioned as causative mechanisms, but with equivocal documentation. Absorptive defects at the arachnoidal granulation level are still hypothetical considerations, having been suggested as factors in the cause of the hydrocephalus by the older terms "otitic hydrocephalus" and "external hydrocephalus." Some patients with superior sagittal sinus thrombosis acquire hydrocephalus presumably as a result of absorption blockade. Ellington and Margolis have supported this hypothesis with a study of arachnoidal villi in subarachnoid hemorrhage, and Shabo and Maxwell have demonstrated that phagocytosis of red cells occurred within arachnoidal granulations following the introduction of endogenous erythrocytes into the subarachnoid space. Without citing specific cases, Cushing in 1914 mentioned that the hydrocephalus in some of his patients could be attributed to an "absence of arachnoidal projections or villi." Winkelman and Fay, in discussing the "Pacchionian system" and its possible relation to epilepsy, listed three patients with hydrocephalus and "aplasia" of the Pacchionian granulations, although they did not mention concurrent problems that might have caused the hydrocephalus.

The purpose of this report is to present two hydrocephalic patients, one with absence of parasagittal arachnoidal granulations, and the other with a markedly diminished number of arachnoidal granulations, in whom no other causes for the ventricular dilatation were found.
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Case Reports

Case 1

This baby boy was first seen at 7 months of age for gradually increasing head size. He was the product of an uneventful 38-week gestation and labor. The family history was unremarkable. There was no history of trauma or infection, and psychomotor development was said to be progressing normally. Admission weight was 5.8 kg, and the head circumference was 50 cm, 2 cm above the 97th percentile. The anterior fontanel was full, no bruits were heard, and there was no transillumination. Neurological examination was normal. Ventriculography and pneumoencephalography revealed a communicating hydrocephalus with air in the cisterna magna, preponitine cistern, and around the spinal cord. Ventricular fluid protein content was 17 mg%, and lumbar fluid protein was 23 mg%.

When the patient was 8 months old, a lumboureterostomy was performed, and he was discharged on a salt replacement regimen. At that time the head circumference was 49.5 cm; he had albuminuria but negative urine cultures. He did well with stabilization of his head growth curve, and by 28 months of age his head circumference was 50.4 cm (50th percentile). Although his parents had been alerted to the problem of severe hyponatremic dehydration that can occur with this shunt in association with emesis or diarrhea of any etiology, he appeared in a moribund state in the emergency room at 3 years of age after a 3-day history of rhinorrhea, cough, and emesis. He was severely dehydrated and had a serum pH of 7.21 (venous), sodium of 137, and BUN of 134. Cardiorespiratory arrest occurred from which he could not be resuscitated. The general autopsy findings revealed only evidence of severe dehydration.

Case 2

This baby boy was first seen at 3½ months of age because of irritability of 3 to 4 weeks' duration and progressive lethargy and poor feeding for 1 week. During the 2 weeks immediately before admission there had been a rapid increase in head size. Gestation and delivery at term had been unremarkable, with a birth weight of 8 lb. 10 oz. Development had been normal. A varus deformity of the right foot had been treated with a cast. There was no family history of neurological disease, and the child's four siblings were well. There was no prior history of trauma or febrile illness.

Examination on admission revealed a weight of 14 lb 5 oz, which was in the 80th percentile. The anterior fontanel was tense. A combined spinal fluid tap with phenolsulfonphthalein revealed ventriculolumbar communication in 6 minutes and an opening ventricular pressure of 450 mm of water. A ventriculogram confirmed this, revealing massive ventricular dilatation and enlargement of the cisterna magna. Air was present around the spinal cord. A lumboureterostomy was performed, and the patient was discharged in good condition on a salt replacement regimen. He was readmitted later that month because of acute obstruction of the shunt at the dural level. This was corrected without incident and he was again discharged well.

He was followed closely. He was underweight for his age, and his psychomotor development was somewhat delayed. Both head circumference and weight at 18 months of age were under the 10th percentile. Between the time of the shunt revision at 4½ months of age and his last examination at 38 months of age, the patient was admitted on four occasions, each time for respiratory or gastrointestinal infections, presumably viral, associated with emesis. He was felt to have a functioning shunt on each occasion and was treated for 1 or 2 days with intravenous fluids until spontaneous recovery took place. At 3 years 2 months of age he was found dead, 10 days following measles vaccination. He had had no prior symptoms and was described as being completely well the evening before death, save for a "slight fever." The general autopsy findings included pulmonary edema, chronic tracheobronchitis, focal interstitial fibrosis, and congestion of visceral organs and gastrointestinal tract. *Streptococcus, Neisseria,* and *Staphylococcus aureus* were cultured from the lung. Viral and bacterial cultures of other organs, blood, cerebrospinal fluid, and urine were negative.

Methods of Histological Study

The entire superior sagittal sinus from Case 1 was cut into blocks at 1 cm intervals from crista galli to torcular, processed, and
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embedded in paraffin. Each block was cut in serial sections at 10 μm. Every tenth section was stained with hematoxylin and eosin, and examined. At intervals of 1 cm, representative blocks of superior sagittal sinus at the vertex were taken from Case 2. Serial sections were cut, stained, and examined in the same fashion as in Case 1. For controls, sagittal sinus was studied from two normal individuals, aged 4 yrs and 4 yrs 8 mos. For this purpose, three and five 1-cm blocks respectively from the vertex of superior sagittal sinus were processed simultaneously with those from Cases 1 and 2.

Results

Detailed gross and microscopic examination of the leptomeninges, brain, and spinal cord revealed no obstructive lesions of congenital, inflammatory, or neoplastic nature that could have been responsible for the hydrocephalus in either case. No bone abnormalities were noted. The superior sagittal, straight, transverse, and sigmoid sinuses in both cases were patent and grossly did not contain arachnoidal granulations, nor were granulations seen over the parasagittal portion of the cerebral hemisphere. In both of the controls, on the other hand, granulations were readily seen in these locations upon gross examination.

Gross examination of the brain in Case 1 revealed old thin subdural hemorrhages and moderately swollen cerebral hemispheres. The brain weighed 1260 gm. The occipital horns and trigones were moderately dilated. The choroid plexus was normal in size, shape, distribution, and morphological appearance. There were no visible arachnoidal granulations in venous lacunae or dural sinuses. The shunt was patent. Microscopically there was moderate congestion of the centrum semiovale, and scattered foci ofependymal loss and subependymal gliosis were present in the ventricular wall. The aqueduct was rounded and widely patent. Fourth ventricular outlets were widely patent. The leptomeninges were thin and delicate throughout. Specifically, there was no desmoplasia of perimedullary, prepontine, basilar, or convexity leptomeninges.

The serial sections of the entire sagittal sinus and adjacent dura of Case 1 contained a total of five arachnoidal granulations, all of which were abnormal. Sinus endothelium was intact throughout. Small collapsed lateral lacunae were identified but were considerably reduced in both size and number. One of the abnormal granulations consisted of strands of arachnoidal mesothelium which penetrated dura adjacent to the sagittal sinus but neither reached the sinus nor lateral lacuna and did not form a normal granulation. A second rudimentary granulation arose from the surface of the arachnoid but did not penetrate the dura. The third abnormal granulation was composed of a small mass of arachnoidal mesothelium which penetrated dura but did not encounter an endothelial-lined lateral lacuna nor did it penetrate sagittal sinus. The fourth and fifth granulations were composed of pegs of mixed arachnoidal and connective tissue which approached a lateral lacuna but did not enter it or the sagittal sinus (Fig. 1). The sagittal sinus in its midpoint was divided by a median fibrous septum for a short distance, but otherwise appeared normal. Entry points of bridging veins were encountered, which were sometimes partially surrounded by arachnoidal tissue but were not associated with granulations. Small numbers of dural arterioles, veins, and nerves were encountered. Relatively little dural adipose tissue was present.

Grossly, arachnoidal granulations were absent over the external surface of the brain.
and along the sagittal sinus in Case 2 (Fig. 2). All sinuses were patent. The brain was of normal weight (1300 gm). The usual frontal and temporal gyral pattern was not present but was replaced by a random distribution of gyri. The cerebellum was somewhat compressed in a rostral-caudal axis and bore a groove on its inferior surface extending around the hemispheres. The lateral ventricles were dilated, and a persistent cavum septi pellucidi was present. The lining of the entire ventricular system was the site of a widespread granular ependymitis with ependymal stripping and nodules of subependymal astrocytes. The choroid plexus was normal in size, shape, distribution, and morphological appearance. Although the cerebellar tonsils extended laterally around the medulla, all fourth ventricular outflow foramina were patent. One vertebral artery was encased by fibrous leptomeningeal thickening, and its lumen was occluded by subintimal fibroblastic thickening. There was no generalized leptomeningeal desmoplasia.

Microscopic examination of the serial sections of the blocks from the vertex sagittal sinus failed to reveal a single arachnoidal granulation. The sinus was of normal size and shape. Small numbers of collapsed lateral lacunae were encountered. All were lined by an endothelium; junctures with the sagittal sinus were demonstrated. The dura overlying the sagittal sinus contained the usual number of small arterioles, veins, and nerve filaments. A relatively large number of fat cells was interposed between the inner and outer layers of the dura immediately over the sinus.

The sagittal sinus examined in these two cases stood in marked contrast to that of the "controls." In each of the eight blocks of vertex superior sagittal sinus obtained from
the two controls, normal arachnoidal granulations were readily found.

Discussion

The history of the early observations of arachnoidal granulations was the subject of a review by Turner in 1961. In April, 1705, Pacchioni first published his observations on the structures that now bear his name, calling them “conglomerate glands of the dura mater.” He later acknowledged his predecessor Mery who had published a thesis on these granulations in 1701. Other authors, including Vesalius in 1543, also had mentioned these “glandulae.” Luschka first pointed out that the Pacchionian or arachnoidal granulations were hypertrophied arachnoidal villi normally present in all brains. Key and Retzius described these structures within the context of the anatomy of cerebrospinal fluid circulation and considered them to be passive filters of cerebrospinal fluid into venous channels.

Weed in 1914 introduced isotonic solutions of iron ammonium citrate and potassium ferrocyanide into the spinal subarachnoid space in dogs and cats and at various intervals sacrificed his specimens, perfusing the carotid vessels with a formalin and acid mixture. Precipitated Prussian blue was found in arachnoidal granulations. His work forms the basis for the concepts of the circulatory pattern of spinal fluid flow now accepted by most authors, i.e., the Pacchionian granulation is a structure necessary for cerebrospinal fluid absorption. He also described accessory pathways of absorption via the perineural lymphatics of cranial nerves and arachnoid vessels. The mechanism by which spinal fluid passes through the granulation and into the venous system is the topic of much recent discussion, but is not immediately germane.

Arachnoidal granulations are not visible grossly at birth but are present as microscopic arachnoidal villi. They first become apparent on close inspection with a hand lens at 6 months, and at about 18 months are visible grossly, appearing first in regions where the parietooccipital and central veins open into the superior sagittal sinus near the vertex. They then become more noticeable with advancing age, spreading anteriorly and posteriorly along the superior surface of the cerebral hemisphere. By the third or fourth year they are widely distributed in the dural venous channels, and form conspicuous nodules in the lumen.

The location of granulations in adults is, in order of frequency, the superior sagittal sinus (including lateral lacunae), transverse sinus, cavernous sinus, superior petrosal sinus, middle cerebral vein, sphenoparietal sinus, straight sinus, and occasionally the torcular. The superior sagittal sinus contained by far the largest number of granulations per given volume of any of the sinuses. In 3- to 4-year-old children who died of non-neurological disease, more than 50 large arachnoidal granulations can be counted grossly along the superior sagittal sinus. The younger the subject from whom the specimen is taken, the more cellular and less numerous are the arachnoidal granulations. Under light microscopy the arachnoidal granulations have been described as invaginations of the dural walls of the sagittal sinus or lateral lacuna with a core of very cellular leptomeningeal tissue. Turner was able to demonstrate their anlage in an 80 mm embryo. Shabo and Maxwell later described the electron microscopic appearance of the arachnoidal granulation in the adult Macaca. It consisted of a core of arachnoidal cells and attenuated collagen bundles, a subendothelial space continuous with the subarachnoid space via lacunae between the arachnoidal cells, and an endothelial covering continuous with the lining of the sinus.

The changes found in these two cases, namely, apparent absence of the granulations (Case 2) along the sagittal sinus, and marked diminution in the total number as well as failure of the remaining five rudimentary granulations to complete a union with either a lateral lacuna or the sagittal sinus, are consistent with a primary dysplasia or developmental failure of this structure. Neither in our experience nor in that of others do we find support for the contention that hydrocephalus, per se, causes these abnormalities in the arachnoidal granulation. Moreover, experimentally increased intracranial pressure in the dog causes changes in the arachnoidal and endothelial cells of the granulation, but apparently not massive loss of granulations. On the other hand, the total number of granulations may be related to...
bulk cerebrospinal fluid flow. In three cases of congenital aqueductal stenosis, there was a decrease in number and size of granulations, but those remaining bore appropriate relationships to the lateral lacunae or sagittal sinus.6 One case of acquired aqueductal stenosis secondary to a mesencephalic tectal glioma failed to show any change in number or size.6 Furthermore, arachnoidal granulations were present in cases with shunts in place for long periods of time in which the hydrocephalus was related to a Chiari I malformation, postmeningitic leptomeningeal fibrosis, arteriovenous malformation, and multiple cases of tumor including medulloblastoma and diencephalic glioma.6

Conclusions

While it is clear that the abnormality of arachnoidal granulations may have arisen from either failure of development or a subsequent destruction of pre-existing granulations, no evidence for the latter was found. In view of the rudimentary nature of the granulations in Case 1, their failure to penetrate the lateral lacunae or sagittal sinus, the apparent decrease in number and size of lateral lacunae, and the associated abnormality of gyral pattern in Case 2, we favor the suggestion that the arachnoidal granulations failed to develop properly in these two children. If that is the case, then the delayed clinical appearance of hydrocephalus may be considered to reflect any of several mechanisms. Although we are unable to find information relating to changes in the rate of cerebrospinal fluid production in the first few months of human postnatal life, we believe that a change in the choroid plexus mass or an increased rate of production of cerebrospinal fluid might have overloaded a minimal perinatal absorptive mechanism after several months of life.

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