Chiari I “malformation”—an acquired disorder?

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Caudal herniation of the hindbrain, indistinguishable from the Chiari I deformity, may occur after the establishment of spinal subarachnoid shunts and become symptomatic years after the procedure. Examples are presented and others are cited from the literature. It is proposed that the force responsible for the displacement is the difference in pressure between the cranial and spinal compartments. On the basis of these observations and other considerations as well, a similar process, disproportionate absorption of cerebrospinal fluid from the spinal region, might account for the spontaneous form of the Chiari I deformity.

KEY WORDS hindbrain herniation □9 Chiari I deformity □9 syringomyelia □9 spinal subarachnoid shunt

Chiari described and numbered several kinds of caudal ectopia of the hindbrain. His Type I, now commonly known as Chiari I deformity, involves displacement of the tonsils of the cerebellum and, to a lesser extent, the remainder of the hindbrain. His Type II malformation (called Chiari II, Arnold-Chiari, or Cleland-Arnold-Chiari malformation) comprises caudal displacement of the more medially situated parts of the inferior cerebellum, together with the brain stem. Chiari considered both types to represent herniation of the fetal brain due to hydrocephalus, a suggestion that has often been repeated by others and, as elaborated by Gardner, may characterize prevailing opinion. There is, of course, the other view, as enunciated by Russell, that the hindbrain hernia is the cause of the accompanying hydrocephalus. The various theories of morphogenesis have been reviewed by Peach, Brocklehurst, and Caviness.

In this paper, we describe acquired chronic downward herniation of the hindbrain, anatomically indistinguishable from the Chiari I deformity, with symptomatic onset years after the institution of spinal subarachnoid shunts. As the result of these observations and others cited from the literature, we present a hypothesis that disproportionate absorption of cerebrospinal fluid (CSF) from the spinal subarachnoid space, with or without hydrocephalus, may account for the genesis of the Chiari I deformity.

Case Reports

Chiari Deformity Consecutive to Spinal Shunting

Case 1. At 18 years old, this woman experienced the onset and progression of dysesthesia and weakness of her legs. This led to myelography and laminectomy at another hospital. The lesion was an intramedullary tumor between C-7 and T-5 and, after biopsy, irradiation was administered with 4400 rads delivered over a 33-day period. The condition of her legs continued to deteriorate, and an "arachnoid cyst" at the site of operation was fenestrated, an operation complicated by infection. After the infection was cured, a cystoperitoneal shunt was instituted.

At 21 years of age, her arms became weak and examination disclosed weakness from the C-6 level downward, with total motor and sensory loss below the midthoracic area. In addition to a cervical-dorsal kyphosis, the intramedullary tumor, and the cystic space, a tonsillar herniation was demonstrated by metrizamide myelography (Fig. 1 left). The herniated tonsils were also outlined with metrizamide enhancement on computerized tomography (CT), which
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Fig. 1. Case 1. Left: Metrizamide myelography of lateral C1–2 puncture. This near-midline lateral polytome reveals the caudal cerebellum placed just below the C-1 level (arrow). The upper spinal cord is surrounded by metrizamide, below and anterior to the arrow. The expanded midcervical cord could be seen on midline tomograms. Right: Gas myelogram 8 months after myelogram shown at left. The cerebellar tonsils are in normal position.

showed slit ventricles as well. The herniated tonsils were decompressed, the tumor (an astrocytoma) was removed, and the shunt was ligated. During the same procedure a spinal fusion was performed. There were many postoperative complications, including atelectasis, vocal-cord paralysis, difficulty in swallowing, and Enterobacter meningitis. Later, because of further deterioration in the function of her arms, the cystoperitoneal shunt was reinstated, but not before gas myelography had shown the tonsils to have resumed a normal position (Fig. 1 right).

At present, she has good function at the C-5 and C-6 levels, and improving function to C-8, but is otherwise quadriplegic. She has given up her place as an art student but continues to paint.

Case 2. This boy was first admitted at the age of 9 weeks because of abnormal growth of the head, the hydrocephalus being due to a confirmed neonatal hemorrhage. A ventriculogram revealed communicating hydrocephalus with a generous cisterna magna (Fig. 2 left). A lumboureteral shunt was placed (J.S.). At 7½ years of age, the patient developed headache, vomiting, and lethargy, with signs of dysfunction in the fifth, seventh, and twelfth cranial nerves on the left. A pneumogram at that time revealed herniated tonsils (Fig. 2 right).

A suboccipital craniectomy revealed the tonsils of the cerebellum to be herniated to below the arch of C-1, the whiteness of their tips betraying the chronic nature of the change. The shunts were subsequently changed and now, at the age of 20, the patient is dependent upon a ventriculoperitoneal shunt. He is asymptomatic and is regularly employed.

Case 3. This patient was born by breech delivery, the second of male twins. At the age of 2 years a lumboureteral shunt was placed for hydrocephalus. His interval history was marked by an episode of meningitis and the correction of an atrial septal defect.

When he was 20 years old, he began to experience loss of balance and weakness of his feet. Examination 6 months later showed generalized muscle weakness (most marked in the plantar muscles) and dorsiflexion of the feet, absent tendon reflexes, and impaired position and pin-prick sensitivity in the legs. There was nystagmus on lateral gaze in each direction and bilateral cerebellar ataxia in the arms. Studies revealed tonsillar herniation and a cervical syrinx, and a suboccipital decompression was carried out. The tonsils of the cerebellum were seen to be herniated to the level of C-2. The inferior 5 mm of the left tonsil was white. As much tonsillar tissue as seemed safe
was amputated. The cervicomedullary region at 11 months and at 21 years are shown in Fig. 3.

Because his condition did not improve, the lumbar shunt was replaced by a ventriculoperitoneal shunt. He continues as a full-time college student.

Case 4. This child was born after a pregnancy complicated by uterine hemorrhage. When he was 4½ months of age, enlargement of the head was noted. A ventriculogram at 6 months of age revealed hydrocephalus, and a generous cisterna magna in communication with a clear high cervical subarachnoid space (Fig. 4). Exploration of the posterior fossa confirmed the presence of a large cistern, and it was specifically noted that there was no Chiari deformity. A lumboureteral shunt was placed at 8 months of age, and during the following years there were several episodes of hyponatremic dehydration.

At the age of 7 years, the patient showed instability of gait, and over the next 2 years an asymmetrical spastic quadriparesis developed, more on the right than the left, together with a suspended sensory deficit involving the right side of the face and C2-4. The diagnosis of syringomyelia was entertained. A laminectomy was made through C-3, and the old posterior fossa craniectomy was reopened. The cerebellar tonsils were found to be herniated to the C1-2 interspace on the left and to C-1 on the right. Although the upper cervical cord seemed enlarged, no fluid could be aspirated.

A year later, the lumboureteral shunt was changed to a ventriculoureteral shunt, and, after many urinary tract infections involving the ventricles, a ventriculoperitoneal shunt was substituted. A progressive subluxation of the upper cervical spine required fusion when the patient was 16 years old. Now, at 22 years of age, he has a right hemiparesis, but works regularly.

Case 5. This patient underwent a lumboureteral shunt for communicating hydrocephalus when she was 4 months of age. Until she was 1 year old, there were a number of episodes of hyponatremic dehydration. Streptococcal meningitis occurred when she was 25, 28, and 29 years of age.

As she approached her 32nd birthday, she complained of numbness on the chest wall on the right and pain in the left arm when she coughed. The symptoms progressed over several months and, when admitted to the hospital, she showed suspended sensory loss from C-3 to T-6 on the left, and T-7 to L-2 on the right. There was areflexia in the left arm. Mild scoliosis was noted.

Metrizamide myelography revealed an enlarged spinal cord and cerebellar herniation to C-2. Delayed CT showed filling of the syrinx by metrizamide. The lumbar shunt was replaced by a ventriculooarial one. Postoperatively, there was increased dysesthesia on the right side and pain in the left arm with return of tendon reflexes in that arm. Repeat myelography 4 weeks postoperatively revealed that the cord was smaller, and no contrast material was seen within the cord upon delayed imaging. Her pain has improved.

Fig. 2. Case 2. Left: Ventriculography at 9 weeks of age. The lateral ventricles were significantly dilated and the air communicated to the cisterna magna. The cerebellum was entirely above the foramen magnum (arrows). Right: Pneumoencephalogram at age 7 years and 7 months. The cervical cord can be seen and the cerebellar tonsils are at the C-1 level (arrow). The lateral ventricles filled and were seen to be much less dilated than at 9 weeks of age.
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FIG. 3. Case 3. Left: Pneumoencephalogram at 11 months of age. The fourth ventricle (x) is seen as slightly dilated and normally placed, as part of the communicating hydrocephalus. The cisterna magna is well visualized. None of the cerebellum is below the level of the cisterna magna. Right: Metrizamide myelography by lumbar puncture, 20 years later. There is a projection of the cerebellum extending down to the C-2 level (arrow).

and her activities were interrupted only during her hospitalization.

Summary

Four instances of chronic herniation of the hindbrain, especially of the cerebellar tonsils, were confirmed by contrast studies and surgical exposure years after the institution of spinal subarachnoid shunts. In three of these, gas-contrast studies had shown no herniation originally and, in one of the three, direct surgical exposure had confirmed that finding. In another case, in which herniation was thought to have been of briefer duration, the hernia was reduced after ligation of the shunt. In a fifth patient, symptoms and signs of syringomyelia appeared almost 32 years after lumboureteral shunt placement: the enlarged spinal cord and cerebellar displacement were demonstrated by contrast studies alone.

Discussion

We report here chronic displacement of the hindbrain, identical in form to the Chiari I deformity, occurring in the wake of spinal shunting. This sequence was described in 1976 by Hoffman and Tucker, who believed the displacement depended upon disproportionate growth of the brain in relation to that of the skull. Fischer, et al., reported three instances in which syringomyelia appeared years after lumboureteral shunting for hydrocephalus; the location of the cerebellum was known in none of these cases, nor was it mentioned in the two instances in which a syrinx was found at autopsy in the patients described by Kushner, et al. The latter authors described the occurrence of kyphoscoliosis after spinal shunting and considered the possibility that syringomyelia might be responsible.

In the search for clues to the origin of the Chiari I deformity, certain constraints must be respected. The
first is concerned with the development of the cerebellum, as described by Larsell and Jensen and Lemire, et al. The central part of the cerebellum or its vermis develops far in advance of the cerebellar hemispheres so that, at least until beyond the 200-mm stage (at about 5 months gestation), the development and situation of the cerebellar tonsils are such that they could hardly be in the forefront of a cerebellar hernia. At the earlier stages of gestation, such herniation involves the vermis of the cerebellum (Chiari II malformation). The beginning of displacement was described by Barry, et al., in a 49-mm fetus, and more advanced deformities were encountered in others up to 160 mm.

A second point is that the Chiari I deformity is usually discovered in adolescence or beyond. It is rare in early childhood or infancy and is unknown, or virtually so, in the newborn. Chiari’s youngest case was diagnosed at 3½ months, and we have seen the lesion as an incidental finding at autopsy in a baby of the same age. The patients of Appleby, et al., were 17 to 54 years of age, a range not dissimilar to that found by Gardner and Goodall: 14 to 54 years.

The third constraint is that, contrary to the belief of Chiari and others who have considered the matter, hydrocephalus is not an essential feature of the Chiari I deformity. In eight of the 22 cases of Gardner in which gas entered the ventricular system, the ventricles were normal in size. If, as theorized by Gardner, the dislocation of the cerebellum was due to a transient hydrocephalus following upon the closure of the neural tube, that herniation would not involve the still undeveloped cerebellar tonsils.

In the latter connection, mention ought to be made that Cameron, recognizing that the Chiari II malformation is not always accompanied by hydrocephalus, ascribed the fetal cerebellar herniation to lowered intraspinal pressure due to leakage of CSF at the myelocoele. That view was endorsed by van Hoytema and van den Berg and by Emery and MacKenzie, who correlated the degree of herniation with the size of the spinal defect. The creation of an artificial type of Chiari I deformity by diversion of CSF from the spinal region indicates that under these circumstances a similar difference in pressure between the cranial and spinal compartments is the force responsible for the herniation, a suggestion that was made earlier.

It might be expected that the difference in pressure would not be very great but could be intermittently increased by assumption of the erect position. In any event, under these conditions and because of the viscous properties of the brain, years are required for the deformity to develop and to become symptomatic.

It may not be intuitively obvious that the tonsils of the cerebellum might be stressed in a negative gradient of pressure and thus be strained even in the absence of impaction at the foramen magnum, but the tissue behaves as a visco-elastic medium as shown by Hakim and his co-workers, and is subject to deformation in response to stress. It is of some interest that in our Case I the cerebellar tonsils resumed a more normal position when the stress was removed.

Might the factitious form provide a paradigm for the spontaneous occurrence of Chiari I deformity? The principal ingredient for such a disorder, a spinal mechanism for absorption of CSF, is in place.

Under normal conditions both cephalic and spinal mechanisms are accessible for absorption of CSF. The latter was first suggested by Elman, who found arachnoid proliferations associated with spinal nerve roots of dogs, structures he considered to be spinal arachnoid villi. These accumulations of cells were attached to and penetrated the root sleeves. Elman repeated the Weed Prussian blue experiment and found the blue precipitate in the clusters of arachnoidal cells, in the tissue around the roots, and in some epidural veins. The possibility that epidural veins might be involved in absorption of fluid led Wislocki to study the spinal veins by postmortem injections. The intradural veins drained into dural veins, which presented sinusoidal dilatations before emptying into epidural vessels. Wislocki suggested that the dural venous sinusoids might be functionally comparable to the cephalic dural venous sinuses in terms of CSF drainage. The close relationship of spinal arachnoid villi to these venous channels in rabbits was later shown by Wollam and Miller, a relationship subsequently confirmed by Welch and Pollay in monkeys.

Several studies involving the subarachnoid instillation and later determination of the distribution of particulate matter suggested the existence of spinal pathways, but it was the perfusion studied by Coben and Smith, Hammerstad, et al., and Lorenzo, et al., that firmly established that there exists a capacity for absorption of CSF in the spinal region. Thus, the maintenance of function in the spinal pathway with impairment of the cephalic mechanisms might be comparable to the situation after spinal shunting, and it is suggested that this unphysiological state might account for the spontaneous occurrence of a Chiari I deformity.

There is a precedent for the recruitment of spinal absorptive mechanisms in compensation for hydrocephalus. The kind that follows external instillation of kaolin in cats is compensated by the dilatation of the central canal of the spinal cord as a collateral pathway to spinal absorptive sites.
The ideas presented here are in agreement with those of Gardner, that syringomyelia in association with the Chiari deformity is due to outlet obstruction of the fourth ventricle; however, they differ in that the obstruction is considered not to be primary but, instead, consecutive to the herniation and impaction of the hindbrain.

References


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