The relationship of Arnold-Chiari and Dandy-Walker malformations

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The posterior fossa is abnormally small in cases of Arnold-Chiari malformation because the tentorium is too low, whereas it is abnormally large in the Dandy-Walker malformation because the tentorium is too high.

KEY WORDS Arnold-Chiari malformation . Dandy-Walker malformation . hydrocephalus . basilar impression

In 1959 it was pointed out that, despite the difference in size of the posterior fossae, the Arnold-Chiari and the Dandy-Walker malformations possess enough of the same anatomical features to suggest that they have a common origin. The shared features are hydrocephalus, either obstructive or communicating, compensated or uncompensated; hydromyelia; failure of the roof of the primitive fourth ventricle to perforate; and herniation of the medulla through the foramen magnum. It was suggested that both malformations result from elevated pressure in the embryonal neural tube (hydrocephalomyelia) caused by inadequate permeability of the rhombic roof. In the infant these hindbrain malformations are more severe, frequently accompanied by spina bifida or cranium bifidum, and the congenital hydrocephalus is more likely to be uncompensated. The present case in an adult is significant in that it demonstrates one more feature that may be shared by both malformations, namely, a large posterior fossa.

Case Report
This 46-year-old woman was first examined on November 22, 1968. She complained of intermittent double vision, first noted 18 months previously while lying down to watch television. Later it occurred when the patient descended stairs, or with close work that required looking downward, and became associated with a vertical oscillation of objects in her field of vision. For 4½ years she had experienced headaches beginning in the posterior nuchal region radiating to the vertex, brought on by coughing, straining, and sneezing. Her voice became hoarse and raspy and she tired quickly. Occasionally she experienced a feeling of numbness on the right side extending up the side of her neck to the earlobe. Her gait had been increasingly unsteady for the past year.

Examination. The patient had a short neck, and a cautious, wide-based gait; she swayed in the Romberg position but there was no dysmetria. The chief neuro-ophtalmologic finding was a striking nystagmus, the fast component being downward. The nystagmus was not only greater in downward gaze than in upward gaze, but was eccentric in that it was greater when gazing down and to the right than when gazing straight down, or down and to the left. Visual acuity and visual fields were normal, as were the optic discs. A diagnosis of basilar impression was made on the basis of the eye findings, and confirmed by lateral skull films which dem-
Sinuses by carotid angiography to demonstrate the size of the posterior fossa. The sinuses, instead of being low as anticipated, were abnormally high (Fig. 1), indicating that the posterior fossa was large rather than small.

Operation. On February 20, 1969, a craniovertebral decompression was performed under endotracheal anesthesia. With the patient in the sitting position and the head well flexed, the occipital bone and upper three cervical laminae were exposed through a midline incision. The posterior rim of the foramen magnum had been pushed inward, and there was lack of movement between it and the lamina of C-1. The laminae of the upper three cervical vertebrae were removed and the foramen magnum enlarged upward. Beneath the foramen magnum there was a deep transverse groove in the dura caused by this indentation at the base of the skull. The bony ridge overlying the occipital sinus was located at the left of the midline. The dura was opened by a vertical incision, and, instead of the anticipated Dandy-Walker malformation, a congenital hindbrain hernia was found with the cerebellar tonsils herniated to the level of the lamina of C-2 (Fig. 2). Below this hernia was a pronounced posterior buckling (telescoping) of the cervicomedullary junction, indicating a downward dislocation of the medulla (a feature of congenital hindbrain hernia first pictured in the comprehensive 1895 monograph of Chiari).

Only the second pair of cervical nerve roots were seen; these pursued a horizontal course to their foramina of exit. The impacted hernia moved sharply downward with each pulse systole and upward with its diastole. Above the exposed portion of the cerebellum, the lower end of a large, translucent cyst-like structure was disclosed, which appeared to have pushed the hindbrain downward and to the left. Its wall was clearly separate from the overlying arachnoid (Fig. 2). The aspirating needle revealed clear, colorless fluid with a protein content of 15 mg%, indicating a ventricular diverticulum rather than a loculated cyst. The exposed portion of the membrane was excised, and, despite its congenital origin as indicated by the displaced tentorium, it still resembled embryonic rhombic roof in that it was partly lined by ependyma. This cyst-like structure, there-
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fore, was believed to have resulted from a bulging diverticulum of the rostral portion of the embryonic rhombic roof. No effort was made to uncover the full rostral extent of the cyst-like structure. That its greater portion lay to the right of the midline was indicated by the lateral displacement of the cerebellum and by the asymmetry of the posterior fossa, demonstrated by the position of the bony ridge overlying the occipital sinus.

The cerebellar tonsils were separated, disclosing some small tufts of choroid plexus attached to the undersurface of the vermis. The floor of the fourth ventricle was elongated downward, extending well below the level of the foramen magnum. The opening of the central canal beneath the obex was funnel-shaped, (a constant finding in syringomyelia); this was plugged by a bit of muscle to lessen the possibility of subsequent development of symptoms of syringomyelia. A silver clip was attached for x-ray localization. Below the telescoped cervicomedullary junction, the cervical cord did not appear to be cystic. (Fluctuation is often difficult to demonstrate through a limited exposure and with the patient in the sitting position.)

At the level of the foramen magnum, the edges of the dural incision were sutured laterally to the muscle, creating a diamond-shaped dural opening which was then covered with a patch of dural substitute. (The purpose of this procedure was to enlarge the dural sac at this level and thus to assure release of the impacted hindbrain hernia as well as to permit the partly imprisoned ventricular fluid pulse wave to escape freely into the cisterna magna.) After closure, with the patient still in the operating chair, a lateral skull film disclosed a large collection of air in the cyst-like structure beneath the high tentorium (Fig. 3).

Postoperative Course. Recovery was satisfactory, and the patient was discharged on the 13th postoperative day. When examined 18 months later, she reported that the headaches were relieved and that, except where descend-
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Fig. 4. Rhombic roof at the 18 mm stage. Rostral to the choroid plexus is the transient area membranacea superior (AMS). This permeable membrane soon thickens to form the vermis with its anterior and posterior medullary vela, the adjoining choroid plexus becoming attached to the undersurface of the nodulus. The area membranacea inferior (AMI) (future foramen of Magendie) is caudal to the choroid plexus. The lower arrow indicates the area of the rhombic roof that bulges to form the usual Dandy-Walker malformation. The upper arrow indicates the presumed origin of the bulging in the present case, the anlage of the cerebellum being located between these arrows. (Reproduced from Weed, L. H., Development of cerebrospinal spaces in pig and man. Carnegie Inst Wash Publ, p. 225, 1917, with permission of the publisher.)

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Discussion

Since the ventricular diverticulum in the present case was located above rather than below the cerebellum, it obviously had originated rostral to the anlage of the cerebellum. The portion of the rhombic roof which progressively thickens to form the cerebellum is in Weed’s area membranacea superior (AMS), situated rostral to the choroid plexus; caudal to this plexus is Weed’s area membranacea inferior (AMI), which progressively attenuates to form the foramen of Magendie.\textsuperscript{19}

To explain the findings in this case, the literature concerning the embryologic origin of the Dandy-Walker malformation was reviewed, and publications by Bonnevie and co-authors\textsuperscript{2-5} were found most pertinent. Their studies on Dandy-Walker malformation showed without question that this malformation, hereditary in the house mouse, originates not in the AMI but in a bulging portion of the AMS that intervenes between the cerebellar anlage and its choroid plexus (Fig. 4). Brodal and Hauglie-Hanssen\textsuperscript{6} subsequently described the postmortem findings in two children with Dandy-Walker malformation. As in Bonnevie’s hydrocephalic mice, they found the underdeveloped cerebellar vermis was displaced rostrally and its stretched choroid plexus displaced caudally by an intervening bulging portion of the AMS. A bulging of the AMI would not have separated choroid plexus from vermis and hence could not explain the usual Dandy-Walker malformation. They concluded that atresia of the outlet foramina could not have been the essential factor in its production (Fig. 5).

In our case, as in the Dandy-Walker malformation, the tentorium had failed to migrate as far caudally as it should have (Fig. 6). The cause of this failure appeared to have been a bulging of that portion of the AMS rostral rather than caudal to the cerebellar anlage. In view of its rostral location, this bulging diverticulum might be described as an “anterior” Dandy-Walker malformation, in contrast to the usual posterior variety.
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Fig. 5. Postmortem photograph of Dandy-Walker malformation in a 7½-month-old infant after removal of the bulging dura and translucent rhombic roof. Cerebellar hemispheres and pale maldeveloped vermis are displaced upward. The choroid plexuses (arrows) are stretched laterally and displaced far caudally from their normal position on the vermis by the bulging AMS. (Photo reproduced from Weed, L. H., Development of cerebrospinal spaces in pig and man. Cleveland Clinic Quart. 1959:26, by permission of the publisher.)

Fig. 6. Drawings showing the normal migration of the straight and transverse sinuses. The choroid plexus of the fourth ventricle (arrow) develops first, but the anterior choroid plexus soon outgrows it. The greater Bering effect of the latter, together with rapid reproduction of neuroepithelium, is believed responsible for the normal encroachment of the forebrain on the posterior structures. (Redrawn from Padget, D. H., Development of cranial venous system in man from viewpoint of comparative anatomy. Carnegie Inst Wash Publ No. 611, pp. 79–140, 1957, by permission of the publisher.)
Conclusion

In the usual Dandy-Walker malformation, a bulging of Weed’s AMS separates cerebellum from its choroid plexus; therefore, we believe this malformation should be redefined since atresia of the outlets of the fourth ventricle is the result rather than the cause of the deformity. Our case lends support to the thesis that the Arnold-Chiari and Dandy-Walker malformations may arise from a similar embryologic defect, namely, a disparity between the rate of formation and the rate of egress of ventricular fluid from the neural tube. If the retained fluid causes a relative overexpansion of the primitive lateral ventricles, a small posterior fossa will result; if it causes overexpansion of the fourth ventricle, a large posterior fossa will result. In the former, with growing of the cerebellum, the AMI becomes impacted in the foramen magnum, so that its subsequent perforation may be prevented. In the latter, the abnormal expansion of the permeable AMS may render perforation of the AMI unnecessary. Since the AMS begins to function with the initial tufting of the fourth ventricle choroid plexus, this would appear to fix the embryologic age at which the Dandy-Walker malformation begins.

The combination of a high tentorium with congenital hindbrain hernia may not be extremely rare. More frequent recourse to sagittal sinus venography in adults with down-beating nystagmus, basilar impression, syringobulbia, or syringomyelia would help to clarify the matter.

References


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