Article *NIMHANS Journal*

Chiari Malfunctions in Childhood

Volume: 02 Issue: 01 January 1984 Page: 49-52

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Abstract

The eponym should read 'Chiari malformations'. The causes of these malformations are obscure. Perhaps different mechanisms operate in each of the four types. Whilst hydrocephalus and spina bifida are the common features, acute respiratory distress or autonomic disturbances may be the only presenting signs. Air studies and angiography were hitherto the mainstay of diagnosis. Some patients do well with CSF shunting but most need decompression of the medullospinal junction.

Key words -
Chiari malformation,
Hydrocephalus,
Spina bifida

A question of priority

Chiari, with his detailed descriptions of these malformations in 1881 and 1885 surely deserves to have. them named after him to the exclusion of Arnold who merely described one case of the second type in 1894 [1].

Causation

Opinions differ as to their causation. Hydrodynamic factors are championed by W James Gardner since 1950 [2], [3]. He holds the primary dialation of the lateral ventricles, downward displacement of the tentorium and marked reduction in the volume of the posterior fossa as factors responsible for downward displacement of cerebellum and medulla. He thus turned upside down Russell and Donald's contention [4] that the primary lesion was obstruction to the reflux of cerebrospinal fluid into the cranial cavity due to plugging of the foramen magnum by the Chiari malformation. Penfield and Coburn [5] suggested a downward pull by the tethered cord in the lumbosacral region as the causal factor. E Gardner and his colleagues [6] postulated a primary mesenchymal abnormality leading to the downward shift of the tentorium. This, as can be expected, was emphatically challenged by W J Gardner [3].

Masters [7] suggested that hydrocephalus caused the aqueduct stenosis associated with the Chiari malformations and was, in turn, due to fibrovascular occlusion of the basal subarachnoid space. In doing so, he ignored Milhorat et al's findings [8] of patent subarachnoid pathways in 14 of 20 cases with these malformations studied at autopsy. Milhorat et al [8] blamed the severity and duration of hydrocephalus as being responsible for the leptomeningeal abnormalities seen in the remaining six patients.

De Reuck and Thienpont [9] described a foetus (4-5 months old) with a Chiari type 3 malformation and concluded that a "defective closure of the roof-plate is the primary cause of this malformation". They noted that the fetus showed no hydrocephalus, proving that the latter was "only a secondary abnormality and not the primary cause of the dystrophic malformations of the posterior fossa".

Obviously, we are, ignorant of the exact mechanisms operating to produce this complex group of malformations. There may, indeed, be several different mechanisms operating in the different types of Chiari malformations. We have at least one report of three members of one family having the Chiari type 1 abnormality with hydromyelia [10] implicating a genetic factor.

Classification and points of radiological interest

Type I:

Elongated, cone shaped cerebellar tonsils and medial aspects of the posterior lobes of the cerebellum as well as an elongated caudal medulla (exclusive of the fourth ventricle) lie within the upper cervical vertebral canal. The herniae reduce or obliterate both intra and extracranial segments of the cisterna magna but the fourth ventricle and medulla are unchanged.

Type II:

Greater elongation and caudal displacement of cerebellum and brainstem such that the caudal part of the fourth ventricle is also in the cervical canal. The fourth ventricle is lowered and the inferiorly placed medulla appears swollen opposite the upper two vertebrae.

Type III:

The entire but diminutive cerebellum is distended over a cystic fourth ventricle and the cerebellum, ventriculocele and lowered brainstem lie caudal to the foramen magnum. The ventriculocele continues through a defect in the posterior arches of the upper cervical vertebrae to present as a cyst on the surface at the base of the skull.

Type IV:

Cerebellum and fourth ventricle lie entirely within the posterior fossa.

a) The inferior vermis is buckled inward and covered by a cystic expansion of the enlarged fourth ventricle resembling the Dandy Walker malformation. A supratentorial occipital encephalocele and caudally displaced tentorium complete the picture.
b) small cerebellar hemispheres are seen, their dentate nuclei lacking the normal convolutional pattern. The fourth ventricle is large.

Whilst the Chiari malformation in adults typically involves herniation of the cerebellar tonsils, in infancy herniation of the vermis is the rule [12].

Clinical Presentation

The infant or child with Chiari malformation usually presents with hydrocephalus and spina bifida. The manifestations seen in adults (pain in the neck, signs of cerebellar involvement and evidence of hydromyelia) are usually not present.

In many children with the Chiari malformation, there may be no spinal abnormality even on x-ray examination.

It is important to note that the patient may be brought in with acute symptoms of respiratory distress and autonomic dysfunction. Breathlessness may follow bilateral, partial or total paralysis of the abductors of the vocal cords [13], [14], [20]. The patient may also show evidence of cricopharyngeal achalasia in the form of pooling of the saliva, regurgitation, cyanotic spells arid aspiration pneumonia. A mistaken diagnosis of asthma has been made. Cine-oesophagography is diagnostic, showing persistent constriction of the cricopharyngeus whilst swallowing [16].

Failure to make a correct diagnosis and follow through with appropriate therapy may prove disastrous. Friede and Roesmann [11] report sudden collapse and death in seven children with Chiari malformations (None of them showed any abnormality of the spinal cord or spina bifida. One had fusion of five cervical vertebrae).

Diagnosis

Hydrocephalus in association with spina bifida at once raises the possibility of the Chiari malformation. During the tests to determine the cause of ventricular dilatation the Chiari malformation may become obvious. In some patients, 'aqueduct stenosis' may be the ventriculographic diagnosis, the contrast medium not entering the fourth ventricle at all. In such cases it may be necessary to resort to pneumoencephalography, paying special attention to the craniovertebral junction.

Inability to introduce air into the cisterna magna and the presence of soft tissue filling defects within the air are the most reliable indicators to the diagnosis. The fourth ventricle may not fill with air even during the PEG. In their figure 14, Fredy et al [11] show very well the thickening of the medullo-spinal junction on air myelography diagnostic of buckling.

Gabrielsen and colleagues, [17] in their review of angiographic findings in the presence of Chiari malformations, emphasise that caudal displacement of the cerebellar tonsils without other angiographic signs of an expanding intracranial lesion suggest a Chiari malformation. Demonstration of a low cephalic loop of the PICA (reflecting caudal displacement of the fourth ventricle) is diagnostic. On their superb sub-traction pictures, the anterior and lateral margins of the lower medulla can be clearly seen below the foramen magnum confirming the diagnosis. one of their pictures also shows the posterior kinking of the cervicomedullary junction.

Treatment

In infants and children, hydrocephalus is the dominant feature and most surgeons shunt the cerebrospinal fluid away. This suffices in some patients who remain asymptomatic after such surgery [18], [19].

Others suffer despite a functioning shunt. Occipito-cervical headaches develop along with signs of cervicomedullary disease. Episodic breathlessness, stridor, cyanotic spells and difficulty in swallowing lived but are ominous, Such patients should be treated with suboccipital craniectomy and excision of the posterior arches of the upper cervical vertebrae so as to decompress the lower medulla and upper cervical cord. The dura should be opened and attempts made at separating the cerebellar herniae. The fourth ventricle should be inspected. If hydromyelia is seen, the opening of the central canal at the obex must be plugged. Some surgeons excise the cerebellar herniae and suture the tonsillar arachnoid to the dura to keep the fourth ventricle in communication with the subarachnoid spaces. A dural graft must be used to prevent leakage of cerebrospinal fluid. During surgery a close watch must be kept for cardivascular instability in the form of fluctuations of blood pressure and bradycardia [20]. Failure to carry out prompt surgery may prove fatal.

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