ON THE SYNDROME OF ARNOLD-CHIARI

REFERENCES

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REPORT OF A CASE

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(Received for publication January 10, 1948)

A malformation of the metencephalon, associated with herniation of the spinal cord and of its meninges through a large lumbosacral spina bifida, was first described by Arnold3 in 1894. Two years later, Chiari4 published a description of a still more serious developmental anomaly consisting of transposition not only of the abnormal cerebellum, but also of the medulla oblongata into the cervical portion of the vertebral canal. Schwalbe and Gredig (1907)17 added 4 similar cases and pointed out that, as a rule, both types of malformation, i.e. of the cerebellum and of the medulla oblongata, appear simultaneously, side by side. They emphasized, however, that this is not always so, and that one may be present without the other. Schwalbe and Gredig subdivided all such cases of malformation of the met- and myelencephalon into several groups, according to the degree of the anomaly, which in extreme instances may consist of displacement of the cerebellar substance through the central canal to the lumbar part of the spinal cord. On the other hand, it may be characterized only by a slight elongation of the vermis cerebelli of an otherwise normal metencephalon. Generally speaking, the difference between the malformation described by Arnold and by Chiari is only that of the degree of the pathological changes that occurred in the hind-brain during embryological life; therefore, it is not surprising that between the extreme cases many intermediary forms are found. This variability in the severity of the anomaly of the hind-brain is due to the difference in the length of time during which the maldevelopment has existed, the severe forms being the result of earlier changes.

Associated with the malformation of the met- or myelencephalon, or with both, is a spinal dysraphism, also differing, from case to case, in its dimensions. Although most frequently found in the lumbosacral region, it can be located in other portions of the spinal column. The disturbance in the development of the vertebral column leading to the absence of fusion of the vertebral laminae, occurs—according to Schwalbe and Gredig17—either at the same time as the disturbance in the development of the hind-brain or even earlier. They admitted that the relationship between the two anomalies may be that of a cause to an effect, the spina bifida causing, mechanically, a maldevelopment of the hind-brain. However, it may happen that both anomalies develop independently, at the same time, and due to the same unknown factor.

The spina bifida is frequently complicated by a myelomeningocele. Yet, Schwalbe and Gredig17 emphasized that the malformation of the hind-brain may be encountered also in the presence of a less pronounced form of spinal and meningeal herniation. In fact, Russell and Donald15 observed a case of malformation of Arnold and Chiari associated with a meningocele only, and it was clear to them that, in such cases, the deformity of the hind-brain would be of a far less severe order than that seen in cases of myelomeningocele, although it would be of a similar kind. A patient of Penfield and Coburn14 suffered from the malformation of Arnold and Chiari associated only with an upper thoracic meningocele. No evidence of spina bifida was seen in the case of Aring2 or in Case I of Adams, Schatzki and Scoville.1
Penfield and Coburn, reporting their observation of an Arnold-Chiari malformation in an adult presenting symptoms that were taken as indicative of a bilateral tumor of the acoustic nerve, induced the study of the clinical side of this condition. Up to this time, it had been considered a pathological curiosity. Although Houweninge Graftdijk3 was the first who attempted to treat the hydrocephalus surgically following the obliteration of the cisterna magna by the displaced cerebellum and the medulla oblongata, it was the report of Penfield and Coburn that drew attention to the desirability of limiting the treatment of the malformation of Arnold and Chiari solely to the decompression of the nervous structures involved. Indeed, Schwalbe and Greig knew that hydrocephalus may often be absent in these cases, especially when the transposition of the hind-brain is less pronounced. After Aring (1938)2, Adams, Schatzki and Scoville (1941)1 and Lichtenstein (1942)10 had published their observations the symptom complex, occurring in cases of the malformation of Arnold and Chiari associated with spinal dysraphism, became rather well-known. It consists, according to Naffziger and Boldrey,12 of symptoms resulting from increased intracranial pressure, symptoms indicative of compression of the brain stem or spinal cord, of cerebellar disturbance, and of symptoms due to irritation or paralysis of cranial and spinal nerves. The demonstration of the subarachnoid block in the cervical region helps to make the diagnosis.

An identical clinical syndrome may be found, as pointed out by Ebenius,7 in so-called platybasia. Up to 1946, some 30 clinical and 40 anatomo-pathological observations of this condition were reported (Welander19). The platybasia, a congenital malformation of the base of the skull, consists of the dorsal arching of the basiocciput and basisphenoid with flattening of the sphenoid angle. The result of this is a projection of the atlas and of the odontoid process above a line drawn from the posterior edge of the hard palate to the posterior border of the foramen magnum (Schüller28). This malformation is sometimes associated with such anomalies of the spinal column as Klippel-Feil's syndrome (Merio and Risak29), cervical spina bifida (Gustafson and Oldberg;3 List;11 Naffziger and Boldrey12), et cetera, all of which are the result of congenital malformation of the various anlagen of those structures. The clinical signs and symptoms seen in platybasia are due to the malformation and displacement of the hind-brain, and are similar to those found in connection with spinal dysraphism. Platynas was remarkably often associated with syringomyelia.

We see, then, a malformation of the hind-brain, sometimes associated with congenital malformation of the vertebral column, and at other times with congenital malformation of the base of the skull, especially when the last is coupled with bony anomalies of the craniovertebral junction. In both instances, there is a similar clinical syndrome, resulting from malformation and displacement of the hind-brain, although it must be admitted that the favorable results of decompression of the cerebellum, the medulla oblongata and the spinal cord with their respective nerves (Aring;2 Chamberlain;2 Gustafson and Oldberg;8 List;11 Adams, Schatzki and Scoville;2 Bucy and Lichtenstein5) seem to indicate that the symptom complex is produced most often by compression of the nervous structures rather than by intrinsic changes occurring in them.

A quite unique observation was reported by Bucy and Lichtenstein.4 They found a displacement of the cerebellum in a woman, aged 40 years, without evidence of any unusual anchorage of the spinal cord or of any bony anomaly. The authors believe that, even under normal conditions of development, the hind-brain is subjected to a force that tries to draw it into the vertebral canal. This comes from the difference in the rate of growth of the vertebral column and that of the spinal cord.

The malformation of Arnold and Chiari and the platybasia are becoming, of late, popular with neurologists and neurosurgeons. It seems, however, that we are still in the stage of collecting data necessary for a fuller understanding of those pathological conditions. In that fact, we find a justification for reporting here another observation of malformation and displacement of the cerebellum associated with platybasia and spina bifida occulta of the atlas. As in other similar cases, the patient was markedly relieved by decompression of the nervous structures of the craniovertebral junction.