Prenatal Agenesis of One Cerebral Hemisphere with Displacement of the Sagittal Sinus

Case Report

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The clinical syndrome of hemiplegia associated with diffuse atrophy or malformation of the contralateral cerebral hemisphere is well known. It has been our privilege to study angiographically a case that demonstrated massive lateral displacement of the sagittal sinus. Review of the literature failed to reveal a previous radiographic or pathological report of this anomaly.

Case Report

This 16-month-old white girl was the product of a 31-week pregnancy which had terminated in a breech delivery. The infant was reported to be cyanotic for about 15 minutes following birth. The weight at birth was 2 lbs 12 oz. The child was apparently developing normally until the age of 3 months when the parents noted a lack of normal movements in the left arm and leg. This became more pronounced until medical attention was sought at the age of 6 months. The child could sit without support at 7½ months and crawled at 10 months. At 16 months she was able to walk only when supported. Speech was limited to early word formation. She had not experienced any convulsive episodes. There was no history of trauma nor significant infection since birth.

The mother was 29 years of age, the father 34. There had been two previous normal pregnancies and deliveries resulting in normal siblings now aged 9 and 7 years. There had been no family history of neuromuscular disorders.

Examination. The patient was small for her age and had an obvious left hemiparesis. She responded appropriately to the examiner by playing and laughing. She generally appeared quite alert. The weight was 17 lbs, height 74 cm, and head circumference 46 cm. The head and face seemed symmetrical. The fontanels were closed, and the cranial sutures were not prominent to palpation.

Neurological examination revealed a left hemianopsia but no pupillary or extraocular muscle abnormalities. There was a left hemiparesis with spastic posturing in the hand and less severe involvement of the left leg. Less response to painful stimulus was noted over the left extremities. The tendon reflexes were increased on the left and normal on the right.

Plain skull x-ray films demonstrated symmetry of the skull and normal cranial sutures. Pneumoencephalography showed filling of only one lateral ventricle, which was minimally enlarged but located almost in the midline. The configuration of the ventricular shadow led to the conclusion of a displaced left lateral ventricle. There was also a relatively large accumulation of convexity air in the right temporoparietal area, which outlined the small, atrophic right cerebral hemisphere (Fig. 1 left).

Bilateral carotid arteriograms revealed marked displacement of the anterior cerebral arteries from left to right (Fig. 1 right). In addition, the sagittal sinus was not located in its usual position beneath the sagittal suture but was displaced far to the right (Fig. 2). The right carotid injection also revealed abnormalities of the middle cerebral vasculature (Fig. 3) similar to those previously reported.

Discussion

There is no scarcity of angiographic data on this condition. Lefèbre, et al., analyzed with arteriography 30 such children. Dyken performed arteriography on 40 patients with infantile hemiplegia including several adults. Ford and Schaffer, in perhaps the definitive etiological study, presented pathological material on 38 patients.

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Agenesis of Cerebral Hemisphere

Fig. 1. Left: Composite drawing emphasizing findings of pneumoencephalogram and angiogram. Note the midline sagittal suture despite the extreme lateral position of the sagittal sinus and falx. Right: Left carotid injection revealing abnormal course of the anterior cerebral arteries. The sagittal suture can be identified in the midline.

The etiology of this disorder has been vascular in an overwhelming majority of patients, in the form of occlusion or abnormal formation of the middle cerebral or carotid arteries on the involved side. The occlusion may occur as a thrombotic or embolic phenomenon.

Despite several angiographic series in the literature, very little mention is made of venous abnormalities. Abnormal veins from the atrophic brain substance have been noted, but these continue to drain via the usual collecting sinuses.

Postmortem material often has revealed findings of massive atrophy with varying degrees of shift from the normal to abnormal side. This may have involved the anterior cerebral arteries and internal cerebral veins but rarely to the degree seen here. No cases have been noted of displacement of the dural sinuses or falx.

This displacement of the sagittal sinus and

Fig. 2. Angiograms, anteroposterior (left) and lateral (right) views of left carotid injection outlining the sagittal and both transverse sinuses. Residual air from previous pneumoencephalography is present.
Fig. 3. Lateral view of right carotid injection outlining the middle cerebral group supplying the atrophic right hemisphere.

torcular Herophili from the midline leads to interesting speculation. The most likely period for occlusion of the cerebral vasculature has been shown to be during delivery and its incident trauma, and during perinatal infections. Indeed, in prematurity as manifested by this patient, such complications are more frequent. At this stage, however, the falx and dural sinuses are well formed, and any degree of displacement of these would be highly unlikely.

Mall and Streeter, in classic work on embryogenesis of human cerebral veins and dural sinuses, have found that the superior sagittal sinus develops in the following manner. In the 21 mm embryo (8 weeks) there is beginning development of a plexus of small channels in the midline area, formed by the fusion of veins from both hemispheres. The falx is not yet developed at that stage. At the 50–52 mm stage (12 weeks), the sagittal sinus is fairly well developed as a single midline channel. The falx has also formed at this time. The torcular continues to be displaced caudally by the enlarging cerebral hemisphere until it reaches the adult position.

As the distinct arterial groups are formed prior to the 50 mm embryonic stage, impairment of a major portion of the blood supply to one hemisphere at this stage could easily lead to migration of the falx and with it the sagittal sinus, in response to a unilateral enlarging cerebral hemisphere. Early occlusion may also have contributed to this patient’s relatively normal cerebral function.

This patient reveals remarkably good mental and physical development, considering the magnitude of her cerebral anomaly. Moreover, she had had no documented seizures, a symptom that becomes a problem in more than 50% of patients with infantile hemiplegia. At present no further diagnostic or surgical procedures are planned.

**Summary**

A case of chronic infantile hemiplegia with cerebral hemiatrophy and marked lateral displacement of the sagittal sinus and falx has been reported, and the embryogenesis of the dural sinuses discussed briefly in an effort to clarify the development of such an unusual configuration.

**References**

7. LINDENBERG, R. Personal communication.