

Unilateral Hydrocephalus Combined with Occlusion of the Ipsilateral Internal Carotid Artery

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The authors encountered a rare case of a patient who developed a mild hemiparesis on the right side at age 8 months with marked enlargement of the left lateral ventricle and occlusion of the ipsilateral internal carotid artery. In computed tomography scans and nuclear magnetic resonance scans 8 years after an operation to install a ventricular shunt, there was marked enlargement of the left lateral ventricle and almost no left basal ganglion. In this case, the differentiation between hydranencephaly and severe hydrocephalus presented a problem because no biopsy had been performed. Diagnosis was difficult, but it appeared that occlusion of the ipsilateral internal carotid artery played a major role.

KEY WORDS: Unilateral hydrocephalus; Hydranencephaly; Internal carotid artery, occlusion; Computed tomography scanning; Nuclear magnetic resonance scanning; Angiography

Unilateral hydrocephalus is a comparatively rare disorder, and various theories have been advanced concerning its cause. We encountered a rare case of unilateral hydrocephalus with occlusion of the ipsilateral internal carotid artery and marked enlargement of the left lateral ventricle. This case is reported here, together with some discussion of the literature.

Case Report

The patient was an infant girl, 1 year 8 months old, with nothing remarkable in her family history. There were no abnormalities during pregnancy or delivery. At the time of delivery, her head showed a normal measurement of 32.5 cm. From about 8 months of age, she developed a mild hemiparesis on the right side, and a

physician was consulted. During the examination, it was noted that the electroencephalograms of the left hemisphere were flat; she was admitted to our hospital.

On admission, the head measurement was 48 cm, the upper limit of the normal range, but there was bulging on the left side of the head. Neurologically, there was a mild hemiparesis on the right side, mild left optic atrophy, and delayed mental development (DQ: 55). In a plain radiograph of the head, there was asymmetry of the cranium, bulging on the left side of the head, and downward displacement of the pyramid (Figure 1). Angiography of the left carotid artery showed occlusion of the supraclinoid portion, no peripheral image, and transdural anastomosis from the middle meningeal artery (Figure 2). An angiogram of the right carotid artery showed no blood flow across the anterior communicating artery to the left cerebral hemisphere could be detected; however a few very small feeders from the right anterior cerebral artery to the left medial frontal region could be seen (Figure 3). An angiogram of the right vertebral artery showed poor development of the left posterior cerebral artery: there was no image of the posterior communicating artery, and no blood perfusion by collateral routes was visible. In a pneumoencephalogram, the left hemisphere showed as a very thin mantle (except in part of the frontal and occipital regions) and marked enlargement of the lateral ventricle could also be seen, but no morphologic abnormalities in the right lateral ventricle were visible.

When a small triangular craniotomy was performed on a trial basis in the left frontal region, the coloring of the frontal region at the site of the craniotomy was normal, but the gyri were not clearly seen and vascularization was poor. These findings extended to the lateral and frontal regions. When a ventricular puncture was performed, the thickness of the frontal mantle was found to be about 1 cm, and a ventricular shunt operation was performed.

After the operation the head measurement temporarily decreased by 1 cm, but it returned to the preoperative size within 2 weeks. Neurological symptoms were comparatively mild, and there was only a mild

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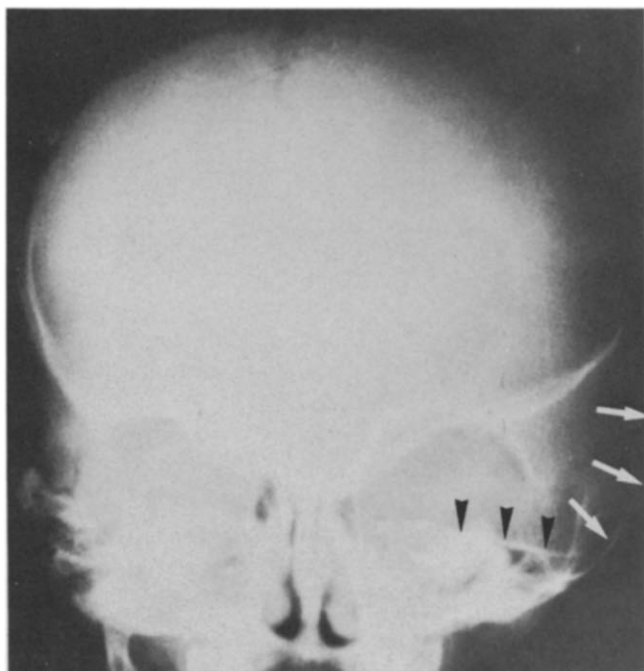


Figure 1. Skull X-P. Asymmetry of the cranium; outward bulging of the left temporal bone and downward displacement of the left petrous bone.

hemiparesis on the right side and a decrease in visual acuity on the left side. After being discharged from the hospital, she was enrolled in a special school. In a subsequent computed tomography scan performed 8 years after the operation there was a marked enlargement of the left lateral ventricle and almost no basal ganglion (Figure 4). The electroencephalogram of the left side

was almost completely flat except for the frontal and occipital regions. Nuclear magnetic resonance scanning showed thicker mantle at the left frontal region than computed tomography scanning (Figure 5).

Discussion

The most widely held theories concerning the cause of unilateral hydrocephalus are (a) that the interventricular foramen has been closed for some reason such as a tumor, infection, trauma or congenital defect, or (b) that the circulation of cerebrospinal fluid has been disturbed by septal formation in the lateral ventricle [1,5].

In the present case, the complete ventricular system was delineated by the pneumoencephalogram, and tests indicated that there were no disturbances in the passage of the cerebrospinal fluid through the interventricular foramen. In the subsequent computed tomography scans performed after the shunt operation, there was still marked unilateral hydrocephalus, and subsequent electroencephalograms also showed almost no activity on the diseased side. From these findings, it appears that this was not due only to a disturbance in the passage of cerebrospinal fluid through the interventricular foramen, but also that occlusion of the internal carotid artery during fetal development was a more important factor.

Clinically, there have been scattered cases of atrophy of the cerebral hemispheres caused by unilateral hypoplasia or occlusion of the internal carotid artery [6,9]. According to a report of Becker [2], various degrees of unilateral hydranencephaly occurred when paraffin was



Figure 2. Left carotid angiogram (lateral view). The left internal carotid artery can be identified up to the clinoid portion (arrow), above which, however, no carotid arterial system is visible. Transdural anastomosis from middle meningeal artery can be identified, too (arrowhead).

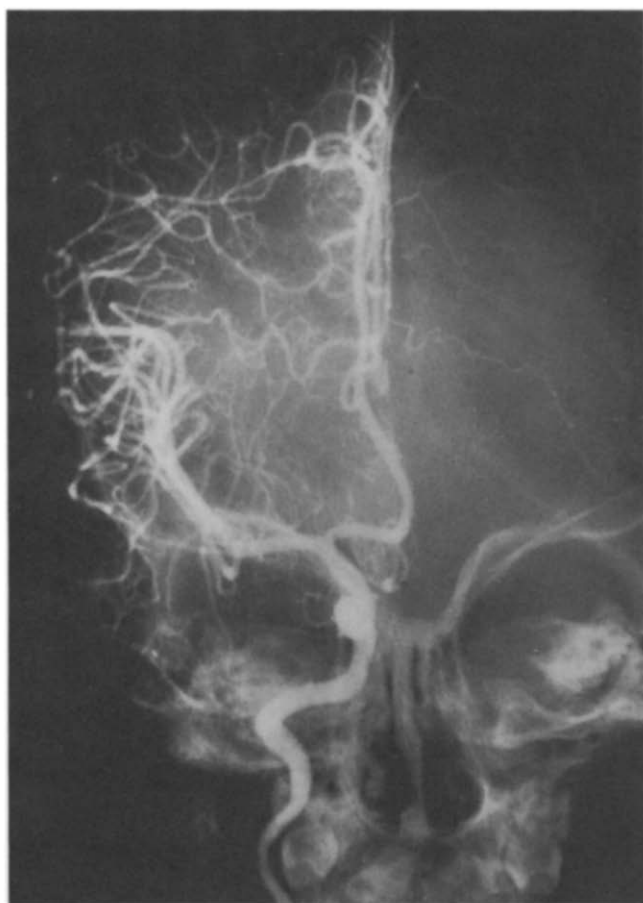


Figure 3. Right carotid angiogram (anteroposterior view). Flow across the anterior communicating artery to the left side cannot be detected. However, a few tiny feeders from the right anterior cerebral artery to the left medial frontal region can be identified.

injected into the unilateral internal carotid artery of puppies, and it appeared that occlusion of the internal carotid artery was a very important factor in the present case.

A major theory concerning the cause of hydranencephaly is bilateral internal carotid artery occlusion (supraclinoid portion) 6 weeks after fetal formation [7,8,10], and several cases of unilateral hydranencephaly have also been reported. According to these reports, the vertebral arterial system was normal, which indicates that part of the basal ganglion or the occipital lobe was present in many cases. The lack of a peripheral image from the supraclinoid portion in the present case is in agreement with this theory, but there were several points that differed from hydranencephaly—such as the lack of any clear structure at the site of the basal ganglion.

Crome and Sylvester [3] has reported on the histopathological differences between hydranencephaly and severe hydrocephalus. They stated that in hydranencephaly there is residual glial tissue corresponding to the

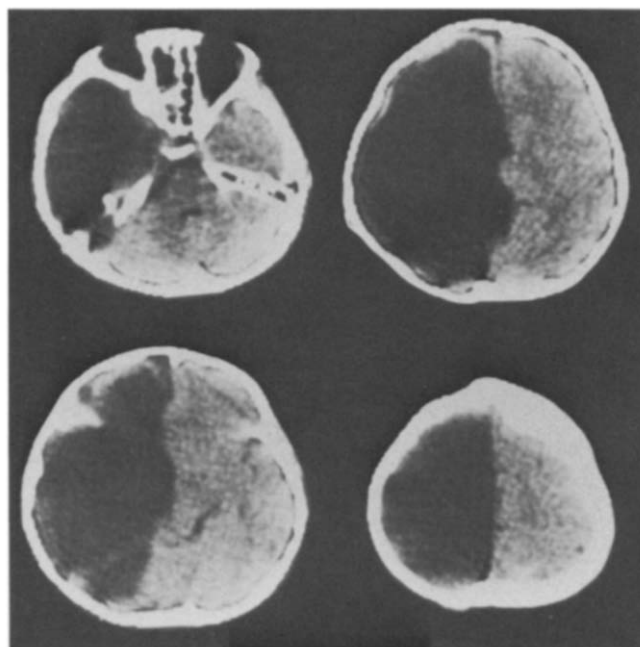
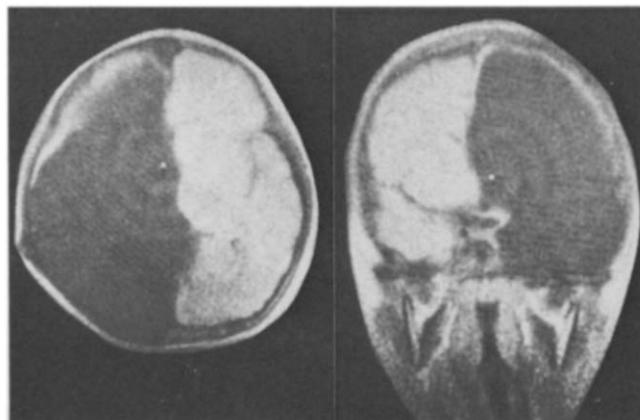


Figure 4. Computed tomography scan. Marked enlargement of the left lateral ventricle and very thin mantle at the left frontal region are seen. The basal ganglia, especially on the left side, are not so clear.

molecular layer of the normal gray matter and leptomeninges, but there is not ependyma. In severe hydrocephalus, however, there are residues of all layers of gray matter, subcortical white matter, and ependyma. Unfortunately, no biopsy was performed in the present case, and it was impossible to differentiate between the two diseases. In any case, we feel that the unilateral hydrocephalus in this patient is a form of unilateral hydranencephaly due to fetal occlusion of the internal carotid artery.

Figure 5. Nuclear magnetic resonance scan (CPMG method). A thicker mantle at the left frontal region is seen here than on the computed tomography scan.



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