HYDRANENCEPHALY

— CLINICAL AND NEURORADIOLOGICAL ASPECTS —

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Received for publication April 13, 1978

The authors encountered a case in which the diagnosis of hydranencephaly was done by CT scanning and cerebral angiography. This paper deals with the clinical feature, characteristics revealed by CT scanning, and cerebral angiographical findings of hydranencephaly.

INTRODUCTION

The term hydranencephaly, or "an-encephalie hydrocephalique" was proposed by Cruveilhier (1835) for the first time. The anomaly consists primarily of absence of the cerebral hemisphere within intact meninges and skull, the resulting cavity being filled with cerebrospinal fluid (Hamby et al, 1950 ; Crome, 1972). Such extensive cerebral deficiency as observed in hydranencephaly involved the whole cerebral hemisphere, but the basal ganglia, brain stem, and cerebellum usually remain in the normal state. The recent development of CT scanning has made it relatively easy to make a diagnosis of hydranencephaly. A more accurate diagnosis will be made if this technique is carried out simultaneously with cerebral angiography.

CASE PRESENTATION

The case presented concerned a 5-month-old girl born on January 30, 1976. The family history, pregnancy and delivery were not remarkable. Nothing particularly abnormal was found at birth, when she weighed 3,400 gr. She presented, however, a weak cry and a feeble power to suck. Moreover, her body temperature was so low that constant warming was required for her. She began to make a grimace 2 days after birth. This attack appeared frequently after that. On June 9, 1977, the patient was admitted to the Department of Neurosurgery, Kurume University Hospital.

Physical examinations revealed a well nourished, irritable baby with moderate cranial enlargement. Head circumference was 42 cm ; the anterior fontanelle

measured 3×3 cm and was showing a normal tension. In neurological examinations, the bilateral optic nerves were markedly atrophic. The pupil presented a moderate mydriasis and was lacking in both direct and indirect light reflex. Both eyes were affected with the incoordination of ocurogyration and nystagmoid jerk. There was no dyskinesia of any extremity. The lumbar puncture was performed; the cerebrospinal pressure was unknown, and was observed clear colorless fluid containing a few red blood cells and a normal total protein content. In electroencephalography, almost flat waves were presented by bilateral frontal, central, and temporal lead, and irregular slow waves by occipital lead (Fig. 1).

The CT scanning picture of the present case is shown in Fig. 2-a and b. As is clear from this figure, the cerebral parenchyma is seen partially in the frontal and temporal base. The ventricular system is lacking. In the posterior fossa, however, the cerebellum retains its normal morphology almost completely. The 4th. ventricle is visible (Fig. 2-a). In Fig. 2-b the cerebral parenchyma is noticed indistinctly at the center, and the occipital lobe retains its normal form relatively perfectly (Fig. 2-b).

Fig. 3-a presents the angiogram of the right carotid artery. In it the internal carotid artery on the opposite side is exhibited through the anterior communicating artery. Although this

Fig. 1 See text

100
μV
1 sec.

artery is presented all the trunks, except one of the anterior cerecal artery failed to be shown. Moreover, the trunk of the middle cerebral artery is expressed distinctly but neither secondary nor tertiary branch of this artery is. It should be noted that a large abnormal branch ($\curvearrowright$) is given off from the

horizontal portion of the middle cerebral artery. Furthermore, the distal portion is extended to from a capillary branch in Heuber's artery and the lenticulostriate arteries (Fig. 3-a). Fig. 3-b is a lateral view. In it, the vascular architectoric is very simple because of the secondary and tertiary arteries of

Fig. 2-a See text

Fig. 2-b See text

Fig. 3-a See text

Fig. 3-b See text

Fig. 4-a See text

Fig. 4-b See text

every artery forming a trunk. Besides, the trunk of the middle cerebral artery ($\rightarrow$) extends and ascends. An abnormal branch ($\Leftarrow$) arising the horizontal portion is assumed to run along the ten-

torium. These arteries are considered to form anastomoses with the external carotid artery in the leptomeningeal circulation in the periphery (Fig. 3-b).

In Fig. 4-a and b are shown the antero-posterior and lateral views of the left carotid artery in angiography. A simple vascular structure is presented on account of the absence of the secondary and tertiary arteries of the anterior and middle cerebral arteries (Fig. 4-a, b). Fig. 5 exhibits the venous phase, and it shows that the deep venous systems are not indicated (Fig. 5).

Fig. 5 See text

The results of angiography of the left vertebral artery are shown in Fig. 6-a and b. The posterior cerebral, superior cerebellar, and posterior inferior cerebellar arteries show normal branching and take a normal course of running on both sides. The distance is shortened, however, in the ambient segment between the left and right superior cerebellar arteries, probably because of stenosis of the aqueduct of sylvius (Fig. 6-a, b). Fig. 7 presents the antero-

Fig. 6-a See text

Fig. 6-b See text

posterior view of the venous phase. In it, the superior sagittal, bilateral sigmoid sinuses of the internal jugular vein are expressed distinctly as shadows (Fig. 7).

Fig. 7 See text

DISCUSSION

In the case of hydranencephaly an extensive cerebral defect is seen in the frontal, parietal, temporal, and occipital lobes. Normal conditions, however, are usually maintained in the basal ganglia, brain stem, and cerebellum (Mori and Handa, 1976). Lange-Cosack (1944) classified hydranencephaly into two types, 1 and 2. In type 1 such part of the cerebrum as above the midbrain is completely deficient. In type 2 basal ganglia, diencephalon, and cerebral mantle remain partially. The case presented by the authors belong to type 2 of his classification.

The frequency of occurrence of hydranencephaly is about 0.2 per cent among newborn infants. In severe cases stillbirth occurs and sucklings die early. There is no agreement of opinions on the etiology of hydranencephaly. A predominant opinion is that this disorder may be induced by the ischemic change of the cerebrum which is caused

by the compression of the internal carotid artery as a result of tangling of the umbilical cord and adhesion of the amnion (Lange-Cosack, 1944). In general, the changes of hydranencephaly are severe in an area supplied by the anterior and middle cerebral arteries and relatively mild in an area supplied by the posterior cerebral artery. Therefore, the opinion mentioned above are supported by many investigators (Sakae et al., 1970). On the other hand, Lindenberg and Swanson (1967) studied five infants suffering from severe encephalomalacia with membranous changes. Since the distribution of pathological changes of this disease was identical with that of hydranencephaly, they proposed the term infantile hydranencephaly for this disease. They stressed the significance of circulatory disturbances in the etiology of hydranencephaly.

The neurological characteristic of hydranencephaly consists in the fact that neonatal reflexes are seen for several weeks after birth and followed by failure of development of mental activity. The clinical signs of this disease frequently observed are convulsive seizure, dysphagia, hypothermia, nystagmus, ocular incoordination, and strabismus. They were also visible in the case presented by the authors. Optical blink reflexes and responses to auditory and visual stimulation are recognized in some suckling infants who have no ability to follow any subject by the eye as yet. This finding has been considered to suggest that the reflex tract may be present in the subcortex in suckling infants.

Poser et al. (1955) and Harwood-Nash and Fitz (1976) put emphasis on obstruction or stenosis of the internal carotid artery, non-development of the

anterior cerebral artery, and abnormality in the course of running of the posterior cerebral artery among the cerebral angiographical findings of hydranencephaly. Thelander and Shaw (1953) mentioned, however, that they had found no abnormality serious enough to be a cause of hydranencephaly. In brief, opinion is divided on the etiology of this disorder. By the way, in the case presented by the authors no stenosis was seen in the internal carotid artery, and the main trunks of the anterior and middle cerebral arteries were presented distinctly, but they had neither secondary nor tertiary artery.

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