CT FINDINGS IN AN ALOBAR HOLOPROSENCEPHALY ASSOCIATED WITH DANDY-WALKER'S CYST

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The authors describe a case of alobar holoprosencephaly with a huge cyst in the posterior fossa, possibly representing a Dandy-Walker's cyst, and marked cerebellar hypoplasia in the absence of midline facial dysmorphias that has recently been encountered at our clinic.

INTRODUCTION

Holoprosencephaly is a malformation of the brain due to a disturbance of the growth of the prosencephalon during the third cerebral vesicle phase of early fetal life. It is an anomaly of the prosencephalon with a single ventricle associated with malformation of the telencephalon and diencephalon. According to the degree of the malformation, this anomaly is classified into three types (DeMyer, W., Zeman, W. et al, 1964; DeMyer W., 1971.): 1) alobar holoprosencephaly in which there is no formation of the cerebral hemispheres, 2) semilobar holoprosencephaly in which the formation is partial, and 3) lobar holoprosencephaly in which the formation is complete. In this paper, the authors describe neuro-radiological features of the alobar holoprosencephaly with possible association of Dandy-Walker's cyst that has recently been encountered.

CASE PRESENTATION

The patient, a full-term boy born

by caesarean section, weighing 3,950 gm, and having an abnormally large head circumference of 42 cm, was admitted to our clinic at the age of 4 months with a diagnosis of congenital hydrocephalus due to a progressive increase in head circumference. At admission, physical examination revealed a normally growing and developing infant, 65.5 cm in height and weighing 7,000 gm, but with marked enlargement of the head (60.5 cm). There was no evidence of gross abnormality in the extremities or the vertebral column, and the patient was capable of active movement of his limbs.

Neuroradiological Findings

Skull plain films revealed a cranio-facial dysproportion due to extreme macrocrania. There was no evidence of hypotelorism or midline cleft in maxilla. Retrograde cerebral angiogram was performed via the right brachial artery. Arteriogram by antero-posterior projection demonstrated a slanting from the midline of the "median trunk" that corresponded to the anterior cerebral

artery, while the "lateral trunk" corresponding to the right middle cerebral artery appeared small indiameter with few furcations. Arteriogram by lateral projection demonstrated well-formed syphons of the internal carotid arteries. Intracranially, the anterior cerebral artery was observed to be in close proximal contact with the inner table of the anterior skull base. Phlebogram by lateral projection failed to visualize the anterior two-third of the superior sagittal sinus. Two cortical veins were observed to merge into a horizontal vessel with an antero-posterior course that continued into a transverse sinus that was situated in an extremely superior position. The diencephalic vein emptied directly into the transverse sinus without evidence of the vein of Galen. Neither the straight sinus nor the inferior sagittal sinus could be visualized. CT scans of the axial plane demonstrated a single ventricle that occupied the greater portion at the supratentorium and was surrounded by small amount of remnant brain tissue. Inferior to the tentorium, the cerebellum appeared markedly hypoplastic, the fourth ventricle was absent, and the brain parenchyma appeared to consist primarily of the brain stem. An extensive, low density area surrounding this brain parenchyma was noted. No cerebral falx could be demonstrated. CT scan of the coronal plane demonstrated a large single ventricle within the enlarged cranium with remnant brain tissue in the right inferior and median inferior regions (Fig. 1-a, b, c, d).

DISCUSSION

The arteriographic finding from a lateral projection of the median trunk in the present case suggests the an-

terior cerebral artery courses over the cerebral surface due to the absence of the longitudinal fissure. In addition, phlebogram has demonstrated the absence of the superior and inferior sagittal sinuses, the straight sinus and the internal cerebral veins, with an abnormally superior position of the transverse sinus and the failure of formation of the vein of Galen. The vein draining from the thalamus, the so-called fetal diencephalic vein, passed directly into the transverse sinus.

These findings are consistent with the morphological characteristics of the deep cerebral veins in the case of holoprosencephaly described by Osaka and Matsumoto (1978). In addition, CT scans revealed an alobar type of holoprosencephaly with complete absence of the longitudinal fissure and the cerebral falx, and a prominent single ventricle occupying the greater part of the supratentorium (Hayashi T. et al, 1979). In infratentorium, the fourth ventricle was absent, with small amount of cerebellar parenchyma. The infratentorial parenchyma evident appeared to be limited to the tissue of the brain stem. It is impossible to determine whether the large infratentorial cyst represents a Dandy-Walker's cyst or an arachnoid cyst, but the pressure of the developing cyst apparently resulted in an arrest in development of the cerebellum and oppression of the fourth ventricle and aqueduct with subsequent internal hydrocephalus. However, the frequent association of Dandy-Walker's cyst with agenesis of the corpus callosum has been reported (Harwood-Nash D. C. and Fitz C. R., 1976). To our knowledge, the case described by Osaka and Matsumoto (1978) has been the only reported case with Dandy-Walker's cyst that is associated with a single ventricle and anomalies of the deep cerebral veins of the brain

Fig. 1. CT in the axial (a, b, c) and coronal plane (d).

- a, d: A huge single ventricle and small amount of remnant brain tissue surrounding the low density area. A large cyst can be seen in the infratentorium.
- b, c: A large cyst around the hypoplastic cerebellum and brain stem in the posterior fossa.

comparable to that of the presently described case. Furthermore, according to Osaka and Matsumoto (1978), holoprosencephaly in the absence of median facial dysmorphias has been observed in as few as 4 of the 42 cases of the anomaly reported thus far in the

literature. If the presently reported case of alobar holoprosencephaly without median facial dysmorphias is, indeed, associated with Dandy-Walker's cyst, then it would be only the second reported case with this association of anomalies to have occurred.

REFERENCES

- DEMYER, W., ZEMAN, W. and PLAMER, C. (1964). Face predicts brain : diagnostic significance of median facial anomalies in holoprosencephaly (arhinencephaly). *Pediatrics*, **34**, 256-263.
- DEMYER, W. (1971). Classification of cerebral malformations. In "Birth Defects : Original Articles." National Foundation March of Dimes, Vol. VII, No. 1, February.
- HARWOOD-NASH, D. C. and FITZ, C. R. (1976). "Neuroradiology in Infants and Children." St. Louis, C. V. Mosby Co., pp. 1026-1030.
- HAYASHI, T., YOSHIDA, M., KURAMOTO, S., TAKEYA, S. and HASHIMOTO, T. (1979). Radiological features of holoprosencephaly. *Surg. Neurol.* **12**, 261-265.
- OSAKA, K. and MATSUMOTO, S. (1978). Holoprosencephaly in neurosurgical practice. *J. Neurosurg.* **48**, 787-803.