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ANTENATAL ULTRASONOGRAPHIC DIAGNOSIS OF FETAL ANOMALIES OF CENTRAL NERVOUS SYSTEM

(antenatal diagnosis / ultrasound / central nervous system)

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We present herein 10 cases with fetal anomalies of central nervous system (CNS), diagnosed antenatally by ultrasound, at Shimane Medical University Hospital. Five anencephalies, 4 hydrocephaluses and one encephalocele could be diagnosed in utero, but associated 3 sacral meningoceles were failed to detect antenatally. Eight cases were stillbirth and one case with hydrocephalus died due to heart anomaly on 142th neonatal day. But, last case with hydrocephalus was still alive under the pediatric management. We discussed the ultrasonographic findings of fetal anomalies of CNS and reviewed the previous papers.

With recent advances in ultrasonographic instruments, detailed studies on fetal central nervous system (CNS) in utero have been done (1) and the antenatal diagnosis of fetal anomalies of CNS was reported(2)-(4). In this paper, we present 10 cases of fetal anomalies of CNS complicated with various fetal malformations diagnosed antenatally by ultrasound at Shimane Medical University Hospital and reviewed the previous papers.

PATIENTS AND METHODS

The fetuses with anomalies of CNS, ranging from 27 to 39 weeks of gestation, were the subjects of this study.
Nine cases out of ten were refered after 27 weeks of gestation from another hospitals due to further examination of fetal

anomalies of CNS.

Antenatal ultrasonic examinations were performed in all cases, and all ultrasonograms were retrospectively assessed, comparing with the final diagnoses which were comfirmed after delivery.

The instruments used were Aloka SSD-256 (3.5MHz).

RESULTS

Ten cases of fetal anomalies of CNS were five anencephalies, four hydrocephaluses and one encephalocele. The gestational age of first detection, antenatal diagnosis and fetal outcome were summarized in Table 1.

The ultrasonographic findings of anencephaly in utero were the absence of fetal cranial vult but the existence of facial bone and the base of skull and the prominence of orbita (Fig.1). All were growth retarded fetuses and resulted in stillbirth. Hydramnions were noted in three cases by ultrasound.

All four cases of hydrocephaluses were detected antenatally by ultrasonographic measurement of the lateral ventricular ratio (LVR). In each case, LVR was greater than the normal range of LVR presented by Johnson and Dunne (1). The ultrasonographic findings of hydrocephalus were the extension of the body margin of lateral ventricle more than halfway to inner table of skull (Fig.2), fluctuation of the midline echo, thin mantle of cerebrum (Fig.3) and the shrunken small choroid plexus just medial to the margin of the ventricle (Fig.4). Three cases revealed the fetal brain mantle less than 1 cm in thickness, which were resulted in stillbirth. Hydramnions were detected ultrasonographically in one case, but the complicated sacral meningoceles were failed to detect in three cases.

In one case of encephalocele, the ultrasonographic findings were the large cystic mass with solid parts and a septum behind the fetal scalp (Fig.5).

DISCUSSION

A normal fetal head could be identified by ultrasonography as early as 12 weeks of gestation (5). Therefore, the absence of fetal cranial vult and the existence of small brain tissue at the end of cervical spine in this weeks was enough to suggest the

anencephaly (6). Johnson et al.(5) reported the case of fetal anencephaly which could be diagnosed at 11 weeks of gestation ultrasonographically, and Kurjak et al.(7) satated that there was no false-positive diagnosis after 13 weeks of gestation. In our experience, anencephalies could not be detected ultrasonically before 27 weeks of gestation, because the cases were all refered from another hospitals. The early detection and termination of pregnancy with patient's permission are considered to be the best way because anencephaly is lethal anomaly of CNS with reasonable accuracy (8).

With ultrasonographic findings of fetal hydrocephalus, verious criterias based on the measurement of biparietal diameter (BPD) were establised (9). It has been recently suggested that with ultrasonography the ventriculomegaly should be identified when LVR is greater than that of the normal range (1)(4), the width of lateral ventricular greater than 1.1cm in the third trimester (10), a shruncken or compressed choroid plexus surrounded by cerebrospinal fluid (9)(11) and five echogenic lines are presented, which are composed by the wall echoes of bilateral lateral ventricles and midline echo (12). In our four cases of hydrocephalus, LVR was all beyond the normal range, and the lateral ventricular widths were greater than 1.1cm in 4 cases of the third trimester. Moreover, compressed choroid plexus was detected sorrounded by cerebrospinal fluid (CSF) in one case. But the five echogenic lines were not detected clearly in each case. In concern with the prognosis of fetal hydrocephalus, the thickness of cerebral mantle was observed carefully (9)(13). Fetuses with the mantle less than 1.0cm in thickness have poor outcomes, especially in fetuses with less than 0.5cm mantle the prognosis was severe (13). In our cases, three cases with less than 1.0cm mantle were delived in stillbirth. On the other hand, one baby whose brain mantle was thicker than 1.0cm was alive under the pediatric neonatal management. This results were in fairly agreement with previous reports.

The characteristic cranial deformity called "Lemon sign" was reported to be identified frequently in fetus with meningocele (14). In our three cases of hydrocephalus with sacral meningocele, "Lemon sign" was noticed in only one case retrospectively (Fig.4).

The ultrasonographic diagnosis of fetal encephalocele was not so difficult, but the differenciation from other mass lesions

arising adjacent to the head, was important (15). In our case, we could differenciate by the detection of cerebral flax and midbrain in the cystic mass ultrasonically.

Early diagnosis and assessment of pathogenic grade of fetal anomaly of CNS are the most important thing, because some fetal anomalies of CNS have been treatable and currable with recent advances of in utero surgical technique (8)(16)(17). Therefore, ultrasound should be a useful diagnostic tool for detection and evaluation of fetal anomalies of CNS, antenatally.

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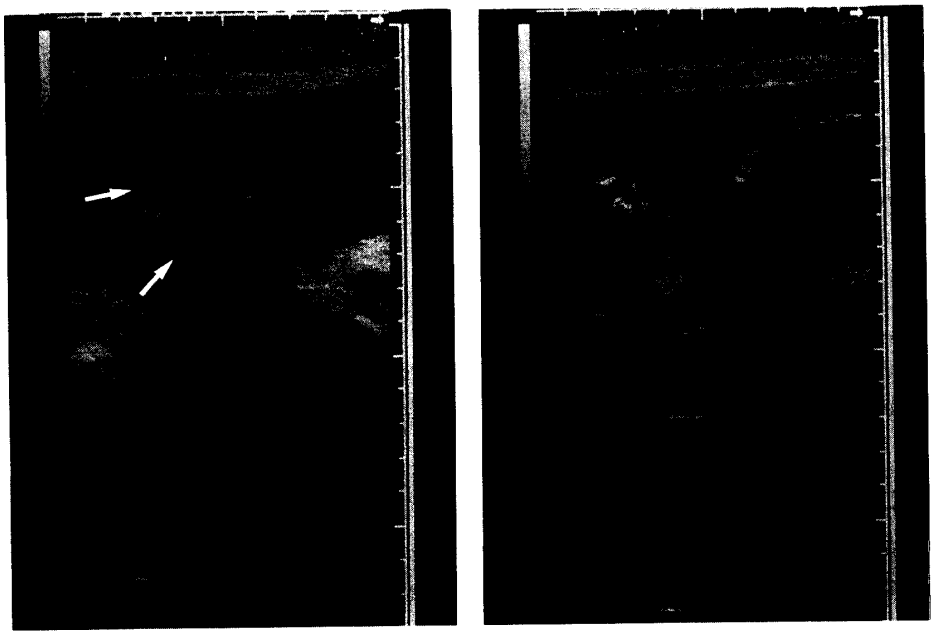

Fig. 1 Ultrasonograms of anencephaly diagnosed at 25 weeks of gestation. (Left : transverse view, right : saggital view) Arrows show the orbita.

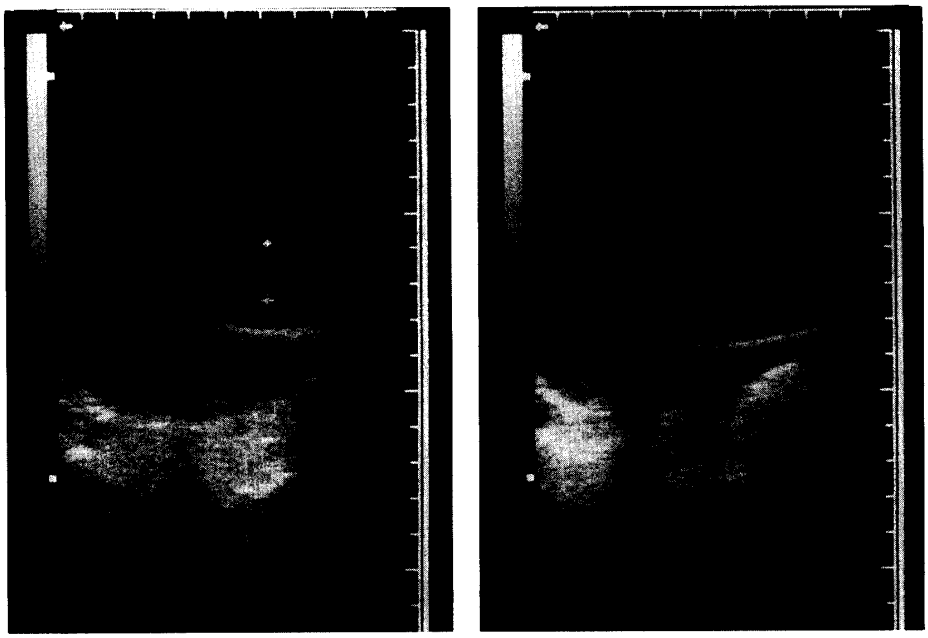

Fig. 2 Ultrasonograms of hydrocephalus at 30 weeks of gestation. (Left : coronal view, right : transverse view) The prominent extension of the body margin of lateral ventricle was observed.

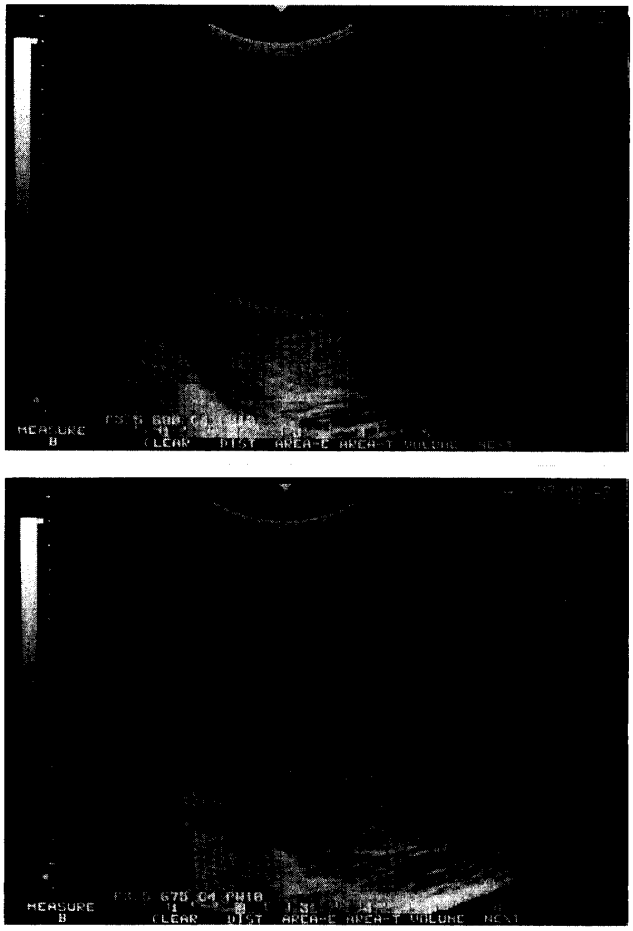

Fig. 3
Ultrasonograms of hydrocephalus at 35 weeks of gestation. The cerebral mantle was significantly thin.

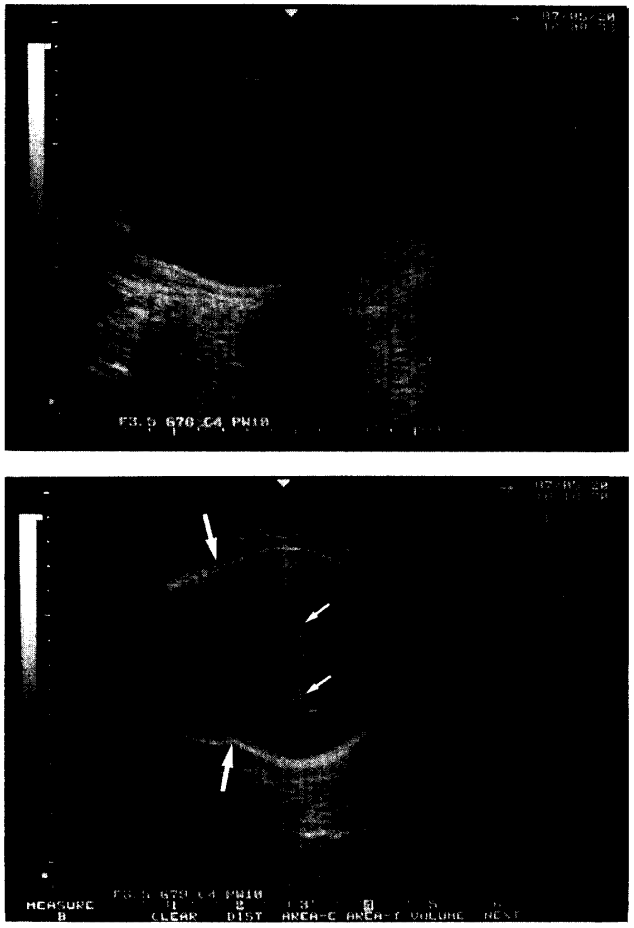

Fig. 4
Ultrasonograms of hydrocephalus at 36 weeks of gestation. (a : coronal view, b : transverse view) Shrunken small choroid plexus was demonstrated.(small arrows) Large arrows show the "Lemon sign".

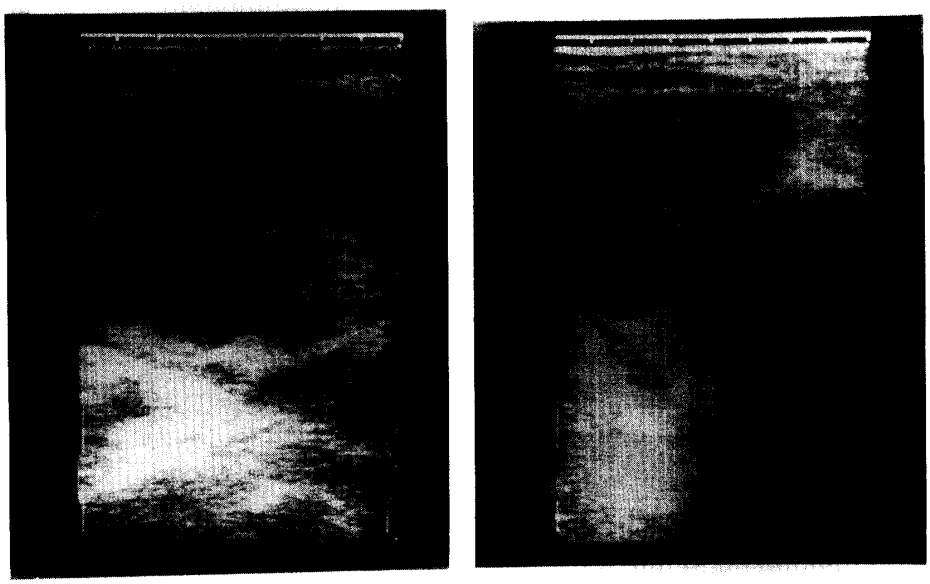

Fig. 5 Ultrasonograms of encephalocele. The cerebral falx and midbrain were evident in the cystic mass.