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CASE REPORT

Prenatal Diagnosis of the Arnold-Chiari Malformation with Spina Bifida : A Case Report

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ABSTRACT

A pregnant woman prenatally diagnosed with the Arnold-Chiari malformation with spina bifida at 18 weeks of gestation was reported. Sonographic evaluation was performed for routine screening. The sonographic findings demonstrated ventricular dilatation, a lemon-shaped cranium (lemon sign), obliteration of the cisterna magna (banana sign) and a dysraphic defect in the lower lumbar spine. The pregnancy was terminated. Postnatal finding confirmed the diagnosis.

Key words : Arnold-Chiari malformation with spina bifida, prenatal diagnosis

Spina bifida refers to a defect in the spine resulting from failure of the two halves of the vertebral arch to fuse. These lesions usually occur in the lumbosacral and cervical regions. Lemon sign, banana sign, and ventricular dilatation were cranial and intracranial findings which attribute to the Arnold-Chiari malformation and have proven to be extremely useful for predicting the presence of spina bifida. Spina bifida is a major type of neural tube defect; the prevalence has been noted, with the highest rates reported in the United Kingdom and the lowest rate in Japan.[1,2] The vast majority of neural tube defects are sporadic and are believed to be multifactorial in origin.[2,3]

Case report

A pregnant woman was seen at the antenatal clinic, Nakornpathom Hospital and was prenatally diagnosed with the Arnold-Chiari malformation with spina bifida at 18 weeks' gestation. Medical and obstetric history were unremarkable and there was no history of familial disease. The demographic data, sonographic findings and postnatal appearance are summarized as follows :

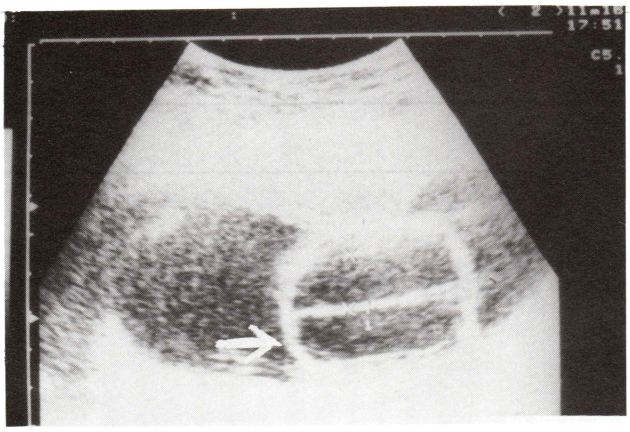

Fig. 1. Ventricular dilatation and lemon sign.

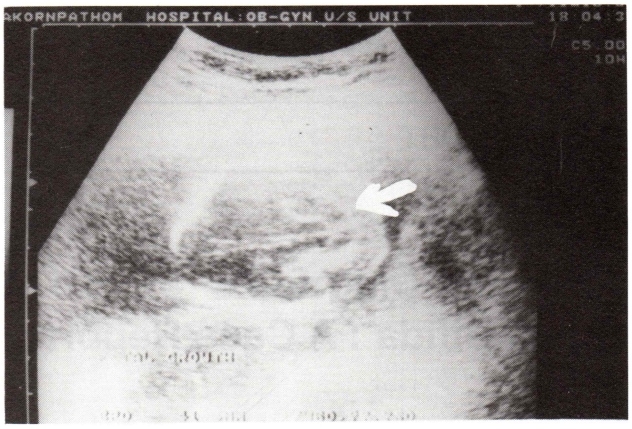

Fig. 2. Banana sign.

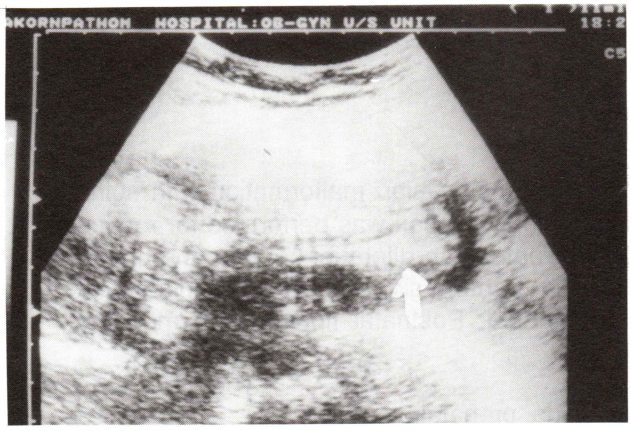

Fig. 3. Dysraphic defect of lumbar spine.

Fig. 4. Photograph with lemon-shaped cranium.

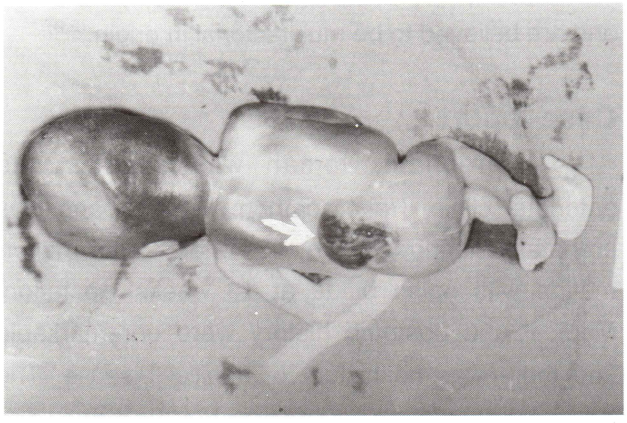

Fig. 5. Photograph with spina bifida.

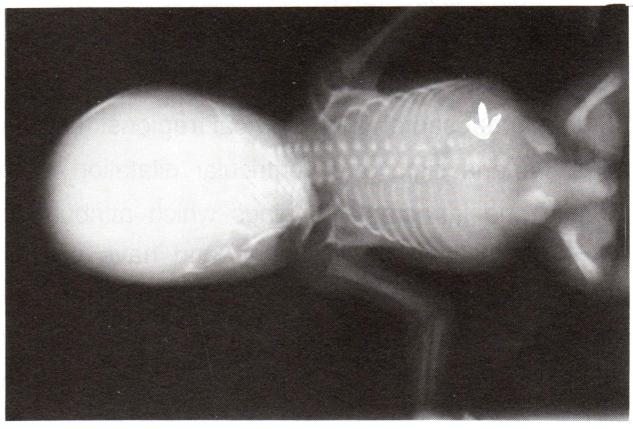

Fig. 6. Radiograph with spinal defect.

Table 1. Mechanism of sonographic findings of the Arnold-Chiari malfomation

Sonographic findings	mechanism/pathophysiology
ventricular dilatation	- obstruction to subarachnoid pathway
lemon sign	- low intracerebral pressure and relatively malleable frontal calvarium
banana sign	- part of the cerebellum displaced into the cervical canal related to tethering of spine

Age 33 Years
Parity 3-0-0-2
Weeks of gestation.....18
Maternal complication.....none
Indication for sonographic examination.....screening
Sonographic finding

- Single viable fetus with breech presentation
- Fetal head : BPD = 41.5 mm
  ventricular diameter 15.5 mm
  lemon- shaped cranium
  banana-shaped cerebellum
- Femur length = 25.1 mm
- Amniotic fluid volume : normal
- Placenta : posterior, upper (grade I)
- Dysraphic defect in the lower lumbar spine

Mode of termination.....prostaglandin with oxytocin
Fetal outcome.....male, stillbirth (300 g)
lemon-shaped cranium
lumbar spinal defect

Based on the sonographic features, the diagnosis of Arnold-Chiari malformation with spina bifida was made and termination of pregnancy was carried out after counseling. Postnatal appearance and radiographs were consistent with prenatal sonographic findings. (Fig. 1-6)

Discussion

In this case, based on sonographic findings (ventricular dilatation, lemon-shaped cranium, banana-shaped cerebellum and dysraphic defect in the lumbar spine), the diagnosis of Arnold-Chiari malformation with spina bifida can definitely be made. The mechanism or pathophysiology of sonographic findings are summarized in Table 1.[4-7]

Of these sonographic findings, the banana sign is highly predictive of spina bifida and is present regardless of the fetus imaged before or after 24 weeks' gestation.[6] In contrast, the lemon sign may disappear after a pregnancy interval of 24 weeks and invariably disappear by 34 weeks.[4]

The case presented was picked up during a routine second trimester ultrasound screening. When detected at such an early stage of pregnancy the condition is invariably incompatible with life. The patient and her partner should be counselled with termination of pregnancy in mind.

Although routine ultrasound screening is accepted in Europe and North America much study is required to evaluate the cost-benefit if it were to be introduced in developing countries such as in Thailand.

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