Case Report

Arnold Chiari Malformation type-2, fetal magnetic resonance imaging findings in a case

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ABSTRACT

Objective: Arnold-Chiari malformation (ACM) is one of the factors that cause death in newborns and infants. Case report: ACM-II detected at 23 weeks 4 days of gestation was presented. The role of prenatal MRI in the recognition of these features, malformation and prognostic value was discussed.

Conclusion: Ultrasonography (US) is the preferred imaging method for pregnant patients. For the diagnosis of ACM II in the developing fetus, direct and indirect US findings must be fully known. However, Magnetic Resonance Imaging (MRI) is increasingly used in patients whose sonographic diagnosis is uncertain. Fetal MRI is performed for patients such as central nervous system (CNS) abnormalities, neural tube defects, congenital diaphragmatic hernia and masses obstructing the airway that are considered fetal surgery.

Keywords: Arnold Chiari Malformation Type II, Fetal MRI, Prenatal Diagnosis

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Introduction

Arnold Chiari malformation (ACM) type II is most common in newborns and infants, characterized by the displacement of cerebellar tonsils, cerebella fourth ventricle sections, pons, and medulla oblongata from foramen magnum to spinal canal. This condition is usually accompanied by hydrocephalus and myelomeningocele \cite{1}. Diagnosis in neural tube defect (NTD) requires experience and careful examination. Small open NTDs can be missed despite careful examination. Therefore, cranial anomalies are now used as a marker of the presence of spina bifida. Symmetrical flattening in the ventral part of the cranium is called the ‘Lemon’ sign. Cerebellar findings are associated with NTD by taking the name ‘Banana’ sign. These findings are used as an indicator of spinal patency \cite{2}.

Ultrasonography (US) is the preferred imaging method during pregnancy. However, Magnetic Resonance Imaging (MRI) is increasingly used in patients where the diagnosis is uncertain. MRI is typically used for the evaluation of abnormal fetal central nervous system (CNS), further characterization of complex anomalies that have not been fully characterized by US, and evaluation of patients who require fetal surgery. In this case report, we aimed to present fetal MRI features of ACM-II\cite{3,4}.

Case presentation

According to the last menstrual date, 23 weeks and 4 days; according to biparietal diameter (BPD) 21 weeks and 4 days pregnant patient admitted to the clinic for routine gestational control. In the patient with the first pregnancy, dilated lateral ventricles and spina bifida were observed on fetal US.

Figure 1. Lateral ventricles on MR imaging

The patient was evaluated with fetal MRI for detailed anomaly screening and further research. The lateral ventricles were clearly visible on MRI (Figure 1). In cerebellum, convex appearance known as ‘Banana sign’ was observed (Figure 2).

There was also ‘Lemon sign’ of cranium (Figure 3). At the level of the atrium, the diameter of the lateral ventricle was measured 12 mm.
Dilatation was not observed in the third and fourth ventricles. The posterior fossa was obliterated. The corpus callosum was thinner secondary to the dilatation of the lateral ventricles and was not clearly observed in the posterior segment. Septum pellicidum was present.

Neural tube defects can easily be recognized with US and in cases the fetal anomaly is incompatible with life, termination of pregnancy is available. Continuation of pregnancy or termination decision should be taken with the family and detailed information should be given [7]. However, in suspicious or delayed diagnoses, posterior fossa findings of ACM and myelomeningocele can be evaluated in detail by fetal MRI [8]. The fetal posterior fossa is small-sized and the tentorium is down-situated, the cerebellum exhibits a dysmorphic pear-shape. Periserebellar fluid fields are obliterated. These findings are often accompanied by hydrocephalus, commissural agenesis and bone-meningeal anomalies [6].

At the lower lumbar segments, there was a 2 cm fusion defect in the posterior part of the vertebra (spina-bifida). There was hyperintense cystic pattern extending outward from the defect and hypointense neural elements (meningomyelocele) were observed in this localization (Figure 4).

US is the first line imaging method for fetal imaging. However, limitations include operator variability, fetal position, gestational age effects (poor visualization, skull ossification), and tissue definition. Early studies using MRI in the evaluation of fetal morphology were prevented by fetal movement. The software and hardware available for MRI now allow fetal imaging without maternal or fetal sedation, allowing MR examinations to be performed with high quality images that are now available in less than 1 second [12]. MRI can assist in diagnosis, patient counseling, and case management of fetuses suspected of having CNS anomalies. US of fetal CNS are restricted in conditions such as; the evaluation of some anomalies with nonspecific appearance, technical factors

Consequently ACM-II and fusion defect in posterior vertebrae in lower lumbar segments and accompanying meningomyelocele were reported in fetal MRI.

**Discussion**

ACM is divided in to four types according to the degree of the disease. In ACM Type 1, the cerebellar tonsils displaces more than 4 mm from the foramen magnum down to the cervical spinal cord. This situation blocks the Cerebro Spinal Fluid (CSF) circulation between the spinal cord and intracranial. In this way, it can cause fluid to accumulate in the spinal cord called syringomyelia or hydromyelia. It is the most common form of Chiari malformations. It is often not noticed until it causes problems in the adolescent or adult life period [5].

ACM-II is characterized by a small posterior fossa with cerebellar compression and posterior brain herniation through the Foramen magnum. Open NTD is always present [1]. Myelomeningocele is a congenital disease, when the spinal cord is not fully closed while developing in the intrauterine period and has an open (sac shaped) spinal cord defect at birth. It may be accompanied by hydrocephalus, cardiovascular anomalies, imperforated anus and other gastrointestinal and genitourinary abnormalities [3].

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Cerebellar appearance findings and neurocranium contour anomalies with US are important markers for spina bifida. Lemon sign is very helpful in the diagnosis of spina bifida in high-risk population before 24 weeks of gestation; however, as the gestational age progresses, the lemon sign may become ambiguous as a result of fetal development; therefore, its reliability is reduced [9,10]. The lemon sign is not specific to spina bifida. Encephalocele may also accompany Dandy-Walker malformation accompanied by encephalocele, cystic hygroma, diaphragmatic hernia, corpus callosum agenesis, fetal hydrops, umbilical vein varices and double-vessel cord anomalies. In the presence of Lemon sign, cranial findings such as ventriculomegaly, microcephaly, cisterna magna obliteration, compression of the cerebellar hemispheres and orientation in the ventral direction (Banana sign) should be examined and the vertebral column should be carefully evaluated [10,11].

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limiting the resolution of the side of the brain near the transducer, ossification and thin parenchymal abnormalities that may not prevent the visualization of posterior fossa structures. MRI provides direct multi-plane imaging of the brain parenchyma, thereby allowing a detailed assessment of the CNS that is not possible by US due to fetal position or advanced gestational age and skull ossification. Preliminary studies suggest that MRI can change diagnostic accuracy and counseling for many fetal CNS lesions [3,12]. But fetal MRI also have some limitations. It is operator dependent and maternal large size effects patient to fit scanner. One other limitation is claustrophobia in some patients [12]. Nevertheless, MRI is superior to US, particularly in the diagnosis of migration disorders, callosal anomalies and pathologies of the posterior fossa in literature [13].

With the development of MRI hardware and software products, very fast MRI sequences with T1 and T2 weighted echo planar and fast gradient echo sequences have begun to be used. High-contrast-resolution fetal tissue imaging was possible without the need for fetal sedation or paralysis. In cases where the US examination is inadequate, there are studies in the literature showing that the most successful examination area of fetal MRI is the CNS and spinal cord [3,4,14]. It has been showed that the fetal brain and spinal cord can be evaluated more successfully than other organ systems. At the same time, adequate plan imaging without being affected by the head bones with MRI makes MRI superior to US in evaluating posterior fossa malformations [12].

The use of advanced fetal MRI imaging techniques allows us to better evaluate the anatomy and pathology of the fetal brain and spinal cord structures in the antenatal period. Prevalence of fetal MRI use in posterior fossa and spinal cord evaluation is important for the clinician to guide and inform families through the detection of rare anomalies, detailed detection of findings associated with US anomalies. Although rapid MRI techniques are widely available, few practitioners are familiar with fetal anatomy and pathology with this technique [15].

As a result; Imaging properties of the ACM should be known very well by radiologists. US is primary diagnostic method in obstetric follow-up and it was found that MRI showed a significant superiority to US in detecting posterior fossa malformations and corpus callosus anomalies especially in the third trimester [15]. Therefore, in cases where a detailed description of the CNS anomalies is required, MRI can be used as an effective complementary diagnostic method [3,4,14].

Disclosure
Authors have no potential conflicts of interest to disclose.

References