CASE REPORT

Ultrafast Sequence in Fetal Magnetic Resonance Imaging for Evaluation Chiari III Malformation

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ABSTRACT

We report a case of Chiari III, a very rare congenital malformation. Fetal magnetic resonance imaging (MRI) was performed at the third trimester. She was referred for fetal MRI after ultrasound revealed an occipital encephalocele. Fetal MRI confirmed the presence of encephalocele encompasses occipital lobes with additional information herniation of cerebellum into cervical canal through foramen magnum change the diagnosis as Chiari III malformation. T2-HASTE sequence clearly depicts of syringomyelia, but the level of cerebellum herniation can demonstrate by TrueFISP sequence.

Key words: Chiari III-fetal MRI-T2HASTE-TrueFISP, occipital encephalocele, syringomyelia

INTRODUCTION

Chiari malformation type 3 is a very rare anomaly, characterized by small size posterior fossa, herniation of cerebellum through foramen magnum into cervical canal and associated with low occipital encephalocele due to un united neural arch in the cervical canal.[1,2] According to Sarnat, Chiari III is an extreme form of Chiari II.[3] The prognosis is not good and sometimes has life-threatening complications. Ultrasound as the main modality for prenatal screening can detect encephalocele, but it has a limitation to evaluation posterior fossa. Magnetic resonance imaging (MRI) with ultrafast sequence resulting high quality of images can delineate cerebellar structures and gives additional information about morphology and abnormal cerebellar and brain stem position. We report a case of Chiari III malformation which was identified using fetal MRI. MRI was performed with Siemens Avanto 1.5T system with body matrix coil. Axial, sagittal, and coronal orientation was acquired with 5 mm slice thickness, 0.5 mm gap covering thoracoabdominal field of view. MRI of the brain was acquired with 3–4 mm slice thickness, 0 mm gap in all three orthogonal planes. Sequence uses are T2-HASTE, TrueFISP, and T1-TurboFLASH. Fetal MRI obviously has seen syringomyelia on T2-HASTE but quantification level cerebellum herniation better depiction by TrueFISP sequence.

CASE REPORT

A 27-year-old woman G2P1A0 was referred for MRI examination at 32 weeks gestation due to encephalocele. Fetal MRI revealed quite large low occipital bony defect, absence of a myelomeningocele distally. An encephalocele size ± 62 mm×42 mm×47 mm, neck 22 mm, involves occipital lobes without cerebellum involvement [Figure 1a]. Head circumference was the 3rd centiles. The posterior fossa was small and foramen magnum was obliterated. There was an inferior displacement of cerebellum and brain stem into cervical canal [Figure 1b and c]. Level cerebellum herniation based on segmentation of cervical bone can be seen clearly on T2-TrueFISP [Figure 1c]. Posterior horn of lateral ventricle and 4th ventricle was not visualized. There was no hydrocephalus. There was a cystic intensity inside the cervical spinal cord extending upper thoracic level [Figure 2a and b].

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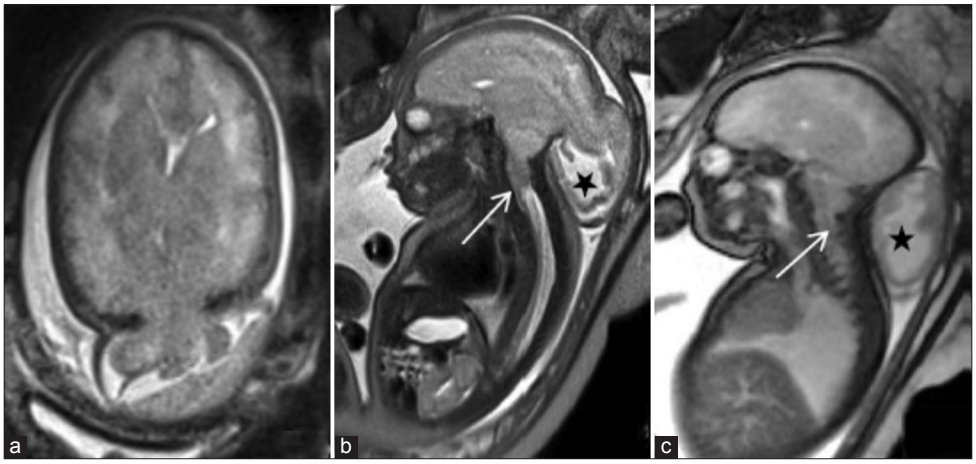

Figure 1: (a and b) Axial and sagittal T2-HASTE showed occipital bony defect and protrusion occipital lobe. (c) TrueFISP showed segmentation of cervical bone. (b and c) Displacement brain stem and cerebellum into cervical canal (arrow) and high signal intensity of neural tissue in sac due to gliosis (star)

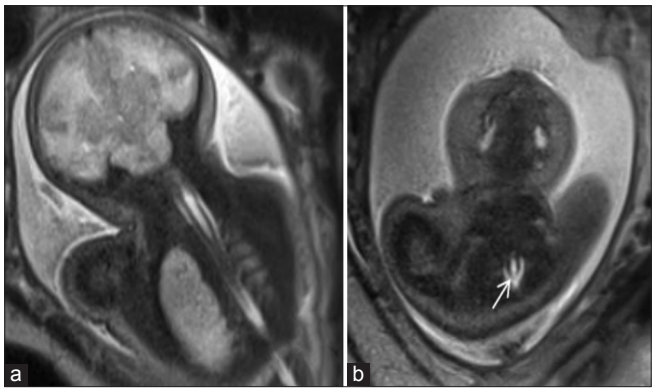

Figure 2: (a) Coronal T2-HASTE. (b) Axial T2-HASTE revealed syringomyelia in cervical cord extending upper thoracic. Bull eye pattern (arrow)

DISCUSSION

From a case above, it is clear that fetal MRI can provide additional information regarding the shape of posterior fossa and organs inside. Small posterior cranial fossa with normal size of cerebellum makes it squeezing out through foramen magnum into cervical canal. Posterior bony defect and part of brain protrusion can be determined by fetal MRI. In our case, the osseous defect at low occipital bone and bilateral occipital lobe herniated through the defect without cerebellum involvement. Based on these findings, we conclude a case above as Chiari III malformations. Fetal MRI can exclude isolate occipital encephalocele and syndromic occipital encephaloceles as a differential diagnosis.[4] Isolated occipital encephalocele lacks of intracranial Chiari II findings. Syndromic occipital encephaloceles such as Meckel–Gruber have multicystic kidney and polydactyly, but in this case, no abnormality was found in other organs.

Chiari III is a very rare case, and the pathogenesis is believed related to Chiari II.[3] The imaging characteristics are the same as Chiari II except in Chiari III; there is a lower occipital or upper cervical encephalocele. Some of Chiari II theories have been suggested, the theory of discredited traction by Lichtenstein (1942). This theory explains meningoencephalocele at distal spine causes tethered of spinal cord and attracted the medulla and cerebellum into spinal canal through foramen magnum. Pulsion hypothesis theory by Gardner 1977 early fetal hydrocephalus forces the entire contents of the posterior fossa to pass through the foramen magnum.[5] In the crowding hypothesis by Marin-Padilla and Marin-Padilla, 1981, the cerebellum and lower brain stem grow beyond the capacity of the posterior fossa.[6] This theory contributes to Chiari late gestation but not as primary pathogenesis because Chiari develops before midgestation when there is no growth restriction within the confines of posterior fossa.[3] The hydrodynamic oligo-cerebrospinal fluid (CSF) theory by McLone and Knepper open neural tubes causes leakage so the CSF is decreasing, causing no accumulation of fluid in the intraventricular. This condition is insufficient for making cerebral vesicle distension during initial development which causes a small posterior fossa.[7,8] Birth trauma hypothesis by Williams, 1991, according to Williams patients with difficulty in labor have significant structural abnormality of the basicranium, leads to invagination of foramen magnum. However, this theory is not suitable because the existing data suggested, Chiari II occurred during midgestation. The theory of segmentation disorder by Sarnat 2008.[3] Chiari II occurs more due to segmentation disorders of multiple hindbrain rhombomeres than secondary disturbances caused by physical and mechanical factors. Sarnat's theory can explain the occurrence of histologic dysgenesis protrusion of cerebellum tissue through the foramen magnum and the occurrence of Chiari II during midgestation.[3] According to Sarnat, Chiari III is an extreme form of Chiari II, but Barkovish has different opinion. Chiari III should be consider a high cervical myelocystocele.[9] Ultrafast sequences in fetal MRI have fewer artifacts result better picture. Lee *et al*. used single-shot fast spin-echo (SSFSE or T2-HASTE).[10] We use both of T2-HASTE and TrueFISP sequences.

TrueFISP has advantages to delineation intervertebral disc level as reported by Abele *et al*., so the level of cerebellar tonsil herniation can be determined as quantitative.[11] Syringomyelia is better depiction by T2-HASTE. Some theories of syringomyelia have been proposed. The water hammer theory by Gardner during systolic CSF flow from the 4th ventricle to foramen magnum, if outflow was obstructed, CSF was forced into the central canal resulted in syringomyelia, but the majority of human do not have patent connection between the central canal and 4th ventricle.[12] New concept pathophysiology of syringomyelia by Greitz *et al*. intramedullary pulse pressure theory is acceptable. The main principles of this theory are the syringomyelia which is caused by repeated mechanical distension of spinal cord and the ensuing cavitation arises from extracellular fluid originating from high-pressure system in the microcirculation of spinal cord.[13,14]

As reported by Smith, fetal MRI has limited for the evaluation of posterior corpus callosum, we cannot see of posterior corpus callosum suspicious hypogenesis of midline structure or distortion due to displacement lower occipital lobe.

The treatment of encephalocele in Chiari III is surgical resection and reestablishment of CSF flow. The encephalocele in our case is enormous and contains large amount brain tissue different from the previous case reported by Lee *et al*. and Smith *et al*.[10,15] Hyperintense in neural tissue can be seen both on T2-HASTE and TrueFISP due to gliosis. We should determine exactly part of brain tissue which is herniated into encephalocele. Herniation of the brain stem in encephalocele is contraindication immediate surgical closure.

Patients with Chiari III malformation have poorer outcomes than Chiari II. The mortality rate is high. In our case, the fetus died at 34-week gestation.

CONCLUSION

Fetal MRI is useful for diagnosis Chiari III malformation and also to determine the exact part of the brain in cephalocele for further planning operation. Both sequences are useful for looking cerebellum and brain stem herniation, but TrueFISP is better for delineation intervertebral disc level.

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How to cite this article: Wirasasmita DA, Abdurrazak G, Adenin I. Ultrafast Sequence in Fetal Magnetic Resonance Imaging for Evaluation Chiari III Malformation. J Clin Res Radiol 2019;2(2):1-3.