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Cephalopagus conjoined twins presented with encephalocele: diagnostic role of ultrafast MR imaging

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ABSTRACT
Conjoined twinning is a rare abnormality and cephalopagus is a very rare form of conjoined twins. We report a case of cephalopagus conjoined twins with encephalocele and omphalocele which diagnosed by ultrasonography and ultrafast magnetic resonance (MR) imaging at 24 weeks of the gestation. Ultrafast MR imaging can provide image quality superior to two dimensional ultrasonograpy and should be considered an adjunct to ultrasound for antenatal characterization of some anomalies. To the best of our knowledge, this is the first case of cephalopagus conjoined twins with encephalocele and omphalocele which diagnosed by ultrasound and ultrafast MR imaging.

Key words: • *cephalopagus* • *conjoined twins* • *encephalocele* • *ultrafast MR imaging*

Conjoined twinning is a very rare abnormality, which occurs in one of every 100 sets of monozygotic twins and in one of every 50,000 to 100,000 births (1). The twins are believed to be monozygotic and do not complete division of the embryonic disc at gestational days 15-17. They are classified according to their most prominent site of connection. Cephalothoracopagus is a very rare form of conjoined twinning, which occurs in one of every million births and in one of every 58 sets of conjoined twins (2). Classic cephalopagus twins are joined from the top of the head to the umbilicus, sharing a single foregut as well as two relatively normal hearts. The intermediate cases share either a single, very abnormal heart, or double aortic arches (3). To the best of our knowledge, this is the first case of cephalopagus conjoined twins presenting with encephalocele and omphalocele. Diagnosis was made by prenatal ultrasonography and ultrafast magnetic resonance (MR) imaging at 24 weeks of the gestational age.

Case report

A 30 year-old woman, gravida 3, para 2, at 24 weeks of gestation was referred from a regional hospital because of polyhydroamnios with a 'singleton fetus'. To herself, she seemed much larger for her gestational age compared to her previous pregnancies, and she perceived movement of more than one fetus. Her history was otherwise unremarkable. Sonographic examination revealed a single cranial structure, irregular contour with one face, and two eyeballs. Head circumference of twins could not be measured and intracranial content was invisible. There was a fused thorax and two separate spines, and 2 pelvises and 8 intact limbs were visible. The livers also appeared to be partially fused and 2 bladders were observed. Both twins had male genitalia. There was a single placenta and umbilical cord with three normal blood vessels. Omphalocele was also present. The spinal columns were joined at the cervical area (Figure 1). There was a single heart in the thoracic cavity. A single abdominal circumference and normal appearance of lower limbs were noted. MR examination was performed in a 1.5T superconductive unit, with a synergy body coil (Intera Master, Philips Medical Systems). Balance turbo-field-echo (TR/TE/FA/scan time: 3,0/1,5/80/25,2 sec-breathhold) images in the coronal plain, and single-shot T2 weighted images (free-breath) (831/80/80/20.8sec) in the axial plain were obtained. Ultrafast MR imaging showed polyhydroamnios, cephalopagus conjoined twins with encephalocele and omphalocele (Figure 2 and 3). There was abnormal brain anatomy, which was difficult to characterize, and there was partial fusion of some structures. Because of poor prognosis, gestation was terminated with intravaginal misoprostol 400µg (Cytotec®). The aborted male twins weighed 765 gr and were joined symmetrically from the face to the umbilicus (Figure 4). The anterior aspect of the head

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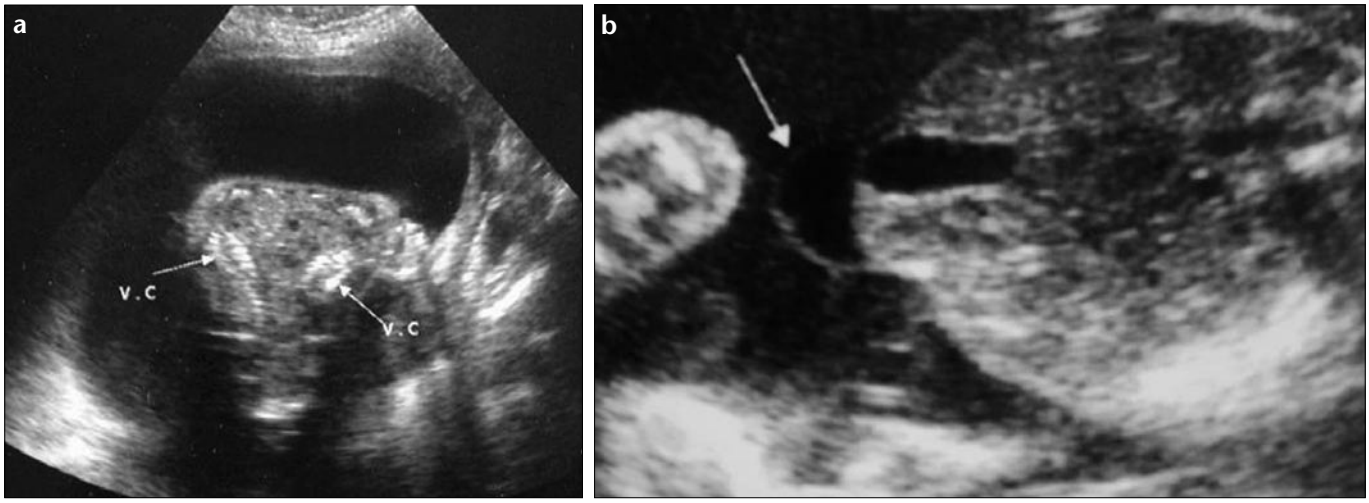

Figure 1. a, b. Fetal ultrasound examination at 24 weeks of gestation revealing two vertebral columns (V.C.) **(a)**, and omphalocele *(arrow)* **(b)**.

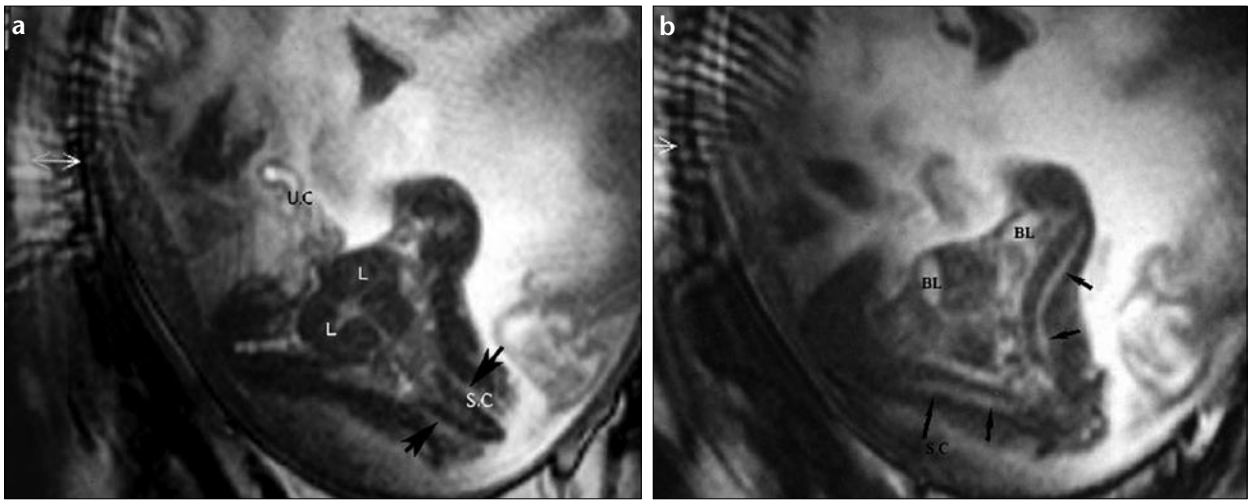

Figure 2. a, b. Coronal MR images demonstrate polyhydroamnios, umbilical cord (U.C), and fused livers (L) **(a)**. Two spinal columns (S.C.) *(arrows)* and bladders (BL) are shown **(b)**.

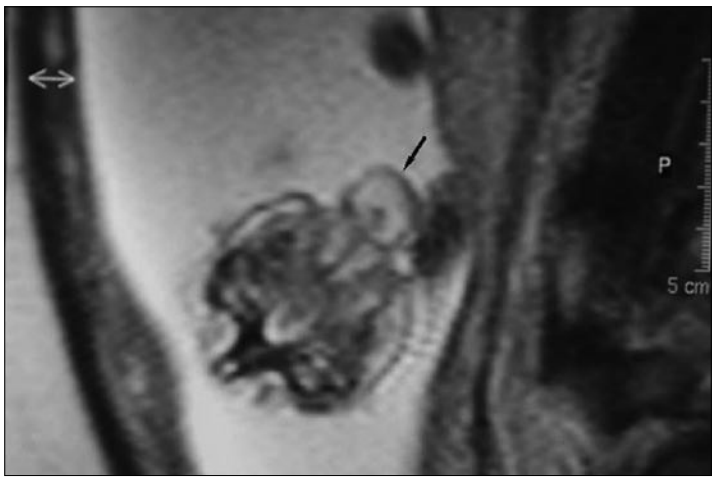

Figure 3. Axial single-shot fast spin echo image shows the encephalocele *(arrow)*.

showed an abnormal face formed by one eye, one ear, half a nose, and half a mouth from each twin. The adjacent corners of the mouths and noses were fused, but the oral cavities and tongues were anteriorly separate and conjoined posteriorly. There was a sac of encephalocele inferior to the nose and covering the mouth. The lower abdomens were separate as were the vertebral columns, genitalia, and extremities. One forearm was rudimentary and contained only two fingers. There were a single large neck and conjoined thorax. Postmortem imaging and autopsy were not performed.

Discussion

Cephalopagus twins are rare. They are fused from the vertex to the umbilicus. There are two faces on opposite sides of the conjoined head; one face is usually rudimentary. The thorax is joined, including the heart, even if only by a single interatrial vessel. The diaphragm and liver are fused, and each twin contributes one lung to each trachea. The pelvis and urinary tracts are separate and normal. There are four arms and four legs. It is not usually possible to separate these twins.

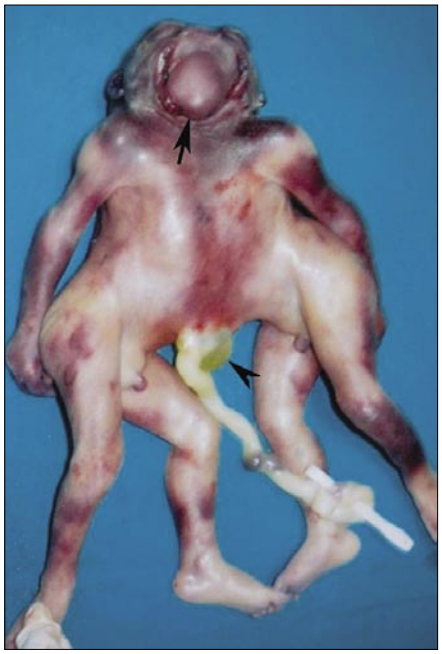

Figure 4. Postnatal image of twins revealing omphalocele *(arrowhead)* and encephalocele *(arrow)*.

In the present case, upon sonographic examination, the livers appeared to be partially fused and 2 bladders were observed. Additionally, omphalocele was present, and there was a single cranial structure, and irregular contour with one face and two eyeballs. Head circumference of twins could not be measured, intracranial content was invisible, and encephalocele could not be visualized. For confirmation of the sonographic findings, ultrafast MR imaging was performed, which allowed us to obtain more precise imaging of the cephalothoracopagus conjoined twins, including enhanced visualization of encephalocele and polyhydroamnios. However, sonographic observation of omphalocele was superior.

Conjoined twins often have discordant anomalies; these often occur in the twin on the right. Levin et al. surveyed 167 pairs of conjoined twins and found that the presence of laterality defects, especially reversal of cardiac situs, depended upon the orientation of the conjoined twins. Such defects were common in thoracopagus and dicephalic parapagus twins, but did not occur in craniopagus or ischiopagus twins (4).

Prenatal sonographic is the primary modality of imaging in pregnancy, which allows direct, real-time examination. It usually permits accurate diagnosis of congenital anomalies, such as conjoined twins, and also reveals such details as degree of fusion. Prenatal diagnosis of shared organs is of great importance for the consideration of possible surgical separation or termination of pregnancy (5). Termination of pregnancy can be done before 24 weeks of gestation by vaginal route with destructive procedures. In the presented case, surgical separation was not technically possible and survival was unlikely according to sonographic findings.

Antenatal MR imaging has been of limited clinical value owing to poor image quality before ultrafast scanning techniques were developed. This was due to the long acquisition times that were needed to achieve a sufficiently high spatial resolution for assessment of small fetal anatomic structures, which resulted in severe motion artifacts. This problem has now been overcome by recent technical improvements. Ultrafast MR imaging has been available since the early 1990's (6), but only recently has been considered as an adjunct to high-resolution ultrasonography in antenatal diagnosis (7). Shakuda et al. compared the efficacy of fast MR imaging (breath-hold fast spin echo T2-weighted and fast gradient echo T1-weighted sequences) to ultrafast MR imaging (half-Fourier acquisition single-shot turbo-spin-echo sequence) in the evaluation of fetal central nervous system abnormalities at late gestational age, and they also compared the capability of fast and ultrafast MR imaging to prenatal US to asses fetal central nervous abnormalities. They found that ultrafast MR imaging demonstrated additional findings over US (8). Casele et al. reported the first case of cephalopagus conjoined twins diagnosed with ultrafast MR imaging (9). They suggested that ultrafast MR imaging can provide superior image quality to two-dimensional ultrasonography and should be considered an adjunct to US for antenatal characterization of some anomalies. Ultrafast MR imaging provides excellent contrast with cerebrospinal fluid between the brain and spinal cord. Compared to ultrasonography, ultrafast MRI facilitates more precise visualization of antenatal brain characterization and spinal cord anomalies. Herein, we presented a case of cephalopagus conjoined twins that to the best of our knowledge is the first case presented with encephalocele and omphalocele. We also suggest that based on our findings, ultrafast MR imaging is superior to two-dimensional ultrasonography in imaging and characterizing antenatal fetal anomalies and should be considered as an adjunctive imaging modality in such cases.

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