Cephalothoracopagus janiceps disymmetros A comparison between antenatal and postnatal findings

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Submission date: 10-May-2023 03:09PM (UTC+0700)
Submission ID: 2089319775
File name: agus_janiceps_disymmetros_A_comparison_between_antenatal_and.pdf (3.17M)
Word count: 1389
Character count: 7704

Article type : Brief Communication

BRIEF COMMUNICATION

Cephalothoracopagus janiceps disymmetros: A comparison between antenatal and postnatal findings

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Synopsis

The present study reports a case comparing 2D and 3D ultrasound prenatal scans with postnatal MRI in diagnosis of cephalothoracopagus janiceps disymmetros.

Keywords

Antenatal; Cephalothoracopagus; Conjoined twin; Diagnosis; MRI; Postnatal; Ultrasound

Conjoined twins are a rare congenital malformation affecting less than 1% of monozygotic twin pairs [1]. The diagnosis should be suspected when monozygotic fetuses are shown to consistently hold the same fixed position relative to each other [2]. Conjoined twins can be divided into symmetric conjoined twins and heteropagus or parasitic twins [3, 4].

A 27-year-old gravida 2 patient at 28 weeks of gestation was referred to our institute with complaint of contraction and congenital anomalies. After the second day of inpatient care, two-dimensional (2D) and three-dimensional (3D) ultrasound scans were performed with a Voluson E6 (General Electric Medical System & Healthcare, Chicago, IL, USA) machine. This examination was then compared with the postnatal findings after delivery of the fetuses (Figs 1 & 2, Figs. S1–3). Antenatal scans revealed that the twins were ventrally fused from the temporal region of the head to the upper abdomen (until the point of the umbilical cord). Fusion of the temporal lobes of the cerebrums was also observed. The faces were distorted due to oblique fusion and two pairs of eyes (Fig. 1). The twins shared a common thoracic and upper abdominal cavity with fusion of two beating hearts, two pairs of kidneys, and two bladders. The 2D and 3D ultrasound revealed fusion of both livers with separated spleens (Fig. S1a–b). There were four pairs of well-developed limbs and two vertebral columns. We identified the gestational age of the first fetus (28 weeks and 3 days) by assessing its femur length (5.4 cm) and transcerebellar diameter (3.30 cm). The femur length of the second fetus was 5.23 cm and the transcerebellar diameter was 3.15 cm (27 weeks and 4 days of gestation). Polyhydramnios was noted in our analysis. A 3D ultrasound examination revealed surface-rendered images of the conjoined region, two clear pairs of limbs, a malformation of craniofacial appearance suggesting hypotelorism, a flat nasal bridge, micrognathia, and low-set ears (Fig. 1b–c).

After obtaining informed consent from the patient, a cesarean section was performed at 29 weeks and 3 days of gestation. The female twin babies, weighing 2100 g, survived for 1 hour. The postnatal examination of the twins, performed postmortem, confirmed the

antenatal diagnosis (Fig. 2a–c, Fig. S2a–b). Postmortem pathological analysis could not be performed as the patient did not provide consent for this.

A whole body MRI assessment with axial, sagittal, and coronal slices, as well as T1- and T2-weighted images without contrast, also revealed fusion from the temporal cranium to the upper abdomen. There was fusion of the temporal lobes of the cerebrums (Fig. 2a) and a cleft in the temporal lobes and subarachnoid space which was not related to the ventricles. Sulci and Sylvian fissures were not formed with a thick and smooth cortex. There was an absence of gyri on the surface of the brain. Gray and white matter differentiation was not clear. The ventricular and the cisternal systems were unclear. There were left and right lateral ventricles in both infants. There was no visible pituitary and optic chiasm. No abnormality of the bulbus oculi and retro-orbital space was observed in both infants (Fig. 2a). In the thoracic part, the twins shared an upper respiratory tract including the trachea and lungs (Fig. S3b). Fusion of the two hearts was observed in the first infant but it was hard to evaluate the border (Fig. S3c). Each twin appeared to have cervical, thoracic and lumbosacral vertebrae (Fig. S3a–b). In the abdominal part, there was fusion of the livers with two separated spleens. The upper gastrointestinal tract, stomach and intestines were fused in both twins. Separated pelvises were observed. Finally, the examination revealed normal rectums, uteruses, and urinary bladders.

Prenatal ultrasonography permitted accurate diagnosis of conjoined twins and enabled a detailed analysis of the degree of fusion. MRI plays an important role in the postnatal evaluation of conjoined twins, particularly in cases where fusion of the head or thorax is observed. Antenatal diagnosis helps in counselling the patient about poor neonatal outcomes and planning prompt pregnancy termination to avoid additional risks and complications of delivery.

Author Contributions

AM and E contributed to the acquisition of data. WTP, NB, and PM contributed to the design of the study and drafting of the manuscript. AKS and PML were responsible for critical revision of the manuscript. RMF was responsible for radiology examination and

interpretation. MHA and CK were responsible for the final approval of the manuscript. All authors contributed to and approved of the final version of the manuscript.

Conflicts of Interest

The authors have no conflicts of interest.

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Figure Legends

Figure 1. (a) 2D ultrasound prenatal images of a transverse section of the skull showing the face distorted with oblique fusion; (b) 3D surface-rendered ultrasound prenatal images of face one and (c) face two

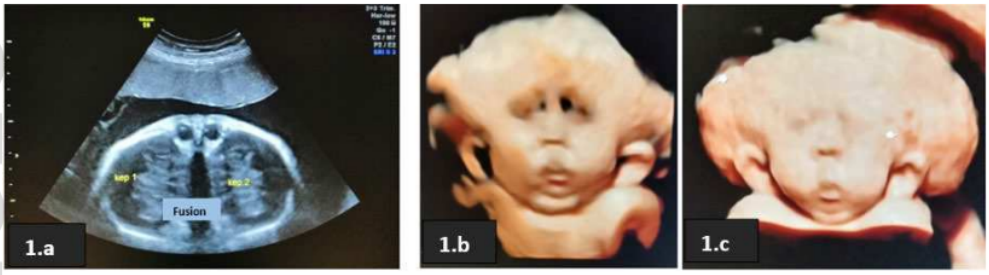
Figure 2. (a) Postmortem MRI of fusion of the temporal lobes of the cerebrums. Postmortem image of the fetal faces after delivery; (b) side one; (c) side two

Supporting Information

Figure S1. (a) 2D ultrasound images showing the twins were ventrally fused from temporal head to upper abdomen up to the umbilical cord. (b) 3D surface-rendered ultrasound images showed one of each fetus' arms hugged each other

Figure S2. Postmortem images of the fetuses at delivery, (a) side one and (b) side two

Figure S3. (a) Postmortem whole-body MRI. (b) fusion at thoracic parts. (c) fusion of two hearts



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