A Monocephalus Diprosopus Fetus: Antenatal Sonographic Findings

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Abstract

Monocephalus Diprosopus is the rarest form of conjoined twins. The etiology of this anomaly is still obscure. We herein report a monocephalus diprosopus case that was diagnosed in week 19 of pregnancy was presented due to its rarity and the significance of its prenatal diagnosis. Recommended a pregnancy termination since it does not have a definitive treatment today, prenatal diagnosis made for such fetuses at an early stage bears importance in terms of lowering the severity of psychological trauma.

Keywords: Monocephalus Diprosopus, prenatal diagnosis, sonography

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Introduction

Conjoined twins are a rare form of monozygotic twins. They occur in 1 per 50,000 to 200,000 live births. Conjoined twins classified into three major groups: twins with a ventral union (cephalopagus, thoracopagus, omphalopagus and ischiopagus), twins with a dorsal union (pygopagus, rachipagus and craniopagus), and twins with a lateral union (parapagus) twins [1,2]. Monocephalus Diprosopus is the rarest form of conjoined twins. A diprosopus twins occurs in 1 per 180,000 to 1 per 15,000,000 births [3]. The etiology of this anomaly is still obscure. A monocephagalus diprosopus case that was diagnosed in week 19 of pregnancy was presented due to its rarity and the significance of its prenatal diagnosis.

Case

The case that presented to her physician in week 19 of pregnancy for the first time for the monitoring of pregnancy was referred to our clinic with cranial anomaly pre-diagnosis. Having 2 gravida and 1 parity, our case of 20 years of age did not have any characteristics “in her personal and family history and the case was a non-kin with her husband. Obstetric ultrasonography demonstrated a 19-week pregnancy according to the last menstruation, and ultrasonographic measurements showed live twin intrauterine pregnancy of 25 weeks conjoined by “cephalic pole” based on biparietal diameter. According to the conducted sonogram, fetuses had two craniums, 4 cerebrums, 4 choroid plexuses, 2 thalami and cerebellums each (Figure 1, 2). However, bodies, vertebral columns, and extremities were single. Normal umbilical cord had a single placenta and comprised of two arteries and one vein. Amniotic fluid index was evaluated as normal. Biparietal diameter measurement was in conjunction with 25-week pregnancy. Following the current condition on results were conveyed to the family, pregnancy of the case was terminated. Total weight of the fetus was 630g and height was 24 cm, and gender was female (Figure 3). Apgar score was recorded as 0 at the 1st minute and 0 at the 5th minute. The case was discharged with full recovery on the postpartum 1st day. Autopsy was not carried out since the family did not grant the permission. When examined the fetus externally, it was observed that two heads were conjoined, entire formations on the faces were double, and that other formations such as body and extremities were single.
Figure 1. Fetuses had two craniums, 4 cerebrums, 4 choroid plexuses, 2 thalami and cerebellums each one.

Figure 2. Fetuses had two craniums, 4 cerebrums, 4 choroid plexuses, 2 thalami and cerebellums each one.
Discussion

Monocephalus Diprosopus, lexically means single head-double faces by definition. It is a historical definition and was adopted by that term since it was accepted in the literature despite not reflecting the actual pathology. On the other hand, this anomaly also bears the name “Monocephalus Diprosopus Tetraophthalmos”. Since access to prenatal sonography is now easier, conjoined twins are better established today. However, perinatal mortality still follows a higher course. Postpartum surgery opportunities are still limited. Mortality and morbidity of the surgery is currently high [4,5].

Although conventional 2D ultrasonography is deemed sufficient for prenatal diagnostic purposes, 3D applications of ultrasonography were also defined in literature [6]. Abnormal shape of the head should pose a warning in conventional 2D ultrasonography. In addition, choroid plexus, thalamus, and cerebellum structures being 2 pairs should be deemed significant hints during diagnosis. Body is one, and extremities are one pair.
Post-mortem examinations of Monocephalus Diprosopuses have also been a matter of debate in literature. In order to better understand the structural foundations of the anomaly such imaging methods as CT are recommended before the autopsy [7]. Since, in our case, postpartum radiologic and pathologic evaluations were not permitted, post-mortem evaluation was possible only externally.

Recommended a pregnancy termination since it does not have a definitive treatment today, prenatal diagnosis made for such fetuses at an early stage bears importance in terms of lowering the severity of psychological trauma.

Disclosure

This study was presented in the 8. Congress of Ultrasound in Obstetrics and Gynecology in September 2012, in Turkey.

Conflict of interest statement

The authors declare that they have no conflict of interest.

References