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Case Report

Dicephalus parapagus conjoined twin: a rare case with review of literature

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ABSTRACT

Conjoined twin is a rare complication seen in 1% of monochorionic twins and associated with severe morbidity and mortality. It occurs due to a division event at the primitive streak stage of the human embryonic development at about 13-14 days after fertilisation, in monochorionic monoamniotic gestations. Early prenatal diagnosis of conjoined twin plays a very crucial role in the management and allows appropriate and timely counselling of couple regarding the different modes of management like early termination of pregnancy or continuation of pregnancy with post-natal surgery. Late diagnoses present with difficult options for parents and obstetrician too. Ultrasound plays a very crucial role in diagnosis of conjoined twin. We are reporting a case of 27 years old primigravida referred to our institute at 13 weeks of gestation with ultra-sonographic diagnosis of dicephalus parapagus conjoined twin and further confirmed after termination of pregnancy.

Keywords: Conjoined twin, Monoamniotic monochorionic, Termination of pregnancy, Ultrasonography

INTRODUCTION

The conjoined twin is one of the rare phenomena seen in 1% of monochorionic monoamniotic pregnancy and is associated with high perinatal mortality. The chances of conjoined twin occur if there is division of zygote later than 12 days of fertilization.¹ They are always identical and occur roughly one in every 200 identical twins. Its incidence ranges from 1/50,000 to 1/100,000 live births with somewhat higher incidence is found in Southwest Asia and Africa.² Five types of conjoined twin are reported that is Thoracopagus, Omphalopagus, Pygopagus, Ischiopagus and Craniopagus.³ There is no association of conjoined twin with maternal age, race, parity, or heredity and the risk of recurrence is negligible.⁴ For an obstetrician and paediatric surgeon, it is a big challenge. The incidence of female conjoined twin is three times more than a male, but the reason is still unknown.⁵ Early diagnosis of conjoined twins helps

in counselling of parents regarding different management options available like termination of pregnancy or continuation of pregnancy with post-natal surgery. Ultrasound plays a very crucial role in diagnosis of conjoined twin.² There are various clues that include unusually close fetal apposition, spinal extension, and a single heart that leads to suspicion of conjoined twin.⁶ We present a case of Dicephalus parapagus diagnosed prenatally by ultrasound at 13 weeks of gestation and confirmed after termination of pregnancy.

CASE REPORT

A 27 year old primigravida referred to our institute at 13 weeks of gestation with ultra-sonographic diagnosis of conjoined twin (Dicephalus). She had regular menstrual cycle prior to spontaneous conception. There was no history of twinning in the family or exposure to any ovulation inducing drugs. She had one ultrasound report

of 6 weeks of gestation performed in one of the peripheral centres, suggestive of single live intrauterine pregnancy. The early anomaly scan at 13.3 weeks was suggestive of conjoined twin with two heads and a single fused body, three hands, two legs, a single stomach, pelvis and bladder, two kidneys and fused multiple chambered heart. There was a single umbilical cord and placenta, which was localized anteriorly (Figure 1).



Figure 1: Ultrasound image showing the conjoined twin with two heads and a single fused body.

Based on this ultra-sonographic picture, diagnosis of dicephalic parapagus tribrachius bipus conjoined twins was made. We informed and counselled the parents regarding the nature of congenital malformation and the poor chance for survival. Patient chose the option of medical termination of pregnancy. After 48 hours of induction patient aborted a dead female conjoined twin with two heads, a single fused body, three hands, two legs with single umbilical cord and placenta (Figure 2). The abortus sent for the autopsy, which further confirmed the finding of ultrasound.



Figure 2: Gross specimen of conjoined twin with two heads, a single fused body, three hands, two legs and a single umbilical cord with placenta.

DISCUSSION

Double monsters or Siamese twins is a mystery and subject of curiosity among the public. Conjoined twins are frequently the mirror image of each other. It is a rare type of monozygotic twin, which occurs when there is faulty division of embryo at 13-15 days.⁷ In literature, two theories exist which explains the origin of conjoined twins. One is theory of fission, in which, the fertilised egg splits partially.

Due to delayed separation of the embryonic mass after 12 days of fertilization, conjoined twin occurs. The other is theory of fusion, in which fertilized eggs completely separate but the stem cells (which search for similar cells) find like stem cells on the other twin and fuse the twin together. A second theory is more widely accepted.¹

Conjoined twins are of five types that is Thoracopagus, Omphalopagus, Pyopagus, Ischiopagus and Craniopagus. Out of these five, the commonest type is thoracopagus with the reported incidence of 75% and second commonest is thoracopagus which account approximately 18.5%.⁸ The least common type is omphalopagus with an incidence of 0.5%.⁷ Based on fusion, we further classify them into three groups (Spencer et al) i.e.

- Twin with dorsal fusion
- Twin with ventral fusion and
- Twin with lateral fusion. Ventral union group includes Cephalopagus (fused with head), Thoracopagus (fused with chest), Omphalopagus (fused with umbilicus) and the last one is Ischiopagus (fused together with hips).

Dorsal union twins are of three types that is Pyopagus (Sacrum), Rachipagus (spine) and Craniopagus (cranium). Only one type of twin mentioned in the third type with lateral fusion is Parapagus (fused by side).⁹

In one of the case series of 74 cases of conjoined reported by Chih-Ping Chen et al. the incidence of parapagus conjoined twins was found to be 13.5% establishing the rarity of this condition.⁴ The prognosis of conjoined twins is poor. Around 40% are stillborn and approximately 35% die within 24 hours of delivery.⁷ The overall survival rate of conjoined twin is only 25%.² Early diagnosis of conjoined twin helps in counselling of parents regarding different management options available like termination of pregnancy or continuation of pregnancy with post-natal surgery. Proper Consultation with a paediatric surgeon facilitates parental decision making regarding actual scenario.¹⁰

Successful surgical separation of conjoined twin is a complicated procedure, which is only possible when twin does not share the vital organ. Literature reveals that in one of the study of 14 cases of prenatally diagnosed conjoint twins, 28% died in utero, 54% immediately after birth, and only 18% survived out of which 50% died

postoperatively.^{1,11} One of the famous conjoined pair named Chang and Eng Bunker lived unseparated for 63 years; they married to two sisters and had 21 children.⁵ However survival rate in conjoined twin is still very less. Conjoined twins are rare, but this complication should always to be there in mind when monochorionic twin is diagnosed during the early anomaly scan that is 11-14 weeks.

Conjoined twin can be diagnosed when any one of the given finding is observed during ultrasonography that is inability to find separation between the anatomical parts or when there is no change in fetal position after movement of mother or after manual manipulations, when fetal heads lie in same plane or unusual flexion of cervical spine.²

The obstetrician plays a very indispensable role in prenatal diagnosis, counselling and organization of interdisciplinary medical care of conjoined twin. In our case, in earliest scan diagnosis was missed, which is very unusual but luckily conjoined twin was picked in early anomaly scan and timely management could be done.

CONCLUSION

This case Highlights the morphological features of the dicephalic parapagus tribrachius bipus conjoined twin. It emphasises the early prenatal diagnosis of conjoined twins, which plays the very crucial role in the management and allows appropriate and timely counselling of couple regarding early termination of pregnancy.

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