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CASE REPORT

A Rare Case of Conjoined Twin: Deradelphus with Review of Existing Literature

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Abstract:
Occurrence of conjoined twins is rare but severe complication of multigestational pregnancies. Many conjoined twin pregnancies are diagnosed before reaching viability. We present a case report of a rare variety of conjoined twins i.e. cephalothoracopagus monster also called deradelphus. It was delivered to a multigravid woman prematurely at 28wks of gestation by normal vaginal delivery and was a stillborn. It was not diagnosed antenatally because of the lack of prenatal follow up. Conjoined fetuses can be detected on prenatal screening and ultrasound. This case reinforces the importance of prenatal follow up.

Keywords: *Conjoined twin, Cephalothoracopagus, Deradelphus, Prenatal screening*
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Introduction

Conjoined fetus is a rare but challenging congenital anomaly for physicians and families. They are rare with an incidence of 1 in 1,00,000 to 2,00,000 births (1). The frequency is reported as being 1/14 000 births in India and Africa and 1/250 000 live births in Europe and the USA, suggesting an increased incidence in black populations (2, 3). They are always monozygotic and are more common in females (3:1). About 40 to 60 % are stillborn and another 35% die within 24 hours after the delivery [4]. These twins can be conjoined at any number of physical points and classified accordingly - Thoracopagus, omphalopagus, pyopagus, craniopagus, Ischiopagus. Occurrence of conjoined twins (CTs) with a chromosomal abnormality like cephalothoracopagus or cephalothoracoomphalopagus is a very rare event (3, 5).

We hereby report a rare case cephalothoracoomphalopagus monster also called deralphus.

Case report

A 23 years old Muslim multigravid woman (para 2 & gravida 4) was admitted in the Department of Obstetrics and Gynecology of our hospital with history of amenorrhea for last 7 months, polyhydraminos and absent fetal movement for last 2 days. There was no significant past medical history.

Her date of last menstrual period (L.M.P.) was

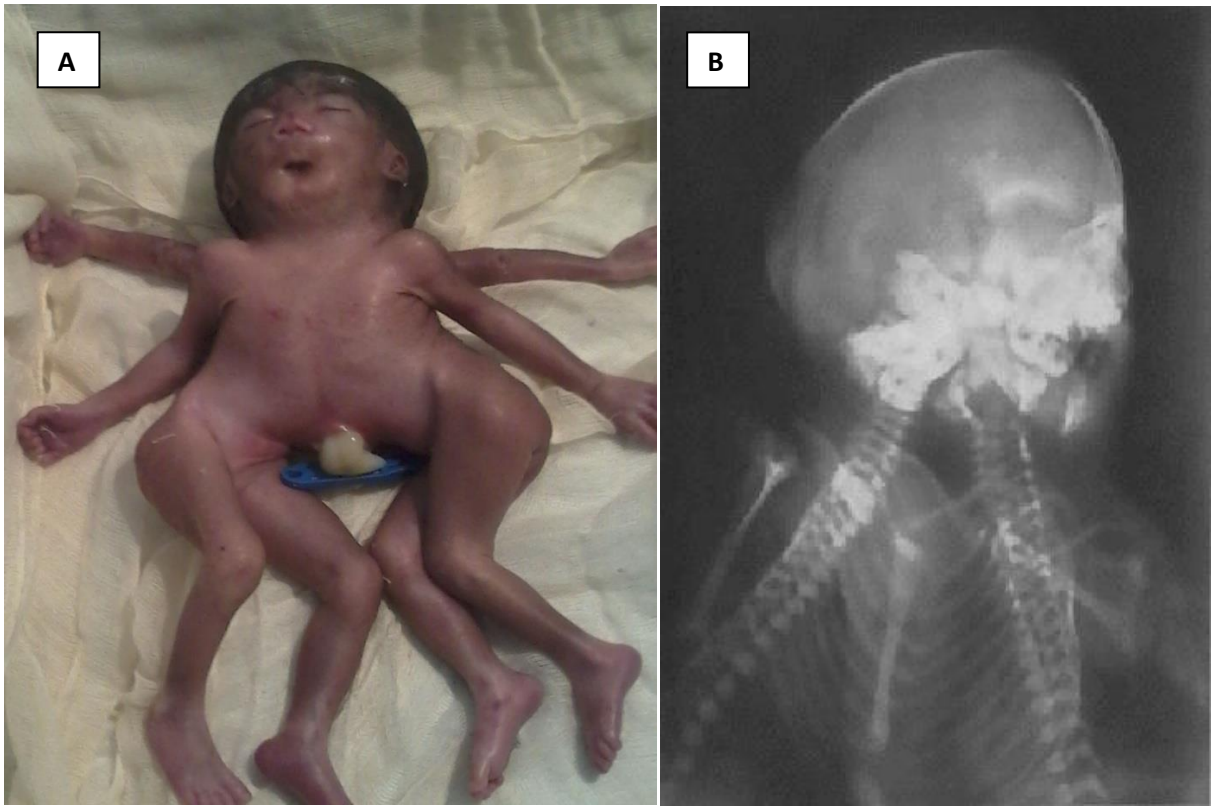

Figure 1. (A) Cephalothoracopagus monster with single head, face, neck, thorax and abdomen with single umbilicus and separate upper, lower limbs with separate pelvis and genitals. (B) The x-ray of the monster showed the two spines joined to the two basi-occiputs in a single skull.

not known. Past menstrual history was regular. She has not gone for any antenatal checkup anywhere or any ultrasonographic evaluation before admission. There was no history of intake of any ovulation induction drugs nor any assist reproductive technique. She was not exposed to any teratogen in first trimester & also didn't take any contraceptive. There was no family history of multiple pregnancy or congenital malformation. Physical examination of the mother was normal except polyhydraminos.

The Ultrasonography (USG) was done before delivery which showed doubtful fetal cardiac activity with four pair of limbs with single cord and single placenta. The delivered was performed by normal vaginal delivery but it was a still-born. Total weight of the twins was 2.3 kg.

On examination, it was seen that the twins were joined from the head down to thorax with one head, one face, one neck, a single thorax and abdomen, and a single umbilical cord. On the face there were two eyes, one nose and two ears. On the thorax, at the front and back, there were existing total four nipples, being two on either side and four upper limbs and single spine. They were separate from pelvis with four lower limbs and two female genitals (Figure-1A).

Photographs were taken, body x-ray of the monster baby was done and tried to obtain consent for autopsy but relatives refused and went away with the dead monster. The x-ray of the monster showed the two spines were joined to the two basi-occiputs in a single skull (Figure-1B).

Discussion

Conjoint twins occur due to incomplete division of a monozygotic embryo at 13-15 days post ovulation i.e. after the embryonic disc and the rudimentary amnionic sac has formed. The precise etiology of conjoined twinning is unknown. The most common explanation is fission of single zygote, or alternatively fusion of

two dizygotic or monozygotic embryos in their very early embryonic development between 13 and 15 days after conception (6). Because conjoined twins develop after differentiation of the chorion and amnion, all conjoined twins are monochorionic-monoamniotic (7, 8). The exact frequency of conjoined twins is not established and estimated incidence varies in the literatures. Spontaneous twinning occurs in 1.6% of all human pregnancies, of which 1.2% are dizygotic and 0.4% are monozygotic [6]. If we see the incidence of various types, these are as follows - thoracoomphalopagus (28%), thoracopagus (18.5%), omphalopagus (10%), craniopagus (6%) and cephalopagus (5%) while cephalo-thoracopagus and cephalo-thoraco-omphalopagus are extremely rare (3, 7, 8).

CTs are classified according to side of union and can be a major prognostic factor. Ventral unions (87%), which can occur rostrally (48%), laterally (28%) and caudally (11%), are the most frequent form. Dorsal unions (13%) are rare (9). Depending on the severity of visceral malformation, survival of CTs is extremely low (10).

First trimester ultrasound screening is useful for early diagnosis of CTs. In our case, the patient did not have prenatal follow up. If two fetuses cannot be visualized clearly, conjoined twinning must be suspected. Umbilical cord anomalies, fetal motion abnormalities and increased nuchal translucency are ultrasound findings which could be helpful in the diagnosis of CTs (11, 12). Polyhydraminos is seen in 50 to 76 % of cases.

We looked for similar cases in literature and found a very similar case described by Turgut et al. in 1998 which is probably the first reported case of deradelphus where they also described the neropathologic features on autopsy (3). All the previously reported cases were thoracopagusbicephalus.

In 2005 Vimercati et al. described prenatal evaluation of cephalo-thoracopagus monosymmetros janiceps twins where they diagnosed it at 18 wks of gestation and terminated the pregnancy (13). Kokcu et al. presented a case of male cephalo-thoraco-omphalopagus in 2007 and they also described neropathologic changes on autopsy (14). In 2014 Koreti et al. reported that a case of Cephalothoracoomphalopagus where they also noticed poor outcome (6).

Prognosis is very poor among conjoined twins. In a study of 14 cases of prenatally diagnosed conjoined twins, 28% of cases died in utero, 54% died immediately after birth, and only 18% survived out of which 50% died postoperatively (7, 8, 15). In another study the prognosis was unfavorable, with approximately 40% of cases stillborn. The prognosis of cephalothoracopagus is extremely poor because single brain and heart are present with fused gastrointestinal tracts (8, 15).

Overall, the prognosis depends on the type of fusion and presence of associated structural defects. In early pregnancy, the parents can opt for a medical or surgical termination. After 18-20 weeks, transvaginal termination will become difficult and the delivery could require a major surgical procedure, i.e., hysterotomy or classical cesarean section (11).

Conclusion

Twinning is a teratogenic event and conjoint twins may have discordant structural anomalies that further complicate decisions about whether or not continue the pregnancy. The prognosis is very often lethal due to the major conjointment, as occurred in the present report. However early prenatal diagnosis, assessment of shared vital organs, multidisciplinary approach, including neurosurgeon, pediatric surgeon as well as obstetrician and pediatrician are essential for optimal obstetric and postnatal management. Although Cesarean section is the delivery method of choice for all CTs to maximize survival and prevent birth trauma, vaginal delivery may be possible in cases for which survival is impossible, as occurred in our case.

References

1. Pant L, Devi IG, Sinha RK. Congenital anomaly similar to craniopagus parasiticus. Asian J of Obs & Gynae Practice. 2007; 11: 42-44.
2. Sills ES, Vrbikova J, Kastratovic-Kotlica B. Conjoined twins, conception, pregnancy, and delivery: A reproductive history of the pygopagus Blazek sisters. Am J Obstet Gynecol. 2001; 185: 1396-1402.
3. Turgut F, Turgut M, Başaloglu H, Başaloglu HK, Haberal A. Extremely rare type of conjoined twins: Cephalothoracopagus deradelphus. Eur J Obstet Gynecol Reprod Biol. 1998;80:191–4.
4. Sharma N, Singh I, Sharma A. An A Rare Case of Syncephalus Thoracopagus Monsterita Sharma. JK Science. 2012;14:93-94.
5. Vural F, Vural B. First trimester diagnosis of dicephalic parapagus conjoined twins via transvaginal ultrasonography. J Clin Ultrasound. 2005;33:364–6.
6. Koreti S, Prasad N, Patell GS. Cephalothoracoomphalopagus: a rare type of conjoined twin. J Clin Neonatol. 2014;3(1):47-8.
7. Graham GM, 3rd, Gaddipati S. Diagnosis and management of obstetrical complications unique to multiple gestations. Semin Perinatol. 2005;29:282–95.
8. Stone JL, Goodrich JT. The craniopagus malformation: Classification and implications for surgical separation. Brain. 2006;129:1084–95.
9. Spencer R. Anatomic description of conjoined twins: a plea for standardized terminology. J Pediatr Surg. 1996; 31: 941-4.
10. Mackenzie TC, Crombleholme TM, Johnson MP, et al. The natural history of prenatally diagnosed conjoined twins. J Pediatr Surg. 2002;37: 303-9.
11. Pajkrt E, Jauniaux E. First-trimester diagnosis of conjoined twins. Prenat Diagn. 2005;25:820-826.
12. Sebire NJ, Souka A, Skentou H, Geerts L, Nicolaides KH. First trimester diagnosis of monoamniotic twin pregnancies. Ultrasound Obstet Gynecol. 2000; 16: 223-5.
13. Vimercati, A, Scioscia, M, Vicino, M, Selvaggi, L. Prenatal evaluation of an extremely rare and bizarre case of cephalothoracopagus monosymmetros janiceps twins . Scopus preview. 2005; 65:981-984.
14. Kokcu A, Cetinkaya MB, Aydin O, Tosun M. Conjoined twins: historical perspective and report of a case. J Matern Fetal Neonatal Med. 2007;20:349-56.
15. Hill LM. The sonographic detection oh early first trimester conjoined twins. Prenat Diagn. 1997;17:961–3.