Case Report

Undiagnosed Parapagus Dicephlus Conjoined Twins: A Case of Assisted Vaginal Breech Delivery

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Abstract

Background: Parapagus (anterolaterally joined at the chest, abdomen, and pelvis) dicephalus (two-headed) twins are rare types of conjoined twins. Vaginal delivery of undiagnosed conjoined twins are a rare occurrence and associated with Dystocia, maternal and fetal trauma. We aimed to share this rare form of conjoined twin delivered vaginally and the need for obstetric ultrasound for early diagnosis and management.

Case Presentation: We reported a 22 years old Gravid -2, Para-1 (alive, vaginal delivery) mother who does not recall the first day of last normal menstrual period but claimed to be amenorrheic for the last 9 months. She had no obstetric ultrasound during her follow-up. She was referred from a nearby health center to Adama Hospital Medical College with the diagnosis of breech presentation + active first stage of labor + intrauterine fetal death. Upon arrival, she was in the second stage of labor, and the fetus was delivered to the level of the umbilicus, up on pushing the whole trunk delivered to the level of the neck. With gentle down traction and suprapubic pressure, two fetal heads were delivered. A 2900-grama 2900-gram male stillborn had two heads, two necks, two upper and lower extremities, and a single trunk. The mother had no significant vaginal bleeding and no genital tract laceration. She was discharged on her 3rd postpartum day.

Conclusions: All pregnant women should have at least one obstetric ultrasound during their antenatal care follow-up to make the diagnosis of conjoined twins at an earlier gestational age, which is very important to guide the subsequent management. [*Ethiop. J. Health Dev.* 2024; 38(3): 00-00]

Keywords: Conjoined Twins, Parapagus Dicephalus, Case Report.

Background

Conjoined twins are one of the rarest complications of monozygotic (MZ) twinning. Conjoined twins occur as a result of incomplete division of a single embryo after fertilization (1). They have monochorionic monoamniotic placentation with fused fetal parts and are estimated to occur in 1.5 per 100,000 births worldwide (2). It is more common in females than males, with a reported female/male ratio of 2:1 or 3:1 (1). Conjoined twins are classified based on the site of union (1, 3). Parapagus (anterolaterally joined at the chest, abdomen, and pelvis) dicephalus (two-headed) twins are rare types of conjoined twins. It account for only 11-13% of all conjoined twins, and they rarely survive (4). Imaging plays a crucial role in the prenatal diagnosis of conjoined twins, with the initial diagnostic modality being ultrasound (5). Since it avoids the risk of dystocia and maternal and/or fetal trauma; cesarean section is preferred mode of delivery for viable conjoined twins (3). Successful vaginal delivery of undiagnosed conjoined twins has been reported, but there is a high risk of dystocia and maternal and/or fetal trauma, including uterine rupture, cervical laceration, and fetal death (4, 6, 7).

We reported Assisted Vaginal Breech Delivery of undiagnosed Parapagus dicephalic conjoined twins with no maternal genital tract lacerations. It emphasizes the need for obstetric ultrasound during antenatal care follow-up for successful in-utero diagnosis of conjoined twins, which is crucial to guide obstetric management. Relevant literature was also reviewed.

Case Presentation

A 22 years old Gravid -2, Para-1 (alive, singleton vaginal delivery) a mother who does not recall the first day of her last normal menstrual period but claimed to be amenorrheic for the last 9 months. She had no early milestones at hand. She had antenatal care (ANC) follow-up at a nearby health center. She had no obstetric ultrasound scan during the follow-up because of a lack of access to obstetric ultrasound at the health center. She was referred to our hospital from that health center with the diagnosis of breech presentation + active first stage of labor + intrauterine fetal death. She had pushed down the pain of 14 hours duration and leakage of liquor of 10 hours. The pregnancy was planned, wanted, and supported. She had no personal or family history of twinning. She had no history of exposure to ovulation induction drugs. She had no known chronic medical or surgical illness.

Up on physical examination, she was in labor pain. Her vital signs were: Blood pressure (BP) 110/70 mmHg, Pulse rate 102 beats per minute, Respiratory Rate 22 breaths per minute, and Temperature 36.8 ^{o}c. Pertinent findings were in the abdomen; it was a 30-weeks sized gravid uterus, longitudinal lie, breech presentation, and the fetal heartbeat was negative. She had 3 contractions in 10 minutes, each lasting 40 to 60 seconds. Upon pelvic examination, the cervix was fully dilated, with no active vaginal bleeding both lower extremities of the fetus were delivered to the level of the umbilicus, up on pushing the whole trunk, and delivered to the level of the neck. Non-pulsating single umbilical cord was also noted. Since the delivery was imminent, obstetric ultrasound was not done. Her laboratory

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studies gave the following results: Hemoglobin - 13.1g/dl, White blood cell count- 13,400 /$mm^{3,}$ Platelet count- 185,000 /$mm^{3,}$ and Blood group and Rh- O^{+}.
With the assessment of 3rd Trimester Pregnancy + Unknown Date + Breech presentation + Second Stage of Labor + Intrauterine Fetal Death (IUFD), the woman was informed about the condition, the IV line was secured, started on DNS, and the bladder was deflated with a urinary catheter. Gentle down traction and suprapubic pressure were applied simultaneously with maternal bringing down effort to effect the delivery of the two fetal heads. The third stage of labor was managed actively. Up on genital exploration, there was no active bleeding, and there was no laceration on the genitalia. Examination of the stillborn reveals a freshly dead 2900-gram male stillborn with two heads, two necks, two well-formed upper extremities, a single trunk, and two lower extremities (figure -1 and 2). There was a single placenta with no dividing membrane. The woman stayed 2 days in the maternity ward and had a smooth postpartum course. She was discharged on the 3rd postpartum day with advice on breast care and the need for preconception, and early antenatal care follow-up.

Discussion and Conclusion

Conjoined twins are also known as Siamese twins, named after the birthplace of the original Siamese twins in Thailand called, Siam, in 1811 (3, 5). There are two main theories regarding the embryology of conjoined twins. The most commonly accepted theory is known as the fission theory, which states that conjoined twins occur as a result of incomplete division of a single embryonic disc, which occurs 13 to 15 days after the ovum is fertilized. The second theory is that conjoined twins occur as a result of a secondary fusion of the two completely divided embryonic discs as the embryos enlarge, which is mainly due to their unusual proximity. This is known as the fusion theory (8, 9).

Conjoined twins are classified based on the site of union (1, 3). This classification includes thoracopagus (union at the chest), omphalopagus (fusion at the anterior abdominal wall), pygopagus (the buttocks), ischiopagus (the ischium), and cephalopagus (the head). The most common is the thoracopagus, followed by omphalopagus(1, 3). Similar to most other types of conjoined twins, parapagus dicephalus conjoined twins have significant problems in their hearts, lungs, and abdominal organs, mainly because of complex malformations of these organs. In addition, they have also duplication of the upper gastrointestinal tract, tracheae, and spinal column have been reported (10).
Prenatal diagnosis of conjoined twins is mainly by imaging modalities, ultrasound being the initial diagnostic modality, followed by computed tomography (CT) and/ or magnetic resonance imaging (MRI) (5). Obstetric u The diagnosis should be suspected based on the first-trimester ultrasound finding of monoamniotic twin pregnancies and when the embryonic/fetal poles are closely associated and do not change in position with respect to each other (11). Other ultrasound findings, which are not specific to all conjoined twins, include increased nuchal translucency, a bifid fetal pole, fetal hyperextension, fewer limbs than expected, a single umbilical cord with more than three vessels, or both heads or breeches consistently at the same level to each other (12).

Perinatal mortality of conjoined twins is generally high. The survival rate of conjoined twins reported from two case series was 18% among 14 sets of conjoined twins managed between 1996 and 2002 (13) and 13.6 % among 36 cases reported from a single center in Brazil between 1998 and 2010. (14) Among 14 cases of conjoined twins managed at the Children's Hospital of Philadelphia, 28% end up in intrauterine fetal death, and 54% end up in early neonatal death. (13) In the other study, the risk of stillbirth among conjoined twins reaches 40–60%, and about 35% of the live births die within the first 24 hours (5).

Evidence on the management of conjoined twins are largely based on data from case reports, small case series, and expert opinion. If the diagnosis of conjoined twins is confirmed before viability, pregnancy termination should be offered (1). If the patient desires to continue the pregnancy, she should be counseled about the prognosis for survival and successful postnatal separation depending on the degree of vascular and organ sharing between the two fetuses(1, 3).

Since it avoids the risk of dystocia and maternal and/or fetal trauma, cesarean section is a preferred mode of delivery for viable conjoined twins (3). Vaginal delivery of undiagnosed conjoined twins was associated with a high risk of dystocia and maternal and/or fetal trauma, including uterine rupture, cervical laceration, and fetal death (4, 6). Vaginal delivery may be attempted with small, very premature, or nonviable fetuses or for pregnancy termination before 24 weeks of gestation (3, 15). Maneuvers to facilitate vaginal delivery have been described (16), but craniotomy, decapitation, evisceration, or amputation may be needed as a last resort (15, 16).

In this case, she had no personal or family history of twin pregnancy. She had no history of exposure to ovulation induction medications. Even though conjoined twins are more common in females, our index case was male (Figure 2). Since she had no obstetrics ultrasound during antenatal care, follow-up, conjoined twins were not diagnosed until delivery. Vaginal delivery of undiagnosed conjoined twins increases the risk of dystocia, maternal and fetal trauma, including uterine rupture and fetal death. Forty–sixty percent of conjoined twins end up in stillbirth; this is mainly because of the congenital anomalies which are always present in conjoined twins. She gave birth by assisted vaginal breech delivery with no dystocia and maternal genital tract lacerations. These congenital anomalies often preclude the survival of one or both twins, even if surgical separation is performed. Intrauterine fetal death or early neonatal

death of conjoined twins can also be caused by the dystocia, asphyxia, or trauma that occurs during the delivery process.

In conclusion, vaginal delivery of conjoined twins is rare and increases the risk of dystocia, and maternal and fetal injury, including uterine rupture and fetal death. Intrauterine diagnosis of conjoined twins, preferably at an earlier gestational age, is very important to guide obstetric management and to improve maternal and fetal outcomes. Obstetrics ultrasound should be done at least once during antenatal care follow-up for all pregnant women in order to pick such types of obstetrics complications.

Abbreviations

AHMC: Adama Hospital Medical College, ANC: Antenatal Care, CT: Computer tomography, IUFD: Intrauterine Fetal Death, MRI: Magnetic Resonance Imaging, MZ: Mono-zygotic.

Declarations

Ethics approval and consent to participate

The patient has given her written consent to participate in this case report. Ethical clearance was obtained from the Institutional Ethics Review Board of Adama Hospital Medical College.

Consent for publication

For publication of this case report and accompanying images, we obtained written informed consent from the patient.

Availability of data and materials

All data generated in the preparation of this document are included in the submitted manuscript and the datasets used in this case report are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

The corresponding author develops the concept of the case report. DT, BA, and HA were a member of the managing team involved in the management and follow up of the patient. The involvement made drafting, execution, and interpretation of data of all authors. In addition, all authors were involved in reviewing different literature and critically reviewing the article before submission. All authors read and gave final approval of the submitted document. Finally, all authors agreed to take responsibility and be accountable for the contents of the article.

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Figure Legends

- Figure 1- Postnatal image of Parapagus Dicephalus Conjoined twins
- Figure 2- Postnatal image of Parapagus Dicephalus Conjoined twins with a single placenta with no dividing membrane

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