Case report

Thoraco-omphalopagus conjoined twin: A rare case report

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ABSTRACT

Introduction and importance: Conjoined twins represent a rare phenomenon and the etiology has not been clarified yet. There is a high rate of stillbirth and neonatal deaths resulting in very few cases surviving long enough for surgical separation.

Case presentation: A 33-year-old gravida 2 para 1 mother without any first and second trimester antenatal care visits was diagnosed to have conjoined twins in the third trimester. Mother and her family chose to terminate the pregnancy for which elective lower section cesarean section was done with the delivery of female conjoined twins, both of them subsequently declared dead within 4 h of birth.

Clinical discussion: A conjoined twin gestation provides inimitable intricacy for obstetric management irrespective of the patient's areas of care. Early diagnosis through ultrasonography can be done and detailed evaluation is necessary along with fetal echocardiography regardless of site of fusion. Cesarean section is the recommended mode of delivery as this reduces various complications.

Conclusion: The obstetricians' role in timely prenatal diagnosis, counseling, and organization of interdisciplinary medical care is indispensable in cases of conjoined twins.

1. Introduction

Conjoined twins represent an extremely rare impediment of monozygotic twinning and are infrequently encountered by obstetricians with an incidence of 1 in 100,000 to 1 in 250,000 live births, and are more common in non-Caucasian populations [1]. Due to miscarriage or termination of pregnancy, many of these pregnancies do not achieve a viable gestational age [2–4]. Expeditionous diagnosis and treatment are warranted because roughly 70 % of conjoined twins expire within 48 h of birth or have a lethal congenital alteration [5]. Due to the high rates of stillbirth and neonatal demise, only 6–8 sets of conjoined twins survive to surgical separation each year [4].

Here, we report a case of conjoined thoraco-omphalopagus diagnosed at 34 + 1 weeks period of gestation (POG) with no prior antenatal visits. This case has been reported in line with SCARE criteria [6].

2. Case presentation

A 33-year-old G2P1L1 with no previous congenital anomalous birth was referred to our center at 34 + 1-week POG for further management of conjoined twins diagnosed three days back during her first antenatal ultrasonography. She never had any antenatal care (ANC) and neither had any dating scan or anomaly scan done during this and her previous pregnancy. She also didn’t take folic acid during her first trimester. She had a spontaneous delivery of male child 17 years back at 39 weeks POG at home. In addition, there was no history of intake of any other drugs or alternative medicine, radiation exposure, or history of fever during the first trimester of her pregnancy.

On examination, she was conscious with a blood pressure of 110/70 mmHg, respiratory rate of 18/min, and heart rate of 80 beats per minute. On abdominal examination, the uterus was of term size, and relaxed; multiple fetal parts were palpable and the fetal heart rate of twin 1 and twin 2 was detected to be 147 and 148 beats per minute respectively. A vaginal examination was not performed. Ultrasonography showed a twin pregnancy with twin 1 at 30–31 weeks period of gestation with a fetal heart rate of 136 beats per minute, and cephalic presentation; twin 2 at 29–30 weeks period of gestation with a fetal heart rate of 136 beats per minute and breech presentation. A single placenta was noted in the anterior upper uterine segment. Both fetal hearts were fused with a common pericardial sac and the 1st twin had a dilated heart. Moreover, the superior aspect of the liver of both fetuses was fused; features suggestive of conjoined twins likely thoraco-
omphalopagus. With this diagnosis, the patient party was counseled about the condition, outcome, and prognosis of the twins. The parents were informed of the malformation and the likely outcome if the twins survived after delivery. The parents decided to terminate the pregnancy and refused further evaluation and investigations pertaining to the twins. Elective lower segment cesarean section (LSCS) was done for conjoined twins at 36 + 1 weeks period of gestation. The first twin presented by cephalic, conjoined by the thoracic region, and the second twin presented by breech and was delivered by breech extraction as shown in Fig. 1. The combined birth weight of twins was 3.22 kg with Apgar scores of first twin 3/10, 6/10, and second twin 3/10, 4/10, and both were of the female sex. There were no other gross congenital anomalies, and the placenta was located in the anterior upper uterine segment and was mono-chorionic and mono-amniotic. Bilateral uterine tubes and ovaries were normal-looking.

After delivery both the twins were handed over to the pediatrician and were shifted to the neonatal unit. However, 2 h after delivery, twin 1 developed gasping, and was kept under the head-box but was declared dead after half-hour and twin 2 was declared dead one and half hours after the death of twin 1 owing to respiratory and cardiovascular compromise. The maternal postoperative period was uneventful and was discharged on the 3rd postoperative day with appropriate compromise. The maternal postoperative period was uneventful and was discharged on the 3rd postoperative day with appropriate counseling.

3. Discussion

Three major groups of conjoined twins (CTs) are known: twins with a ventral union, twins with a dorsal union, and twins with a lateral union [2,7]. Within these groups, based on the most eminent position of conjunction, conjoined twins are classified as the skull (cephalopagus), thorax (thoracopagus), abdomen (omphalopagus), pelvis (ischiopagus), sacrum (pygopagus), and back (rachipagus) [7]. Ventral fusion is the commonest type of conjoined twinning with a wavering span of seriousness commencing from the thorax above down to the pelvis and the fetal chest is the most common fusion site, making the majority of conjoined twins thoracopagus [1]. This indicates that the embryonic disc splits from both cranial and caudal ends, and if there is defective division, the chest remains fused [1].

Though the exact cornerstone for CTs has not been established, there are two refuting theories. Incomplete division of a single zygote twelve to fifteen days after fertilization results in conjoined twins with fuses sharing portions of their bodies, which falls under the traditional theory of fusion [8–10]. The other theory is fusion where two originally discrete mononuclear embryonic disks undergo secondary unification resulting in conjoined twins [10–12]. Conjoined twins are always of the same sex as they are monozygotic, monochorionic, and monoamniotic at all times [13]. Some risk factors like positive history of twin delivery, use of drugs for induction of ovulation, infertility treatment, and exposure to detrimental radiation were proposed to have a likely effect on the development of this rare condition [14]. However, none of these risk factors were covered by the past, family, and pregnancy history of our case. Additionally, there is no evidence of an alliance between the development of CTs and increased maternal age, and demographic, genetic, or environmental factors [1].

Although most of the cases of stillbirths in CTs were reported in males, CTs tend to occur three times more in female fuses [10,15]. CTs in our case were also of the female sex. Overlapping of the timing of monozygotic twinning and X chromosome inactivation thus leading to the development of monozygotic twins and potential survival advantage due to XX karyotype are the proposed reasons for the observation of higher incidence of CTs among females [7].

Thoracopagus twins usually have an impoverished prognosis because of a greater incidence of cardiac anomalies and the more complex hepatic and biliary fusion [16]. These twins usually share hearts and have composite cardiac anomalies as well, which imperil the successfulness of disunion surgery [17]. 92 % of conjoined twins have cardiac defects, extracardiac anomalies (e.g., facial, abdominal wall, limb defects) occurred in 62 % of cases, a common pericardial sac is present in 90 % of thoracopagus twins, and the prevalence of conjoined hearts is 75 % [17]. The extent of the union of the hearts in thoracopagus twins varies but can include fusion of the large vessels, the atria, the atria and ventricles, and rarely, a single heart in one of the twins [17].

Early diagnosis of CTs is possible by high-resolution transvaginal sonography done in the first trimester during prenatal follow-up and confirmation can be done by a definite imaging modality such as magnetic resonance imaging (MRI) which is complementary to ultrasound and can provide more precise anatomical clinical data [18–20]. A comprehensive anatomical fetal survey at 18–20 weeks will be diagnostic for patients who did not undergo fetal ultrasonography in the first trimester [21]. Detailed evaluation by ultrasound to delineate the stretch of fusion, common viscera, and the cardiac condition is of paramount importance for prognostic counseling [21]. When CTs are detected by USG, fetal echocardiography should be done regardless of the fusion site because of increased incidence of cardiac defects in monozygotic twinning, as shared cardiac anatomy is associated with a poor prognosis [21]. In our case, the mother did not have any ANC visit as well as an ultrasound done in the first and second trimesters, so the diagnosis was made in the third trimester.

A timely finding of CTs provides immense help in the management of pregnancy and planning of delivery techniques. In the present case, the diagnosis was made in 34 + 1 week POG and the parents chose to terminate the pregnancy and did not opt for any management of the born twins. Both of the twins expired in our case and no further imaging procedures were considered. Early diagnosis of conjoined twins provides the parents an opportunity for well-informed decision-making. However, exact details about united visceral involvement of heart structures that would highly influence counseling about distant viability may not be feasible until later in gestation [22]. According to data from a single-center study, almost 50–70 % of patients select termination of

![Fig. 1. Photograph of thoraco-omphalopagus conjoined twins delivered through elective lower section cesarcean section at 36 + 1 weeks of gestation.](image_url)
pregnancy after extensive consultation [23]. Obstetric advising must traverse the values of expectant parents along with the dissemination of pragmatic and elaborative expectations regarding the postnatal journey [24]. Counseling should be commenced as soon as the diagnosis of CTs is made and images from the performed radiological investigations like ultrasound or fetal MRI can be used in order to impart fetal anatomical illustration, along with associated outcome [24]. Choices for management of pregnancy including termination of pregnancy and expectant management should be rigorously discussed [24].

The commended mode of delivery is cesarean section regardless of future neonatal care schemes for conjoined twins in the third trimester as this decreases the incidence of possible damage to shared fetal tissues, the associated danger of internal hemorrhage, and intrapartum death [4]. Conventionally a classical cesarean delivery (vertical skin incision with a vertical uterine incision) has been practiced however, in certain scenarios based on proportions of the fused fetal mass, anatomy of fetal adjuncts, fetal presentations, location of the placenta, and maternal and uterine anatomy, a Pfannenstiel skin incision followed by either classical or low transverse incision on the uterus may be pondered [4]. A Pfannenstiel skin incision and LSCS were done in our case.

4. Conclusion

A conjoined twin gestation is a rare event, which has unique complexity for obstetric management regardless of the patient’s goals of care. However, early diagnosis with a good prediction of associated anomalies would help in earlier decision-making. The obstetricians’ role in prenatal diagnosis, counseling, and organization of interdisciplinary medical care is indispensable in cases of conjoined twins.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

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Author contribution

Asmita Adhikari (AA) and Sunita Bajracharya (SB): Study Concept, editing, and surgical therapy of the patient. Diptee Poudel (DP), Suraj Shrestha (SS) and Roshan Aryal (RA): Writing- original draft preparation, editing, and reviewing. SB: Senior author and manuscript reviewer.

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