Severe twin-to-twin transfusion syndrome in a scared uterus successfully treated with serial amnioreduction

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Abstract

Background: Twin-to-twin transfusion syndrome (TTTS) is a vascular abnormality unique to monochorionic pregnancies. The associated polyhydramniosportends significant risk to uterine rupture especially in a scared uterus. Serial amnioreduction is an established technique that reduces the risks of severe polyhydramnios thereby preventing over-stretching of the uterus and ameliorates maternal symptoms. **Methods**: The case records of a patient who has had three previous caesarean sections and presented with respiratory distress following severe polyhydramnious were reviewed. **Result**: A total of 7,225mls of amniotic fluid was removed over a 3 month period; and grade II discordant intrauterine growth restriction (IUGR) was confirmed at delivery. **Conclusion**: Serial amnioreduction was successful in relieving maternal symptoms and prevented the risk of uterine rupture in a woman with three previous caesarean section scars.

Keywords: Twin-to-Twin Transfusion Syndrome, Polyhydramnios, Amnioreduction.

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Introduction

Twin-to-twin transfusion syndrome (TTTS) is a vascular abnormality unique to monochorionic pregnancies. It affects 5-17% of monochorionic pregnancies and is responsible for 15% of the overall fetal mortality in twin pregnancy. [1]The associated polyhydramnios of the recipient fetus can reach extreme levels with a significant risk to uterine rupture especially in a scared uterus. Discordant growth occurs in 15-29% of monochorionic twins, and percentage discordancy in estimated fetal weight (expressed as a percentage of the larger twin's weight) is used as an index of discordant IUGR[2]. Prenatal therapy of polyhydramnios consists of amnioreduction, septostomy, selective fetoscopic laser photocoagulation of the communicating vessels on the chorionic plate or selective foetocide by umbilical cord occlusion. Serial amnioreductionhas been shown to achieve 100% reduction in maternal dyspnea and a gain of about 3 week means gestational age[3].

Case Report

Mrs. O.S, a 36 year G_6P3^{+2} alive was referred from a private hospital at about 29 weeks of gestation with complaints of abdominal swelling/pain and difficulty in breathing for one week. She also had an early scan at about 19 weeks gestation confirming monochorionic diamniotic twin gestation. The swelling was gradual in onset, generalized and progressive. It was associated with dull aching pain that was continuous and severe enough to prevent her from sleeping. It was worsened by lying flat and relieved by analgesics. She had no vomiting or constipation, and she ate little for fear of pain. There was associated difficulty in breathing. She had no cough, chest pain or paroxysmal nocturnal dyspnea. There was no fever, urinary symptoms, vaginal bleeding or drainage of liquor. The index pregnancy was planned and spontaneous. The pregnancy was booked at 22 weeks of gestation at the referring clinic. The pregnancy was uneventful until she started experiencing the above symptoms. Her first and second pregnancies were 16 and 14 years ago respectively. They were both unplanned and she had termination of both pregnancies at eight weeks gestation with no post abortal complications. The third pregnancy was nine years ago. It was planned, she had uneventful antenatal period and the pregnancy was carried to term. She had an emergency lower segment caesarean section on account of prolonged labour with fetal distress. The baby died few hours post-delivery. Her fourth and fifth pregnancies were four and eight years ago respectively. They were planned and she had caesarean section at term in both cases. There were no puerperal complications.

She was a trader from Southeast Nigeria. She neither smoked cigarette nor drank alcohol. She was aware of contraceptives but had not used any method. There was no family history of hypertension, diabetes mellitus, and sickle cell disease. Her mother had a set of twins.

She was a young woman who weighed 93kg with a height of 169cm. She was in pains and respiratory

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distress, but not pale, afebrile, and anicteric. She had mild pedal oedema. Her respiratory rate was 18 cycles per minute, and the lung fields were clinically clear. The pulse rate was 84beats per minute with a blood pressure of 120/80mmHg. The heart sounds were normal. The abdomen was uniformly distended and shiny. There was a pfannenstiel scar that healed by primary intention. The liver, spleens and the kidneys were difficult to palpate because of the distention. The symphysio-fundal height was 45cm which was far greater than her gestational age of 29weeks and 4days. The fetal poles could not be palpated. Fetal heart tones were not appreciated by auscultation. Ultrasound scan done showed diamniotic monochorionic twin gestation. Both fetuses were active with no gross anomaly. The first twin age 28 weeks three days, while the second twin age 25 weeks four days. Both fetuses were males. There was massive polyhydramnios. A diagnosis of twin-to-twin transfusion syndrome in a patient with three previous caesarean sections was made.

The diagnoses, risk and the need for conservative management till delivery were explained to her. She consented for hospital admission. Routine investigations and fasting blood sugar were requested. She had weekly packed cell volume and ultrasound scan. Intramuscular dexamethasone 12mg 12hourly for 24 hours was administered. She was given intramuscular pentazocin 60mg as needed. Intermittent tocolysis was administered prophylactically using nifedipine. She also had tablets of folic acid 5mg daily and fesolate 200mg daily. The fetuses were monitored with the sonicaid regularly. She had serial amnioreduction after counseling. Ultrasound scan was used to select the largest pocket of amniotic fluid ventral to the fetus, and avoiding the umbilical cord and the placenta. A sterile surgical site was created with appropriate skin scrubs and sterile towels. The amniotic fluid was removed using a 50ml syringe via wide bore needle introduced through the anterior abdominal wall while lying supine with a left lateral tilt and support pillows to enhance maternal comfort. A total of 7225mls of amniotic fluid was drained. We ensured that the fluid was not removed any faster than1000ml in 30 minutes in each session. She had a successful caesarean section at 36 weeks of gestation due to worsening symptoms. Fetal discordance, which is the difference in birth weights expressed as a percentage of the larger twin's weight was 31.3% at birth, confirming Grade II discordance (Figure 1).

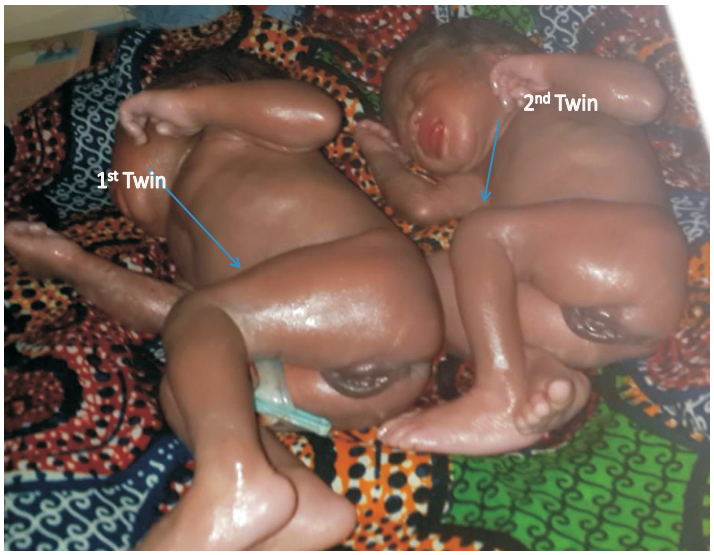

Figure 1. Discordant twins

Discussion

Chronic TTTS usually presents in the mid-trimester as seen in this case. Prematurity and the absence of anymajor fetal abnormalities on ultrasound informed the need to allow the pregnancy to progress. Serial amnioreductionis the therapeutic modality most frequently used for the treatment of TTTS. It has the advantage of being a technically simple procedure, does not require sophisticated technology or specialized centers, and can even be performed without administration of anesthetics agent. Also, it was particularly useful in alleviating the maternal discomfort and the respiratory difficulty, and more importantly the risk of uterine rupture cause by uterine over-distension. It is associated with an overall survival rate ranging from 38 to 81%[4].

However, this symptomatic therapy does not remove the etiology of TTTS. This is because as the placental anastomoses remain patent, polyhydramnios relapses and the need for serial amnioreduction. A definitive fetoscopic laser ablation therapy of placental anastomoses has a better outcome. [5]However, this invasive procedure is not available in our centre. Septostomy, the intentional perforation of the intertwin amniotic membrane in an effort to equilibrate the amniotic fluid volume of the twins has some role in early stage disease. Selective fetocide of one twin by umbilical cord ligation or bipolar coagulation under ultrasound or fetoscopic guidance is the method of selective fetocide more commonly employed when one of twins is affected by a chromosomal abnormality or a multifactorial effect[6].

The amnioreduction of 7,225ml of amniotic fluid was under careful ultrasound guidance to avoid injury to the fetuses and placenta. We also ensure that the fluid was not removed any faster than 1000ml in 30 minutes in each session to avoid risk of abrutio placenta. Corticosteroids were administered to accelerate lung maturity because of the high probability of preterm delivery; and intermittent tocolysis was administered

prophylactically to prevent uterine contractions.

The early ultrasound scan finding of one twin with a gestational age of 28 weeks three days and the other twin 25 weeks four days suggest discordant growth. This prompted the weekly ultrasound scan follow-up as discordancy of greater than 25-30% has some predictive value for poor fetal outcome in TTTS[7]. Grade II discordance of 31.3% was confirmed at delivery. The normal should be less than 20%.

In conclusion, serial amnioreduction was successful in relieving maternal symptoms and preventing risk of uterine rupture in a woman with three previous caesarean section scars.

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