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Case Report

Radiology Section

Twin to Twin Transfusion Syndrome (TTTS): An Interesting Entity in Twin Pregnancy

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ABSTRACT

Twin-Twin transfusion syndrome (TTTS) is a condition with high rates of perinatal morbidity and mortality due to its poorly understood etiology and difficulty in diagnosis and treatment. Early diagnosis during antenatal ultrasound is very important in reducing the morbidity and mortality rates. TTTS is a phenomenon exclusive to monochorionic twin pregnancies. The donor twin is characterized by oligohydramnios or anhydramnios, growth restriction and abnormal umbilical artery by Doppler velocimetry. The recipient, on the other hand, is characterized by polyhydramnios, abnormal venous dopplers, cardiac enlargement /failure, and eventually hydrops. Sonologists while evaluating the monochorionic twin should have high index of suspicion for TTTS. The present study reports a classical case of TTTS which was detected accurately by ultrasound and managed efficiently resulting in birth of healthy twins.

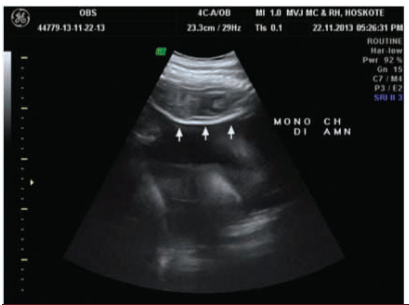
Keywords: Anaemia, Hypothermia, Intensive care, Jaundice, Low birth weight, Neonatal

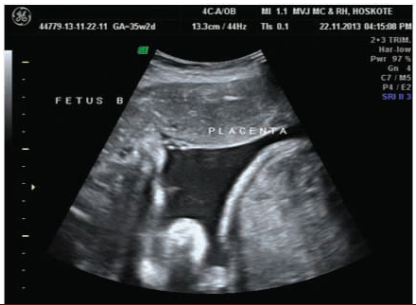
CASE HISTORY

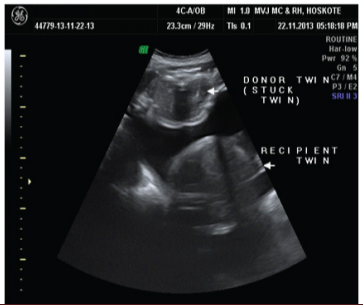
Pregnant female (G4P1L1A2) was referred to our Department of radiodiagnosis, MVJ MC and RH for antenatal sonography at 8 ½ months of amenorrhoea with previous antenatal scans done elsewhere. She had her ANC's done outside and her scan at 8wks had showed twin gestation with chorionicity not mentioned. Subsequent scan done at 16 wks was normal without any discrepancy in growth or liquor. She had no history suggestive of TTTS like abdominal distension, respiratory distress or preterm labour.

USG and MANAGEMENT

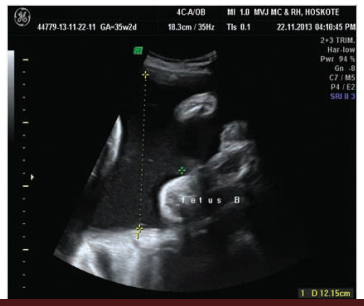
On performing sonography we found Monochorionic diamniotic [Table/Fig-1,2] twin live intrauterine gestation. Twin A (donor twin) was seen 'stuck' to the anterior uterine wall, corresponding to 29 weeks [Table/Fig-3], small for gestational age with anhydramnios [Table/Fig-4] and empty urinary bladder. Umbilical artery doppler demonstrated absent diastolic flow suggestive of moderate placentofetal insufficiency [Table/Fig-5]. Twin B (Recipient twin) demonstrated breech presentation, corresponding to 34 weeks [Table/Fig-6], polyhydramnios (Deepest Vertical Pocket -12cm) [Table/Fig-7], over distended urinary bladder [Table/Fig-8]. Doppler demonstrated normal umbilical artery flow pattern [Table/Fig-9]. Because of the above mentioned findings, diagnosis of Monochorionic diamniotic twin live intrauterine gestations with features of Twin to Twin transfusion syndrome (stage-3) with stuck donor twin was given. With adequate blood transfusion patient was taken up for emergency LSCS. Twin A was delivered first by breech extraction with excess liquor, second twin was extracted with membrane tightly wrapped around the baby. Recipient twin was a female baby weighing 2.01kg with good APGAR score. Donor twin was a

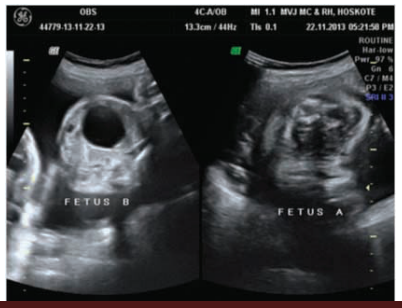


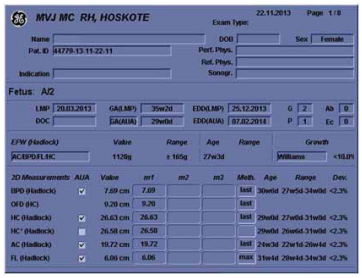




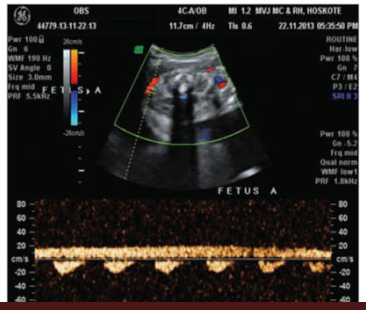
[Table/Fig-1]: Monochorionic twin gestation **[Table/Fig-2]:** Diamniotic twin gestation **[Table/Fig-3]:** Fetal biometry of donor twin demonstrating significant growth lag

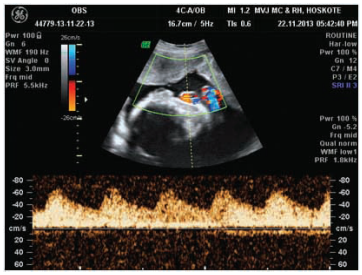






[Table/Fig-4]: Donor twin 'stuck' to the uterine wall with anhydramnios **[Table/Fig-5]:** Umblical artery doppler of donor twin demonstrating absent diastolic flow suggestive of moderate placento-fetal insufficiency **[Table/Fig-6]:** Recipient twin demonstrating growth appropriate for gestational age





[Table/Fig-7]: Recipient twin demonstrating polyhydramnios **[Table/Fig-8]:** Recipient twin demonstrating over distended urinary bladder and donor twin demonstrating empty urinary bladder **[Table/Fig-9]:** Umbilical artery doppler of recipient twin demonstrating normal flow

female baby weighing 1.3kg with good APGAR score. Placenta was single. There was difference of > 5g Hb between the twins, weight difference of 0.7kg (40 %). Blood group of the babies was 'O' Positive.

DISCUSSION

Approximately 75% of monozygotic twins are monochorionic. Twin to twin transfusion syndrome (TTTS) is a specific condition complicating 10-15% of monochorionic multiple pregnancy [1]. The natural history of severe TTTS is well established. Mortality rates approach 80 to 100 percent if left untreated, especially when it presents at less than 20 weeks gestation [2]. Vascular anastomoses are present in virtually 100% of monochorionic twin pregnancies, whereas twin-twin transfusion occurs (with rare exception) in 5-10% of monochorionic pregnancies [3,4]. The reason for the occurrence of twin-twin transfusion syndrome in only a small proportion of the monochorionic twin pregnancies with vascular anastomoses is unknown. The intraplacental anastomosis is usually situated in a single, shared cotyledon of the common placenta, and is usually arteriovenous but may be arterial-arterial anastomoses. A distinction between the acute and chronic forms of the syndrome can be made on the basis of weight discrepancy and haemodynamic changes. Infants with the chronic form have discrepancies in birth weight exceeding 15% and the peripheral blood film of the donor twin may show hypochromic microcytic anaemia and erythroblastosis [1]. TTS has been classified into five stages based on sonographic findings by Quintero RA et al., [5].

The best treatment of cases presenting before 26 weeks of gestation is fetoscopic laser ablation of the intertwin anastomoses on the chorionic plate [6]. There are no formal contraindications of laser therapy, although a short cervical length (<15 mm) may indicate a higher-risk of preterm delivery. Preliminary evidence suggests that cervical cerclage might reduce this risk. Moreover, perinatal survival rates after laser therapy were higher when the TTTS was Stage 1 or 2 than when it was Stage 3 or 4. Increasingly, therefore, attention is being paid to the timely diagnosis and treatment of TTTS [7,8]. In the absence of complications after laser treatment, planned delivery is recommended from 34 weeks and no later than 37 weeks.

Sonographic findings include

1. Monochorionic placentation
2. Estimated fetal weight discordance (>20% of larger twin's estimated weight)
3. Abnormal amniotic fluid volume - One sac with oligohydramnios, deepest vertical pocket < 2.0cm and the other sac with polyhydramnios, deepest vertical pocket > 8.0cm
4. Urinary bladder findings-Small or no bladder visualized in twin with oligohydramnios and large bladder visualized in twin with polyhydramnios
5. Appearance of a " stucktwin"
6. Hydrops fetalis-Presence of one or more of the following in either twin:

a) Skin edema (> 5mm thickness of scalp skin)

b) Pericardial effusion

c) Pleural effusion

d) Ascites.

6. Doppler findings-Abnormal Doppler S/D ratio at the umbilical cord. The absent end diastolic flow in the donor's umbilical artery accompanied by venous pulsation in the recipient's umbilical vein are usually associated with a poor prognosis.

Other sonographic findings that may prove to be of prognostic significance include - The presence of a hypertrophied, dilated heart, with absence or reversal of flow in the ductus venosus during atrial contraction. In the donor, the heart may be dilated, the bowel is hyperechogenic, and there is absent end-diastolic flow in the umbilical artery. Prognosis is dependent upon gestational age at birth, the lower the gestational age at birth, the greater the risk for long standing neurologic or pulmonary sequelae.

CONCLUSION

High degree of suspicion of TTTS should be there while performing sonography of Monochorionic twins. Timely diagnosis of twin-twin transfusion syndrome by ultrasound is crucial to initiate the treatment early. Delay in diagnosis may result in a delay in treatment and increased perinatal mortality and morbidity.

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