**INTRODUCTION**

Monoamniotic twins accounting for about 1-5% of all monozygotic conceptions are rare, with an estimated prevalence of 1 in 5000 to 1 in 25000 pregnancies. Most commonly they are females with only 25 – 35% being male pairs. These fetuses are generally considered at high risk for fetal death, neonatal morbidity and mortality due to several conditions including umbilical cord entanglement, abnormal vascular anastomoses, congenital anomalies and premature birth. Recent studies have documented that cord entanglement is uniformly present in all mono amniotic twins and is an infrequent cause of fetal demise.1

**DISCUSSION**

Embryology

Monozygotic twins originate from the fertilization and subsequent division of one egg. The stage at which division occurs determines chorionicity and amnionicity. Monochorionic mono amniotic twins arise from division of the fertilized egg 9-12 days post fertilization, ie, in the post implantation blastocyst period. It has been proposed that either a delay in implantation (Van der Auwera & D’Hooghe, 2001 Steinman 2001) or hardening of the zona (Longo 1981, Edwards et al. 1986, Derom et al. 1987) induced by the ovulatory hormones may be the contributory factors to monozygotic twinning.

Incidence and significance

Umbilical cord entanglement in monoamniotic twins is a biologically plausible phenomenon that can be demonstrated antenatally in all monoamniotic twins when systematically evaluated with ultrasound and color doppler. After commenting that cord entanglement is uniformly present in all mono amniotic twins, Dias et al has suggested that, this is an infrequent cause of fetal demise. There are only a few studies which address the causes of early and mid pregnancy loss in mono amniotic twins, which is not that rare. These losses are in large attributed to TRAP sequence, conjoined twins and major congenital anomalies.2

Diagnosis of Cord entanglement

Cord entanglement was diagnosed by 2D, color and pulse Doppler. This was evident at the initial scan as a lump of cords with two different heart rate patterns. The different methods for diagnosis are:

1) Apparent branching of the umbilical artery ( Belfort et al). Sherer et al has reported that even first trimester diagnosis of cord entanglement is possible using this sign.

2) Narrowing of the umbilical vessels involved in cord entanglement can produce Doppler flow variations like notching of the waveform.

3) Galloping heart rates, ie, the presence of two distinct arterial flow velocity waveforms can be demonstrated by insonation of the entangled cord lump. This was how we reached the diagnosis in our patient.

Placental cord insertions

Recent studies have approved that proximal cord insertions are quite common in mono chorionic monoamniotic placentas. In our case, the cord insertions were of marginal type which was different from the usual type of proximal or (para)-central insertions.

**CASE REPORT**

A 26 year old primi gravida who conceived by ovulation induction and Intra Uterine Insemination reported for first trimester screening at 12 weeks 4 days gestation at the Fetal Medicine Unit, KARE Centre Thrissur, Kerala. The diagnosis of mono chorionic mono amniotic twin pregnancy was made on first trimester ultrasound by demonstrating two fetuses with a single placenta and absence of a dividing membrane and the family was counseled. Cord entanglement was evident on the first scan and was confirmed at subsequent visits. Marginal type of cord insertion was noted. Follow up comprised of antenatal ultrasound scans including a targeted mid trimester anomaly scan. Both the babies were found to be structurally intact. The dopplers were normal and there was no evidence of twin to twin transfusion syndrome. Ultrasound at 23 weeks of gestation showed intrauterine death of both the fetuses. Postnatal morphological examination of the placenta, cords and fetuses confirmed the ultrasound findings.

**CONCLUSIONS**

We present here a case of structurally intact male mono chorionic mono amniotic twins with cord entanglement, conceived with ovulation induction and IUI, which ended up in mid trimester intra uterine demise of both babies. Cord entanglement, which is universally present in all mono amniotic twins, can be diagnosed prenatally, if searched for meticulously. Though considered as a minor cause of fetal death, cord entanglement may be regarded as one important cause of fetal demise in mid gestation.

References

2) Dias T. et al. Ultrasound Obstet Gynecol 2010
3) Rossi A. C. et al Ultrasound Obstet Gynecol 2013

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