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CASE REPORT

Conjoined Twins with Thoracoabdominopagus Anomaly in the Third Trimester

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ABSTRACT

Conjoined twinning is rare; occurrence is about 1% of monochorionic twins. Here, we report a case of 26-year-old multigravida with 29 weeks pregnancy with conjoined twins-thoracoabdominopagus. Two live female babies joined at the chest and abdomen were delivered by cesarean section. The parents refused a separation operation and despite resuscitation the twins died of cardiopulmonary arrest after 16 hours. A review of the literature suggests that early diagnosis by a combination of ultrasound and magnetic resonance imaging is essential for predicting prognosis, management and success of surgical separation. It offers an opportunity for early counseling for parents and option for termination.

Keywords: Conjoined twins, Thoracoabdominopagus, Thoracopagus, Ultrasonography, Surgical separation.

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INTRODUCTION

Conjoined twinning is very rare occurrence. It in about 1% of monochorionic twins, with an incidence ranging from 1:30,000 to 1:200,000 live births[1,2] and 1 in 650 to 900 twin deliveries. Conjoined twins are genetically identical and are of the same sex.[3] They develop from the same fertilized egg, and they share the same amniotic cavity and placenta.

In various parts of Southeast Asia and Africa, an increased incidence of 1:14,000 to 1:25,000 has been described.[4] Forty to 60% of stillborn reported to be of conjoined twins, and 35% of live births conjoined twins do not survive beyond the first 24 hours.[2] Female predominance in the order of 3:1 have been reported.[2]

Classification is made usually according to the most prominent site of connection: the thorax (thoracopagus; 30-40%), abdomen (omphalopagus; 25-30%), sacrum (pygopagus; 10-20%), pelvis (ischiopagus; 6-20%), skull (craniopagus; 2-16%), face (cephalopagus), or back (rachipagus).[2,5-7]

Vaginal delivery of dicephalic parapagus conjoined twins at 37 to 38 weeks gestational age with birth weight of 3.5 kg have been reported.[8] Here we report a case of thoracoabdominopagus conjoined twins diagnosed in the third trimester of pregnancy.

CASE REPORT

A 26-year-old, gravida 2, para 1, living 1, at 29 weeks and 3 days of gestational age, was referred at her third trimester of pregnancy, Her previous medical, surgical, and obstetrical history was unremarkable. There was no family history of twinning on maternal side, and there was no history of medication or radiation exposure. Ultrasound was never done till date.

EXAMINATION

On abdominal examination, fundal height was more than the period of amenorrhea corresponded to 34 weeks gestational age. Multiple fetal parts felt.

On vaginal examination, cervical os uneffaced, os closed. Ultrasonographic evaluation revealed: (i) A conjoined twin of the thoraco-omphalopagus subtype, (ii) a single cardia with intercommunication of atria, (iii) kyphoscoliotic deformity of spine in both twins. Doppler measurements on ultrasonography was unremarkable. Placenta was monochorionic monoamniotic.

A magnetic resonance imaging (MRI) was performed in order to confirm the diagnosis (Figs 1 to 4). Two live fetuses joined at the chest, with combined weight of 3.1 kg (Figs 5 to 8).

MANAGEMENT

A joint meeting with senior obstetricians, neonatology, pediatric and cardiac surgeons was held since the gestational age was 29 weeks. As the results of surgical

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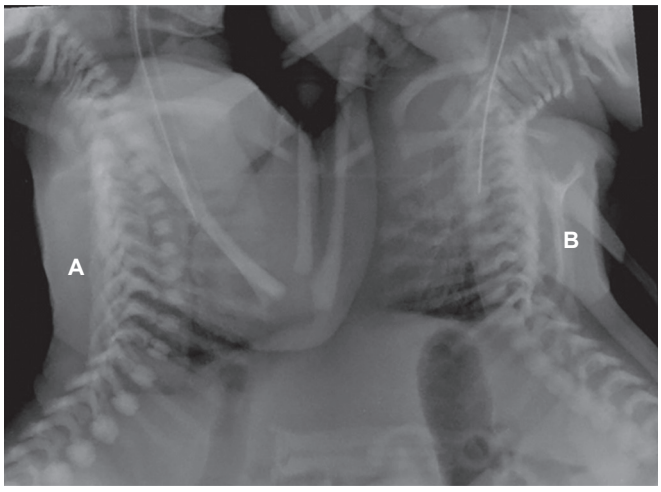

Fig. 1: Twin A is on the left lateral position while twin B is on the right lateral position

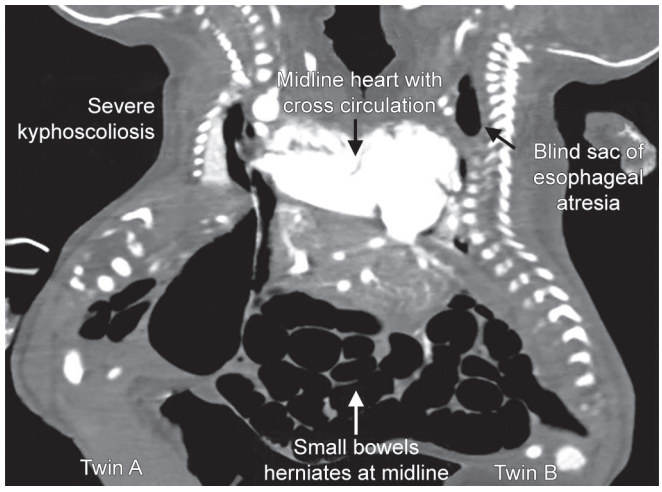

Fig. 2: Virtual reality reconstruction of twins A and B

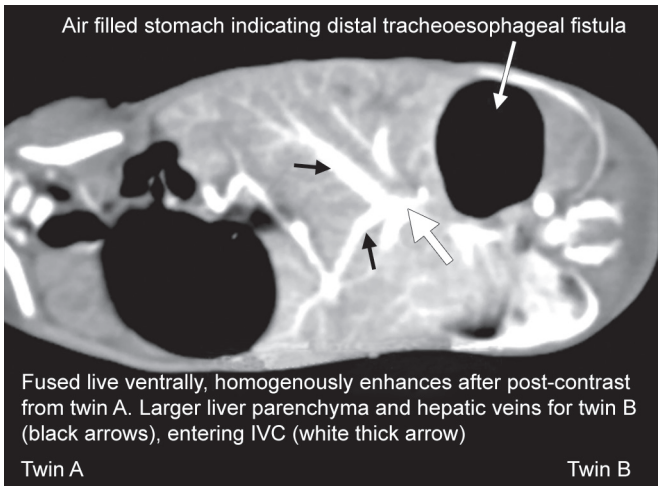

Fig. 3: Contrast-enhanced image of abdomen

Fig. 4: Three-dimensional image of the cardiac fusion in twins A and B

Fig. 5: Curvilinear uterine incision

Fig. 6: Resuscitation of twins A and B

separation would be poor due to sharing of major vital organs it was decided jointly to terminate the pregnancy, as allowing the twins to grow would only cause difficult delivery at a later date with no improvement in outcome.

Patient and her attenders were informed of the ultrasound and MRI findings the poor prognosis, the need for NICU care and resuscitation, post operative separation surgery and cost involved. The need for cesarean section to deliver the babies was also explained.

After obtaining informed consent and reserving adequate blood, patient was taken up for elective lower segment cesarean section. The conjoined girl babies

Fig. 7: Twin A (left lateral)

Fig. 8: Twin B (right lateral)

weighing 3.1 kg were delivered. Twin 1 was delivered as cephalic and had cord around the neck once. Twin 2 was delivered as breech. Both babies did not cry at birth and were intubated electively. Omphalocele was present, and both had cleft lip and cleft palate. Postoperatively, uterus was contracted and there was no postpartum hemorrhage. Patient recovered well and was discharged on 3rd postoperative day. Parents refused a separation surgery and the twins died from cardiopulmonary arrest after 16 hours.

DISCUSSION

Conjoined twins are rare, occur in monochorionic monoamniotic twins. Twinning is usually initiated once the rudimentary amniotic sac and the embryonic disk have been formed and fission of the embryonic disk is incomplete.[9]

Thoracopagus twins are usually united face to face from the upper thorax to the umbilicus, with a common upper abdominal wall, diaphragm and sternum.[10] Ninety percent common pericardium, 75% conjoined heart and liver, 50% common gastrointestinal tract.[11]

The management is mainly based on the anatomy of cardiovascular system. The success rate of separation surgery is higher where only the pericardium is shared.[12] In our case, the fetuses had shared cardia.

The survival, prognosis and feasibility of separation depends on the severity of the cardiac abnormality. Separation sugery for conjoined twins with ventricular conjunction have not been successful.[10]

The transvaginal ultrasound and MRI are very much useful in diagnosing, a conjoined twin even at 8 weeks of gestational age.[13] Early diagnosis of conjoined twins is the most important factor for the management of pregnancy.

Criteria for diagnosing conjoined twins include: (i) bifid appearance of the fetal pole in the first trimester, (ii) more than three vessels in the umbilical cord, (iii) conjoined body parts, (iv) absent separating membrane, (v) inseparable bodies or heads despite changes in fetal position, (vi) complex structural anomalies, (vii) hyperextended spine, heads or bodies at the same level, (viii) unusual proximity of the extremities, (ix) persistence of the relative positions after movement or at the follow-up scan.[7] Approximately, 50 to 76% of cases found to have polyhydramnios, but not in the first trimester.[9] The antenatal diagnosis of conjoined twins using 3D ultrasonography has also been reported.[14,15]

To predict the prognosis of the fetuses detailed evaluation of the degree of union and number of shared organs is required. Computed tomography and MRI both provide excellent bone and anatomic detail, limited vascular anatomy, shared viscera, and demonstrating organ position,[10] but the consensus appears to be that optimal evaluation is obtained with a combination of ultrasound and MRI. Early diagnosis enables proper counseling of the family and gives the parents the option of termination.

However, in our case, as patient presented only in the third trimester as unbooked case termination of pregnancy at earlier stage was not possible.

CONCLUSION

Thus early diagnosis of conjoint twins, details of organ sharing by imaging modalities will help to predict prognosis and aid in patient counseling. Ultrasonography and MRI in combination is a good diagnostic modality. A joint management with obstetricians, neonatologist and pediatric surgeon would optimize good outcome.

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