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Case Report

Caesarean Section For Thoraco - Omphalopagus Conjoined Twin - A Case Report

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Abstract

Conjoined twins is one of the rarest forms of twin gestation. The reported incidence varies from 1 in every 200 identical twin pregnancies to 1 in 50 000 to 1 in 100 000 live births. Majority of cases reported in literature were diagnosed in first trimester of pregancy and have undergone termination on parents request in view of poor fetal outcome. We report a case of live thoraco omphalopaguspagus conjoined twins, presented at 36 weeks of geatation with breech presentation, weighing 4.2 kg, delivered by elective caesarean section. Babies were sharing heart and greater vessels. Babies were referred to cardio thoracic centre for further evaluation and surgical intervention.Parents were counselled about the possible adverse perinatal outcome.

Keywords: Conjoined twins, Elective caesarean section for twins, Surgical separation of conjoined twins, Siamese twins, Thoraco-omphalopagus

1. Introduction

Conjoined twins are a rare event in mankind, with a reported prevalence of 1:100,000 [1] to 1:20,000 births[2]. Such occurrences have attracted the attention of scientists and also of the general population for decades, as demonstrated by the notorious public life of Chang and Eng, a pair of thoracopagus twins born in old Siam. They gave rise to the popular expression ‘‘Siamese twins’’ to designate any type of conjoined twins.[3] There are seven well-known types of conjoined twins, classified according to the area of conjunction.[4] There is debate on whether conjoined twins are the result of fission or fusion of monoamniotic twins[5] especially based on ventral or dorsal conjoining.[6]

2. Case report

A 28-year-old, third gravida was referred to our university hospital at 35 weeks of gestation with ultrasonographic diagnosis of conjoined twin (thoraco omphalopagus). There was no history of birth of twins in her family. She did not receive prenatal care during index pregnancy. Initial sonography was performed at 28 weeks of gestation, which showed

two fetuses in breech presentation,having maturity of 28 weeks. There were 4 arms, 4 legs, and 2 heads in a single amniotic cavity. After admission,obstetric ultrasound was repeated to confirm the diagnosis. Report confirmed that the twins were in breech presentation with first baby weighing 3.5 kg and second baby weighing 2.7 kgs and were joined at the thorax and upper abdomen. There was a single umbilical cord and only one fetal heart was observed. The placenta was localized anteriorly. There was evidence of polyhydramnios. One of the babies had cleft lip. On the basis of these findings, the diagnosis of thoraco-omphalopagus conjoined twins was made.

Physical examination revealed that the woman was averagely built but poorly nourished. She had mild degree of pallor and her blood pressure was normal. Per abdominal examination revealed over distended uterus due to twins and polyhydramnios. Multiple fetal parts were felt in the uterus. Both babies were in breech presentation. Fetal movement were not felt and fetal heart was indistinctly heard. Per vaginal examination showed posteriorly placed undilated cervix. Her investigations revealed haemoglobin of 8grams percent. Her blood group was A Positive .

Patient and the relatives were informed about the malformation and the twins' poor chance for survival. A written informed consent was obtained from the family .In view of the advanced gestational age and poor prognosis of fetuses, decision of termination of pregnancy was made. Considering the combined expected body weight of the fetuses, decision of elective caesarean section was taken. After making necessary arrangement for blood, patient was posted for caesarean section under general anaesthesia. Senior paediatrician attended the surgery. In view of the large size of the babies ,abdomen was opened by vertical mid line incision. There was no difficulty in extraction of the babies. as the uterus was thinned out and relaxed. The twins were in breech presentation and were joined at the thorax and upper abdomen. There was a single umbilical cord. (Figure 1-4) Babies were handed over to the paediatrician for resuscitation and subsequent care. Babies cried after resuscitation. The Apgar score at birth was 3 and 2 at one minute and was 6 and 7 at five minutes. Total birth weight of babies was 4.2 kgs. Both babies were female. One baby had cleft lip. There was no post-partum haemorrhage. Examination of placenta and membranes revealed monochorionic monoamniotic placenta, weighing 780 grams. There was no obvious morphological abnormality except for bifurcation of umbilical cord near placental insertion . (Figure 6-8) Patients abdominal wall was very much lax and rectus sheath was extremely thinned out due to over distended uterus. Necessary care was taken while closing the abdomen to prevent post operative incisional hernia.

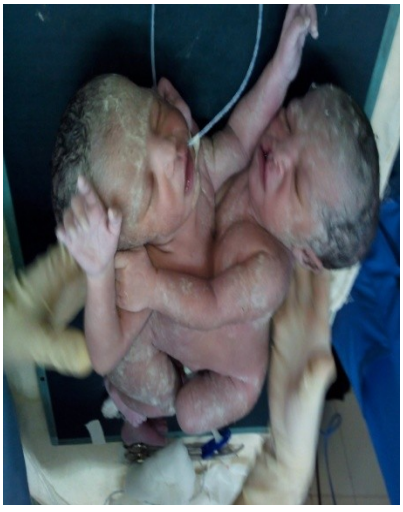
Babies were shifted to intensive neonatal care unit and were given oxygen by mask. They were maintaining oxygen saturation on mask. Babies became tachypnoic after two hours of birth. Kidigram of babies revealed two separate stomach shadows. (Figure 5)Other investigations such as ultrasonography and echo cardiography could not be performed due to deteriorating cardio-respiratory condition of the babies. Babies were transferred to cardio thoracic centre under paediatric supervision. Mother had uneventful post operative period after caesarean section and was discharged from hospital on 7th post operative day.

Clinical photographs-thoraco-omphalopagus conjoined twin

Fig.1,2,3 Showing different views of Thoraco-omphalopagus conjoined twins

(1)

(2)



(3)

Fig. 4,5 Showing Thoraco –omphalopagus with its X-ray

(4)

(5)

Fig. 6,7,8-Showing maternal and fetal surface of placenta and cord attachment

(6)

(7)

(8)

3. Discussion

Conjoined twins have been a source of fascination since time immemorial. Their birth was initially viewed as an ominous sign of impending disaster. This was followed, in western by prolonged periods through the Middle Ages and into the nineteenth century, when they were regarded as freaks or monstrosities and were exhibited with substantial financial reward at circuses and sideshows. Recently, conjoined twins have attracted intense media interest, which coincided with increasing success in their separation.[7]

Conjoined twins represent one of the rarest forms of twin gestation. They occur in roughly 1 in every 200 identical twin pregnancies and are always identical. The incidence ranges from 1 in 50,000 to 1 in 100,000 live births.[8] Because this situation carries high risk, early diagnosis and management of delivery is extremely important..Conjoined twins are classified according to the most prominent site of conjunction: thorax (thoracopagus), abdomen (omphalopagus), sacrum (pygopagus), pelvis (ischiopagus), skull (cephalopagus), and back (rachipagus). Depending on the aspect of the embryonic disc, the most common types are thoracopagus(19%).[9]

Conjoined twins discordant for cleft lip and palate have been reported on thoracopagus twins, with cleft lip in one

twin and no alteration in the other [10]; two pairs of thoraco-omphalopagus twins, with cleft lip and palate in one twin and submucous cleft palate in the other[11] and another pair with cleft lip and palate in only one twin.[12]

Its etiology is unknown, but an incomplete division of the zygote between 13th and 15th days after fertilization probably occurs.[13] The overall survival rate for conjoined twins is approximately 25%.[14] The condition is more frequently found among females, with a ratio of 3:1.[13] Two contradicting theories exist to explain the origins of conjoined twins. The traditional theory is fission, in which the fertilized egg splits partially and conjoined twins represent delayed separation of the embryonic mass after day 12 of fertilization. The second theory is fusion, in which a fertilized egg completely separates, but stem cells (which search for similar cells) find like-stem cells on the other twin and fuse the twins together.[13,15,16]

Early diagnosis of conjoined twins was previously reported, but not before the 10th week of gestation.[17] On careful transvaginal sonography and serial scanning, there appears to be an inability to separate between the anatomical parts of the fetuses. Once conjoined twins have been diagnosed, characterization of the type and severity of the abnormality can be performed with ultrasound, three-dimensional ultrasound, computed tomography, or magnetic resonance imaging.[18,19] Sonographic characteristics of conjoined twins include the absence of a separating membrane, conjoined body parts, inseparable bodies or heads between the twins despite a change in fetal position, and a bifid appearance of the fetal pole in the first trimester.[20] Other sonographic findings suggestive of conjoined twins include more than 3 vessels in a single umbilical cord, complex fetal anomalies, heads or bodies at the same level, a hyperextended spine, unusual proximity of the extremities, and persistence of the position relative to one another after movement or at follow-up scanning.[20]

Surgery to separate conjoined twins may range from relatively simple to extremely complex, depending on the point of attachment and the internal parts that are shared. Most cases of separation are extremely risky and life-threatening.

If the condition is diagnosed early, as it happens in most of the cases, termination of pregnancy is offered on ethical and social grounds, as the long term prognosis of the babies is not good. The condition is likely to remain undiagnosed till advanced gestational age in women who do not receive antenatal care, as it happened in this case. After the diagnosis of the condition at advanced gestational age,it is important to counsel the woman and the relatives about the plan of management and the probable fetal outcome after birth. These cases must be managed at tertiary care centre as there is increased risk of maternal and fetal complications. Caesarean section is the preferred mode of delivery to avoid problems of dystocia, prolonged labour and rupture of the uterus. There is likely hood of atonic post-partum haemorrhage due to over distended uterus. Arrangement of blood must be made before taking patient for caesarean section.

Counselling of relatives regarding need for admission of babies to neonatal intensive care unit and subsequent surgical separation at higher centre is necessary. It is ideal to do in utero transfer of the baby to higher centre to avoid the risk and problems that may arise while subsequent shifting the baby to higher centre. In the present case relatives were not willing to take woman to higher centre for financial reasons. Babies started showing signs of deterioration after 3 hours of birth due to sharing of heart and greater vessels. Paediatrician some how managed to arrange transfer of the babies to cardio thoracic centre few hours after birth .

4. Conclusion

Conjoined twin is a rare obstetric condition that obstetrician may come across in his lifetime. Termination of pregnancy is preferred option ,when diagnosis is made during early pregnancy. In utero transfer of babies to higher centre should be ideal thing to do in advanced pregnancies. There are many social, psychological, financial and surgical issues associated with the management of conjoined twins. Overall fetal prognosis is unsatisfactory.

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