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CASE REPORT

Conjoined twin

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Abstract

In the recent past we encountered a pair of newborn who were symmetrically conjoined. We found a 30 years old woman who delivered those conjoined babies by caesarean section at full term. Antenatal ultrasonography at 26 weeks revealed twin pregnancy but no comment regarding conjunction. After delivery, babies were found fuse from upper thorax to umbilical region. Postnatal ultrasonography revealed single liver. Till date many such cases have been reported worldwide but so far no case report has been published in our country.

Introduction

Conjoined twins are rare presentation of monozygotic pregnancies, incidence 1 in 50,000-1,00,000 live births and 1 in 400 monozygotic twin birth. There is female preponderance 3:1 ratio and are always of same gender.1 Both are monozygotic, monoamniotic and monochorionic. They can be symmetric or asymmetric (heteropagus) forms. Symmetric conjoined twin cases are: thoracopagus (chest), omphalopagus (umbilicus), pygopagus (rump), ischiopagus (hip), craniopagus (cranium), parapagus (side), cephalopagus (head), and rachipagus (spine). They are always joined at homologous sites. But asymmetric cases result from demise of one twin with remnant structures attached to the complete twin. Fetus in feto refers to asymmetrical monozygotic intraparasitic twins.2 Anatomic site shared by conjoined twins can be complex e.g. thoraco omphalopagus. In such cases, 75.% have conjoined hearts, 50% have fusion of intestinal tracts and virtually all have a shared liver.1,3 Diagnosis can be made as early as the 9th week of gestation by ultrasonography (USG). Majorities are dead born (28%) or die soon after birth (54%). Those born alive may have complex congenital problem. Only around 20% survive. In thoracic conjunction, prognosis is usually very poor.2,3 Different varieties of conjoint twin cases has been reported.4-7 But attempts to surgery have been done only in small number of cases.8 Many alive conjoined twin cases have been reported. Ronnie and Donnie from Ohio, USA, the oldest living conjoined twins as they reached their 63rd birthday last year.9 A Bangladeshi born conjoined twin (craniopagus) girls Trishna and Krishna were successfully separated at the Royal Children's Hospital in Melbourne, Australia and thereafter returned back to their parents in Bangladesh.10

Case report

A 30 years old women, para 3, 4th gravida, hailing from a poor village farmer's family, delivered those symmetric conjoined twin (Fig- 1,2,3) weighing 3400 grams by caesarean section at full term in a private clinic of a Upazilla in Bangladesh. This was her unplanned pregnancy. She had been taking oral contraceptive pill for last four years following last delivery upto this conception. Her first day of last menstruation (LMP) was 11.09.15 and expected date of delivery (EDD) was 18.06.16. Antenatal Ultrasonography done at 26 weeks by a physician trained in sonography, found twin pregnancy but did not mention any comment regarding conjunction. She took three antenatal visits at Upazilla, health complex at 24, 31, 37 wks pregnancy consecutively. She had no medical illness during

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antenatal. period. Elective caesarean section was done at term in a private clinic at Upazilla and found the babies conjoined.

Bilateral tubal ligation was done per operatively after taking written consent and prior counseling. Her past medical history was also unremarkable. Previous three children were delivered per vaginally without any complication.

Age of last child was 5 years. All family members were in good health. She had no family or personal history of twins. After delivery, babies were immediately referred to district hospital. Physical examination showed, both were positioned face to face and fused from upper

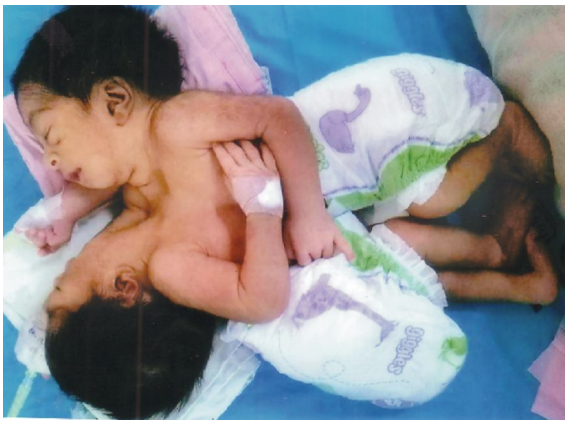

Fig 1: Conjoined twin showing thoracopagus with typical neck extension.

thorax to umbilical region sharing single umbilical cord. Head, neck and limbs were normal in both the babies. Postnatal ultrasonography done by a qualified sonologist in

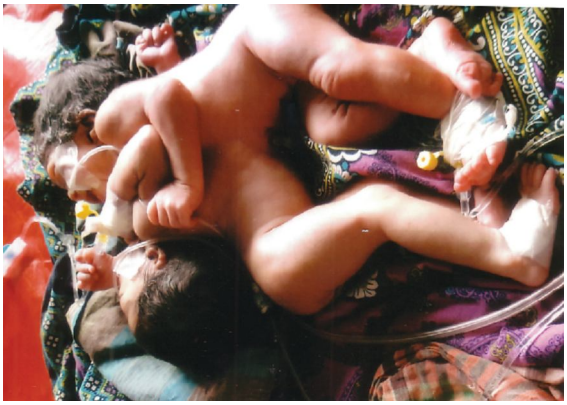

Fig 2: Same conjoined twin showing omphalopagus with single umbilical cord.

sadar hospital revealed single liver and rest of the organs were normal. They were further referred to Medical University on 3rd post natal day for further evaluation and better management. But unfortunately babies died on 5 th day of age due to early onset neonatal sepsis with apnea and shock.

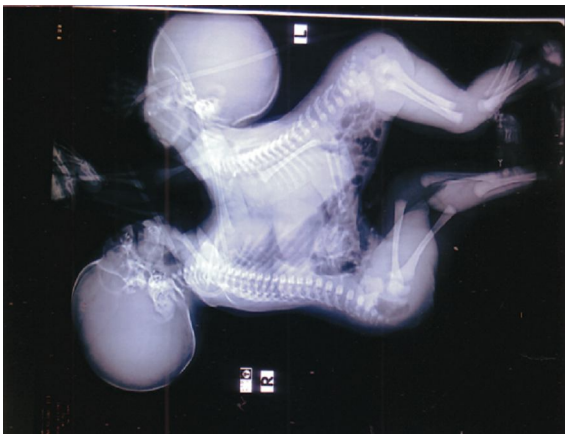

Fig 3: Radiograph showing symmetrical conjoined thoraco-omphalopagus twin.

Discussion

Symmetrical conjoined twins are one of the rarest major congenital malformation/ anomaly. There may have additional congenital defect/s. They are individual and deformed but symmetrical and proportional; develop dissimilar personalities from an early age.[2]

Two different theories have been suggested in the formation of conjoined twins. One is "fission" theory and another is "fusion" theory. 1) "fission" theory: failure of separation of the embryonic plate between 15th to 17th days gestation. 2) "fusion" theory: secondary union of two separate embryonic discs at the dorsal neural tube or ventral yolk sac areas at 3 to 4 weeks gestation. Recently the fusion theory is accepted because it can explain all conjoined twin phenomenon. Long term usage of contraceptive drugs, abnormal calcium metabolism and extremely underweight woman with ovulatory dysfunctions are suspected in etiology.[1,2]

Antenatal USG and MRI can help in diagnosing conjoined twins. During early antenatal period, conjoined fetuses should be evaluated for conjunction aspects: location, extension of adhesion, presence of fetal anomalies, existent of organ sharing. Diagnosis can be made as early as the 9th week of gestation.1 Diagnostic USG criteria for conjoined twins include 'relative fixed

position of the two fetuses facing each other, as in thoracopagus, with hyperextension of the cervical spine; Continuity of the skin and mirror image body parts with limbs close together, presence of single heart, fused liver, solitary umbilical cord, fused spine, fetal body parts at the same level, etc' confirm the diagnosis.2,3 The decision to terminate the pregnancy may be taken after detailed evaluation and counseling after confirming diagnosis.

Immediate postnatal management consists of resuscitation and stabilization.. This is followed by a thorough physical examination with special investigations to define the relevant anatomy. The site of conjunction will determine the type and order of special investigations. In thoracopagus cases following investigations should be done: USG of the abdomen and skull, Echocardiography, Radiography, Fetal echocardiography, Computed angiotomography, Doppler USG of the abdomen, Magnetic resonance imaging (MRI). Important structures to evaluate are the cardiac, hepatobiliary, intestinal, urogenital, and spinal systems. In typical thoracopagus cases, they are positioned face to face and very commonly sternum, heart, diaphragm and upper abdominal wall are conjoined. Twins with classic omphalopagus cases are conjoined only in the umbilical region, but often containing the lower thorax. Liver is around 80 % mutual and this sharing is usually not equally balanced. Heart is almost always separate, but pericardial. adhesion may be present. Congenital heart disease present in 30% of the cases.1 This case showed that twins were found face to face with typical neck extension position, whole of the chest and upper abdomen upto umbilicus were fused sharing single umbilicus. Ultrasonography and radiology showed single liver.

Information obtained from investigations will determine further management e.g. surgical approach, timing of separation, allocation of organs and structures, and eventual prognosis regarding survival and functional outcome. If separation is possible and desirable, surgery should be performed within the first 6 to 9 months, before the twins develop an awareness of their condition. Despite successful separation, some children are left crippled and disabled, requiring lifelong follow up and care. The overall survival for symmetrical twins is 33.3%, but it is 64.7% for those who underwent surgery. Better preconceptual maternal nutrition with folic acid supplementation is likely to reduce the incidence.2

Diagnosis can be made during antenatal period by careful ultrasonography. Decision to terminate early in complicated cases can minimize the unnecessary harassments and untoward medical circumstances. In viable cases, timely referral to appropriate health facility, prompt medical treatment & surgical interventions can save either one or both the child's life.

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