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Case Report

Dicephalic parapagus twin - A rare case report

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ABSTRACT

We are reporting a rare case of a dicephalic parapagus twin born to multigravida mother diagnosed antenatally few hours before birth by ultrasonography. The baby was born by emergency cesarean section. The baby was full-term live male baby; however, the baby expired few hours after birth. Due to the rarity of the case, we are prompted to report this case.

Key words: *Dicephalic parapagus, Conjoined twins, Neonate, Congenital malformation*

Conjoined twins are identical monozygotic twins joined *in utero* that do not separate fully from one another and are partially united due to incomplete division of one fertilized ovum. Incidence ranges from 1 in 50,000 to 1 in 180,000. Approximately 50% of these babies are still born. Conjoined twins are typically classified by the point at which their bodies are joined. The most common types of conjoined twins are thoracocephalopagus, thoracopagus, omphalopagus, craniopagus, cephalopagus and xiphopagus. The most famous pair of conjoined twin was of Chang and Eng Bunker (1811-1874), the Thai brothers who were born in saim, now Thailand. They traveled in P.T. Barnum's circus for many years and were labeled as Siamese twins. Due to brothers' fame and the rarity of the condition, the term Siamese twin came to be used as a synonym for conjoined twin. In 1957, Bertram Katz and his surgical team made international medical history performing the world's first successful separation of conjoined twins named John Nelson and James Edward Freeman born in 1956 who were omphalopagus.

Dicephaly is a rare form of conjoined twins. This rare condition of dicephalic head occurs when before the implantation, the zygote fails to divide at the level of neck and the twins are joined below the neck. There are many occurrences of multiheaded animals in the both reality and mythology. Two headed people though rare have been known to exist and documented. The Scottish brothers were conjoined twins allegedly dicephalic were born in 1460, had attained 28 years of age. So far, we have only one successful modern case of famous Abhigail and Brittany Hensel, born in 7 March 1990 in Minnesota, USA. They have two spine which join at pelvis. They each have individual brain, spinal cord, and heart. They have four lungs, two stomach, two gall bladder, three kidneys and three arms. The arm between the head was useless and was amputated in infancy. Each twin controls the limb and body part on her side with coordination that allows them to walk, run, play piano, swim, drive, etc.

Conjoined twins must be suspected in all monochorionic monoamniotic twin pregnancies, and careful sonographic assessment must be undertaken to exclude any classical sign suggestive of conjoined twins and to identify the severity of shared fetal organ for the perinatal management [1]. Dicephalic twin though rare must be kept in mind. When serious malformation that is incompatible with life are diagnosed in early gestational age, termination of pregnancy must be advised.

CASE REPORT

A dicephalic twin was born with complete maturity to a multigravida mother in India. The baby was delivered by emergency cesarean section. The mother presented in active labor. There was no history of abortion, infertility, medical problem, or drug history in mother. Ultrasonography done at this admission diagnosed; this rare occurrence of conjoined twin with dicephalic head, and hence, an emergency cesarean section was taken up. The twin had one trunk, two arms, two legs but two head joined at neck. The two head were joined side-by-side. The chest was wide in dimension. The genitalia looked normal. The placenta was monochorionic. The umbilical cord was centrally placed with artery to vein ratio 2:1. Apgar at 1 min was 5/10 and at 5 min was 7/10.

After being born, the baby was intubated under all aseptic precaution due to impending respiratory failure (Figure 1). Left head was intubated first and then the second head. During intubation, larynx appeared normal. After intubation, air entry was equal on both the sides of the chest from both the endotracheal tube. No investigation could be done on the twin due to the poor general condition. One head of the baby developed progressive cyanosis due to compromised blood supply to one head. The baby developed progressive shock and cyanosis due to likely associated heart defect which could have been not compatible with life. Resuscitation was started, but the twin could not be revived by all

possible measures. Post-mortem examination could not be done as parents of the baby did not give consent.

DISCUSSION

Conjoined twins are derived from one fertilized ovum and are monochorionic, monoamniotic. There is incomplete fission of a single embryonic disc which should occur at 13-15 days after the ovum has fertilized. There is an alternate theory which suggests that the fertilized ovum divides completely into two embryonic disc whose unusual proximity results in secondary fusion into the conjoined twins, as the embryo enlarge. Most conjoined twins face each other. In contrast, dicephalic twins are joined side-by-side. Which results in disturbed cross-signaling between the two primitive streak giving rise to numerous anomalies of the organs due to defective embryogenesis.

Cunniff et al. (1988) hypothesizes had rotation of heart at the atrial level initiate lateralization of the embryo and that interferes with heart rotation and causes abnormalities in some right dicephalic twins [2]. There are only a twelve reported cases of modern dicephalic live birth, and only a few among them have made it to the adulthood. Among them, only three were male. Most dicehalic babies are stillborn or aborted before 24 weeks if diagnosed early. Human dicephalic twins look like a two-headed person with each brain supporting a distinct mental life. Stillbirth and mortality are very high in dicephalic twins. They usually die within few hours to days after birth. Those who survive have high morbidity with short life span due to anomalies which occur in extensive sharing of the organ. Live dicephalic babies have also been reported from Turkey in 2000, Egypt and Indonesia in 2003, Phillipines in 2007, Bangladesh in 2008, Indonesia in 2009 again, Brazil in 2001 and 2011, India in 2003 and 2014 and from Atlanta in 2014. All these babies expired during their neonatal or early infancy period due to the complex anomaly commonly found in dicephalic twins.

Diagnosis must be made before birth to minimize injury and planning a suitable delivery. Criteria for ultrasound examination for conjoined twins include the absence of separating amniotic membrane, inseparable fetal bodies, lack of relative change of bodies, and fetal head on repeated examinations [3]. Investigations that must be done before birth are three dimensional ultrasound, magnetic resonance imaging, and fetal echocardiography. Investigations to be done after birth are ultrasonography of whole abdomen, skull, spine, echocardiography, angiography cardiac catheterization, radionuclide scanning, cystography or urethrography, and gastrointestinal contrast studies to clarify the degree of conjoining and ideal obstetrical and perinatal management.

Management of conjoined twins includes three options. The first option is to withhold or withdraw the life support to allow the twins to die. One may question whether parents can justifiably refuse intervention due to legal issues. Second is to separate one twin with the possibility that one will die. This is not possible in dicephalic head. The third option which is most appropriate for conjoined twins is allowing the twins to remain conjoined [4]. Dicephalic head twins are generally inseparable, and surgical management is generally not possible in dicephalic twins due to extensive sharing of organs. Hence, comfort measures are the most appropriate management for them.

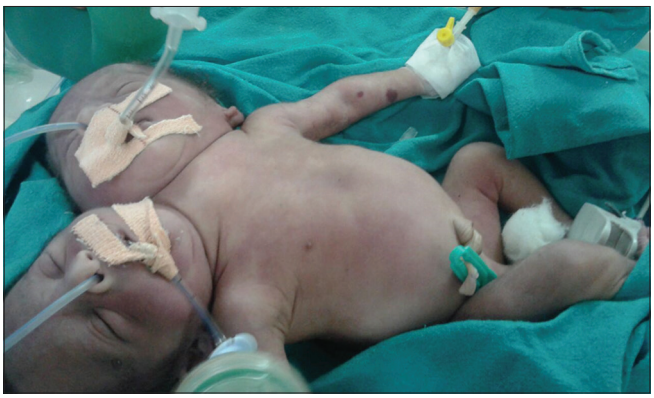

Figure 1: Clinical photograph of the dicephalic twin

CONCLUSION

Conjoined twins are rare, and parapagus dicephalic twin represents only a small proportion. Anomalies of the heart and abdominal organs occur in most of the dicephalic twin, which was the likely cause of death in our case. Early diagnosis facilitates termination or optimal obstetric and perinatal preparation. Practitioners may unexpectedly encounter conjoined twins during labor,which happened in the present case, or at delivery.

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