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Radio Diagnosis

DICEPHALUS DIPUS DIBRACHIUS CONJOINT TWIN RAREST OF RARE

Dr. Swati Gupta	(DMRD), Radiologist
Dr. Abhishek Gupta*	(MS), Assistant Professor, Department of Surgery, Government Medical College, Orai (Jalaun). *Corresponding Author
Dr. Vishal Jain	(MD), Radiologist

KEYWORDS :

INTRODUCTION

Conjoined twins are a rare congenital malformations. Rarest among them is the dicephalus parapagus twin. Conjoined twin pregnancy of the parapagus dicephalus dibrachius dipus (PDDD) type. This type of conjoined twin is defined by lateral fusion with two heads, sharing a single thorax, abdomen and pelvis with two arms and two legs. [10]

There have been two opposing embryologic theories for the development of conjoined twins: "fission" and "fusion." The former attributes the development of conjoined twins to incomplete division of the developing embryo, while the latter argues that all eight types occur by secondary fusion of two embryonic discs beyond the 13th day post-conception.[11]

Here we are presenting a case of dicephalus dipus dibrachius conjoint twin diagnosed on transabdominal sonography.

CLINICAL HISTORY

A 26 year old patient, second gravida with 6 months ammenorhea came to our setup for routine fetal examination. The patient had a previous history of miscarriage . The patient was clinically stable and with normal vital parameters. No associated complains were given by the patient.

ULTRASOUND FINDING

On initial ultrasound examination the fetus which was corresponding to 24 weeks and 5 days according to last menstrual period , showed a large head with increased biparietal diameter and head circumference, corresponding to approx 40 weeks parameter. The rest of the parameters of abdominal circumference and femur length were within the normal limits.

On further detailed examination it was found there were two head with fully developed brain parenchyma which were attached to each other from the occiput. The facial features of both the head were well formed with very well visualized orbits, nose and lips. Two seperate spine were seen in the neck region with well formed vertebrae.

However below the neck single thorax , single abdominal cavity, two upper limbs , two lower limbs, single umblical cord and single genitalia(but ambiguous) were seen.

The other associated anomalies were cardiomegaly with ill visualized great vessels. dilated ventricle , and ventricular septal defect was seen. The stomach bubble was seen in the thoracic cavity at the level of heart suggesting diaphragmatic hernia.

Bilateral renal, bladder were normal. The genitalia were single but ambiguous type. Both the upper and lower limbs were well formed and normal in appearance.

The patient was further referred to gynaecologist. Later on the preganancy was terminated on medical and ethical grounds. Post delivery the diagnosis was confirmed to be dicephalus dipus dibrachius conjoint twin.

DISCUSSION

Conjoined twins (CT) are a very rare developmental accident of uncertain etiology. Prevalence has been previously estimated to be 1 in 50,000 to 1 in 100,000 births.[1]

Dicephalus is a subset of parapagus in which twins share a common body from the neck or upper chest downwards, having only a pair of limbs and a set of reproductive organs. This anomaly represents less than 0.5 per cent of all reported cases of conjoined twins .[2,3]

Conjoined twins form when one sperm fertilizes one egg, but at some point during the stage where the single egg splits, the process stops, and the twins develop attached to one another. Identical twins will only be conjoined if they separate after the 12th day of conception. No one knows why conjoined twins occur, although genetic and environmental factors have been explored. Conjoined twins are always of the same sex, same types (dicephalus & ischiopagus for example) may only have one set of external genitalia (or none at all).[7] They are most likely to occur in India or Africa than in China or the US (although the rates in Vietnam have been much higher in recent years, possibly due to Agent Orange exposure).

The conjoined twins when united laterally can lead to types like:

a) Parapagy - twins that share a conjoined pelvics, one symphysis pubis and one or two sacrum's united side by side. This type account for about 5%. It is possible for them to share a heart (if one has situs inversus of the heart), but rare. They may share the liver, kidneys and other organs.
b) Dithoracic parapagus - when the union is united to the abdomen and pelvis (does not involve thorax). This is a subset of parapagus twins. They will be of the dipus type. They will likely to have their own hearts, although some will have situs inversus and would possibly share a heart as well.
c) Dicephalic parapagus (dicephalus) - when the union includes the abdomen, pelvis and thorax. Dicephalus have one trunk with two heads. They are always of the dipus (two legs) type, and 3rd and 4th arms may present on their backs. This is a subset of parapagus twins. They may share a heart and other organs.

The rarest type of pappagus twin is dicephalus dipus dibrachius.

- Dibrachius- two arms are present (about 10% of all dicephallus/ dithoracic twins).
- Tribrachius-three arms are present (about 10% of all dicephalus/ dithoracictwins).
- Tetrabrachius-four arms are present (about 80% of all dicephalus/ dithoracic twins).

d) Diproscopic parapagus- a single trunk and single head with two faces. Various level of duplication of the face and brain can be seen. This is the most uncommon and rare type of conjoined twinning, also called as Monocephalic Diprosopos.[7]

Parapagus conjoined twins can be divided broadly into two types: Parapagus dicephalus (two heads, single trunk/ abdomen/pelvis with 4-7 limbs) and parapagus diprosopus (two faces on a single head, single trunk/abdomen/pelvis and four limbs). [11]

The distance between the rostral ends of the embryonic disc increases as the spectrum of parapagus conjoined twins progresses from diprosopus to dicephalus. As this distance increases, there is increasing separation of the conjoined twins; the heads separate first, followed by the thoraces, while the number of limbs increases from four to a maximum of seven.

Conjoined twins often have several associated anomalies in various organ systems, including musculoskeletal, cardiac, gastrointestinal, and the central nervous system. As the number of limbs and degree of separation increases, so does the likelihood of organ duplication. For example, parapagus diprosopus twins (parapagus type with the least degree of physical separation) will share a single heart. However, parapagus dicephalus twins have greater separation that may result in a partially duplicated or two separate hearts. This phenomenon is not limited to the thorax and the same principles apply to the abdomen, pelvis, and CNS.[11] The frequent anomalies which are associated with conjoined twinning are the duplication of the visceral organs, omphalocele, facial clefts, meningomyelocele, an imperforated anus and cardiac defects .[6]

CONCLUSION

Prenatal diagnosis helps us to give a mental and physical support to the patient and the concerned family by proper counselling. It helps us in planning the delivery to ensure mothers safety which might in later ignored status may lead to significant maternal morbidity/ mortality. Therefore it is important to diagnosis the anomalies in early stages for the better outcome of the patient and reducing the comorbid conditions.

Fig 1:Transverse image of the fetal head and thorax showing discrepancy in the size . The brain parenchyma appears separate with intervening falx delineated clearly.

Fig 2: Transverse section of the head at the level of orbit showing two separate facial profile with well formed orbit (O) and nose (N).

Fig 3: Transverse section of the fetal neck region showing separate neck and spine with a fluid filled space in between posteriorly, which was in continuation into the intracranial space in between both the brain parenchyma.

Fig 4: Transverse section of the thorax showing single thoracic cavity with a single enlarged heart(H). Another cystic area seen in the contra lateral side (S) stomach herniating into the thorax - diaphragmatic hernia.

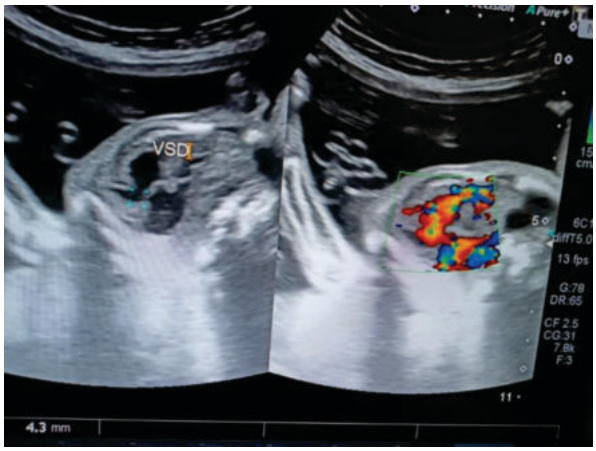

Fig 5: Transverse section at the level of heart shows enlarged abnormal shape heart with prominent ventricles and interventricular septal defect.

Fig 6,7and 8: - Post delivery images of the fetus with separate face, with ventrally fused head and neck , single trunk, single umbilical cord, two upper and lower limbs and ambigouis genitalia confirming it to be dicephalus dipus dibrachius conjoint twin.

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