Parapagus dicephalus dibrachus dipus: A case of conjoined twins

Parapagus dicephalus dibrachus dipus: Yapışık ikiz olgusu

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Abstract

In this report, we describe the case of a woman with a prenatal diagnosis of parapagus dicephalus dibrachus dipus conjoined twins at 14-15 weeks of gestation via two-dimensional ultrasonography. The parents elected to terminate the pregnancy and the patient delivered a 15-cm 130-g male fetus. There were two heads and necks which appeared grossly normal. The thoracic and abdominal cavities were shared. The fetus had four normal limbs. On internal examination, there were two separate structurally normal hearts. There were two larynges and four lungs. Two esophagi fused to enter a single stomach. The diaphragm was common and separated a single abdominal cavity. Distally, the alimentary system including the liver and gallbladder was single. (J Turkish-German Gynecol Assoc 2009; 10: 241-3)

Key words: Conjoined twins, parapagus twins, ultrasonography

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Özet

Bu vaka sunumunda 14- 15. gebelik haftasında ultrasonografi aracılığıyla tanı konulan parapagus dicephalus dibrachus dipus yapışık ikiz olgusu tanımlanmaktadır. Ebeveynler gebeliğin sonlandırılmasını tercih etmişler ve hasta 15-cm boyunda 130-g ağırlığında erkek fetüs doğurmuştur. Dıştan görünümü normal, iki baş ve boyuna sahip fetüsün göğüs ve karın boşlukları ortaktır. Fetus dört adet normal ekstremiteye sahiptir. Otopsi incelemesinde fetüsün yapısal olarak normal iki ayrı kalbe, iki larenkse ve dört akciğere sahip olduğu görülmüştür. İki özofagusun distalde tek bir mideye girdiği görülmektedir. Diyafram ortak olup, tek karın boşluğunu göğüs boşluğundan ayırmaktadır. Fetüsün bir adet karaciğer ve safra kesesini de içeren tek gastrointestinal sisteme sahip olduğu görülmektedir.

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Anahtar kelimeler: Yapışık ikiz, parapagus ikiz, ultrasonografi

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Introduction

The incidence of conjoined twins is between one in 50,000 pregnancies, and one in 650-900 twin pregnancies. The incidence of live born conjoined twins is a one set per 200,000 live births (1, 2). Conjoined twinning is a random event, unrelated to heredity, maternal age or parity (3).

Two theories have been proposed to explain conjoined twinning. Classically, the theory asserts that incomplete fission of a single embryonic disc occurs 13 to 15 days after the ovum is fertilized (fission theory). More recently, embryologic studies of conjoined twinning have indicated an alternative postulation that this developmental anomaly could originate from the secondary fusion of two separate monovular embryonic discs (fusion theory) (4).

Conjoined twins are mainly classified according to incomplete duplication (parasitic) or complete duplication. There are eight types of completely duplicated conjoined twins according to the most prominent site of union. (Table 1) (5).The most common type of union is thoraco-pagus and/ or omphalo-pagus (anterior thoracoabdominal fusion) and is found in 40-75% cases. Parapagus twins represent an extremely rare type of conjoined twins.

Here we describe the prenatal diagnosis of a case of parapagus (dicephalus, dibrachius, dipus) conjoined twins diagnosed during the first trimester with two-dimensional ultrasound at 14-15 weeks of gestation.

Case report

An 18-year old woman, gravida 2, para 1 was referred for routine scan at 14-15 weeks. Her previous pregnancy was uncomplicated and she had a 5 year old healthy child. She had no personal or family history of twins. The patient and husband were non-consanguinous. However, she is an identical twin herself.

Two-dimensional (2D) ultrasound scan demonstrated a conjoined twin pregnancy. There were two heads, two upper limbs and two lower limbs (Figure 1a). The twins were joined at the thorax and abdomen and there were two hearts. The diagnosis of dicephalus parapagus was made on the observation of two heads, one body, and one umbilical cord ultrasonographically. The couple was informed about the findings and poor outcome. They opted to have a termination of pregnancy. At autopsy, the weight of the conjoined twins was 130g, the

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Table 1. Embryologic Classification of Conjoined Twins [5]

Embryonic aspect (%)	Type	Incidence (%)	Extent of union
Ventral (87)			
Rostral (48)	Cephalopagus	11	Top of head to umbilicus
	Thoracopagus	19	Thorax, upper abdomen, conjoined heart
	Omphalopagus	18	Thorax, upper abdomen, separate heart
Caudal (11)	Ischiopagus	11	Lower abdomen, genito-urinary tract
Lateral (28)	Parapagus	28	Pelvis, variable trunk; diprosopus 2 faces, dicephalus 2 heads
Dorsal (13)	Craniopagus	5	Cranial vault
	Pygopagus	6	Sacrum
	Rachiopagus	2	Vertebral column

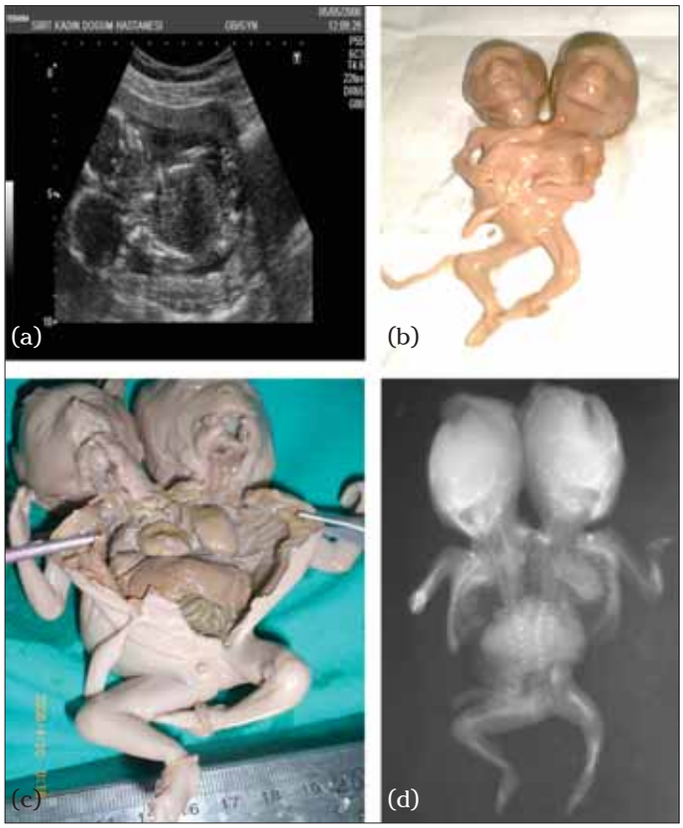

Figure 1. (a) Two-dimensional ultrasound scan of conjoined twins at 14-15 weeks of gestation. (b) Post-mortal image of the fetus anteriorly demonstrating two hands, conjoined at the thorax and the abdomen with two upper and lower limbs. (c) At autopsy, there were two separate structurally normal hearts. There were two larynxes and four lungs. The diaphragm is common and separated a single abdominal cavity. (d) A plain X-ray film showed two vertebral columns joining at the common pelvis and two separate chest cavities

head circumferences of the fetuses were approximately 90mm, the thorax circumference was 130 mm and the abdomen circumference was 100mm. The crown- heel length was 150 mm consistent with 14-15 weeks' gestation. There were two heads and necks which appeared grossly normal. The thoracic and abdominal cavities were shared. The conjoined twins had two upper limbs and two lower limbs (Figure 1b). Both upper and lower limbs were normal with five digits on each hand. The anus was normal. There was male external genitalia.

On internal examination there were two separate structurally normal hearts (Figure 1c). There were two larynxes and four lungs. Two esophagi fused to enter a single stomach. The diaphragm was common and separated a single abdominal cavity. Distally, the alimentary system including the liver and gallbladder was single. There was one pair of kidneys and adrenal glands. Two ureters drained into a single bladder. There were two pairs of testes within the abdominal cavity. There was a normal placenta and a normal umbilical cord.

X-rays demonstrated that each twin had the normal complement of vertebrae and 12 pairs of ribs. A plain X-ray film showed two vertebral columns joining at the common pelvis and two separate chest cavities Upper and lower limb bones were normal. (Figure 1d).

Discussion

Spontaneous twinning occurs in 1.6% of all human pregnancies, of which 1.2% are dizygotic and 0.4% are monozygotic. Approximately 5% of monochorionic pregnancies are monochorionic monoamniotic and 1% is conjoined.

A conjoined twin is a rare occurrence and the parapagus variety is even rarer. The developmental processes that underlie conjoined twinning are imperfectly understood, but recent research based on the analysis of large numbers of conjoined twins suggests that they form early in development due to the secondary union of two monovular embryonic discs (5). Parapagus twins lie side by side with venterolateral fusion. Most commonly; these twins are conjoined at the chest, with joined liver and diaphragm but separate respiratory and upper gastrointestinal tracts, two arms; two legs, and two complete spinal cords and vertebral columns, a single shared genitourinary system and lower gastrointestinal tract (4). All parapagus twins have one umbilicus and a conjoined diaphragm and liver. In this case, there was a single umbilical cord and a single shared pelvic region and abdomen, but two joined thoraxes having two

complete vertebral columns, two hearts and two pairs of lungs, two arms and two legs.

Although findings at autopsy were normal in this case, other dicephalus twins have had characteristic abnormal findings. Past studies described complex anomalies of the heart and abdominal laterality heart abnormality. Ventricular cardiac union is reported in parapagus twins. Many of our parapagus twins have had a common pericardium (6). Defects of lateralization including right and left atrial isomerism and mirror imagery are known to be particularly common in parapagus twins. There are generally two sets of lungs, which may be underdeveloped or anomalous. Neural tube defects, cystic hygroma, clubfoot, and imperforate anus have occurred in parapagus twins (7).

Before the recent improvement of quality of ultrasound depiction, the diagnosis of conjoined twins was not always easy and therefore often missed. Sonography is now used widely in obstetrics, and it can detect conjoined twins as early as 12 weeks of gestation. Whenever monozygotic twinning is observed, conjoining should be suspected. The presence of polyhydramnios and both twins in breech or in face-to face position alert the sonographer to the possibility of conjoined twins (8).

In conclusion, conjoined twins have intrigued physicians for centuries. Their management is often extremely complex and experience with large numbers restricted to a few centers worldwide. Most of the conjoined twins are born prematurely, around 40% are stillborn, and 35% die in the first 24 hours of life (9). Postnatal separation can be achieved in rare cases. Overall, the prognosis depends on the type of fusion and presence of associated structural defects. Accurate antenatal assessment allows the parents to be counseled as to the probable outcome of the pregnancy and the likelihood of successful postnatal separation. First or second trimester detection of conjoined twins enables obstetricians to counsel parents about potential termination, or about delivery and treatment options if pregnancy is continued.

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