A CASE OF PARAPAGUS DICEPHALUS CONJOINED TWINS DIAGNOSED AT 12TH WEEK OF GESTATION

Case Report

ILK TRİMESTERDE TANISI KONULMUŞ PARAPAGUS DİCEPHALUS YAPIŞIK IKİZ

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ABSTRACT

Objective: Dicephalic parapagus is a rare anomaly of conjoined twins with severe morbidity and mortality. Early prenatal diagnosis is crucial for the follow-up of the pregnancy. In this article, prenatal diagnosis of parapagus twins with ultrasonography in the first trimester is presented.

Case report: Twenty-four-year-old pregnant woman with gravidity 2 whose personal and familial background was uneventful was diagnosed with parapagus dicephalus conjoined twins in the 12th week of gestation by ultrasonographic examination. Fetus with 2 heads with seperate necks, 2 arms, 3 legs, and fused vertebral column was observed by ultrasonographic examination.

Conclusion: Early diagnosis of parapagus dicephalus conjoined twins seems to be very important for the management and termination of pregnancy when required.

Keywords: *Conjoined twins; parapagus dicephalus; prenatal ultrasound.*

ÖZET

Amaç: Yapışık ikizler oldukça nadir görülen morbidite ve mortalitesi yüksek olan durumlardır. Erken tanı gebelik tabibinde önemlidir.Bu yazıda ilk trimester ultrasonografi değerlendirilmesinde tanı almış bir Parapagus Dicephalus yapışık ikiz olgusunun sunulması amaçlanmıştır.

Olgu: 24 yaşında ikinci gebeliği olan hasta rutin ilk trimester rutin muayene için kliniğimize başvurdu. Ultrasonografik değerlendirmede 12 hafta ile uyumlu parapagus dicephalus yapışık ikiz görünümü mevcuttu. Ayrıntılı değerlendirmede iki boyun ve iki baş ile birlikte tek toraks ve abdomen izlendi. Kolumna vertebralis torakstan itibaren birleşik olup üst kol ve üç alt ekstremite izlendi.

Sonuç: Birleşik ikizlerde erken tanı konulması gebelik yönetiminde ve gerekli olgularda erken gebelik terminasyonu yapılmasına olanak sağlaması açısından önemlidir.

Anahtar kelimeler: *Yapışık ikiz; parapagus dicephalus; prenatal ultrasonografi.*

INTRODUCTION

The frequency of conjoined twins has been estimated as 1 in 50000 gestations. However, the true incidence is around 1 in 250000 live births since over 60% succumb in utero or are stillborn (1). To describe the type of conjoining, the suffixes "pag" (meaning fastened or stuck) and "us" are added to the term used for the most prominent site of union (2). Though the exact etiology of conjoined twins is not known, two theories are suggested (1). According to the first theory, there is a failure to undergo complete separation of the embryonic disc at around the 15th to 17th day of gestation. A secondary fusion occurs between two originally separate monovular embryonic discs in fusion theory (2). Eight types of pagus conjoined twins should be recognized such as omphalopagus (umblicus), thoracopagus (chest), cephalopagus (head), ischiopagus (hip), craniopagus (helmet), rachipagus (spine), pynopagus (rump) and parapagus (side) (3). The term parapagus is recently introduced to include extensive "side to side" fusion. In this article, prenatal diagnosis of dicephalic parapagus case at 12th gestational week with ultrasound is reported.

CASE REPORT

Twenty-four-year-old pregnant woman with gravidity 2, and who had a healthy child was referred at 12th week of pregnancy. Her personal and familial background was uneventful. Neither the woman nor her husband had a family member with a history of twinning or congenital anomalies. The referral pre-diagnosis was conjoined twins. Obstetric ultrasonographic examination performed by Logiq P5 (transabdominal 3.5 MHz probe, General Electric) revealed a fetus with measurements consistent with intrauterin gestational age of 12 weeks 5 days having 2 heads and 2 necks on one body, 2 arms and 3 legs, conjoined two vertebral column (**Figure-1**).

Figure-1: *Image of ultrasound examination of fetuses.*

Ultrasonography showed single cardiovascular system, placenta, and umblical cord. These findings suggested a diagnosis of parapagus dicephalus conjoined twin. There was undetectable cardiac abnormality in fetal heart. However, increased nuchal translucency was detected in both neck as 5.4mm and 4.5 mm, respectively. The couple was informed in detail that this is possibly surgically inseparable twin pregnancy and there is a poor life expectancy. The case was discussed with the patient and her family; and the pregnancy was terminated by the written consent of the patient and her husband. The termination was performed by using misoprostol. Post abortive period was uneventful. After the termination, it was seen that the fetus had 2 heads, 2 necks, 2 arms, and 3 legs one of which fused pelvis in the macroscopic examination (**Figure-2**).

***Figure-2*:** *Image of fetuses after abortion*.

Fetal anatomic dissection was not performed because the family did not approve.

DISCUSSION

Conjoined twins are easily diagnosed by transvaginal or transabdominal ultrasonographic examination in the first trimester. In additionally, prenatal ultrasonography is capable of diagnosing conjoined twin pregnancies as early as 10th – 12th weeks of gestation (4,5). Ulker et al. reported a parapagus dicephalus twin diagnosed by tree-dimensional ultrasonography in 12th gestational week (6). Another parapagus twin case was presented to be diagnosed by two-dimensional ultrasonography in the 14th week of gestation (7). Our case was diagnosed as parapagus dicephalus in the 12th week of gestation by two-dimensional ultrasonography. Technological developments like high-resolution-ultrasonography have facilitated the detection of abnormalities during the first trimester. Magnetic resonance imaging can be performed prenatally to diagnose conjoined twins as parapagus dicephalus (8).

All conjoined twins are monochorionic and monoamniotic. Ultrasound images of one yolk sack - two fetuses or monoamniotic twin in any gestational week should arouse suspicion conjoined twins (9). The diagnosis of conjoined twins should be considered in any twin pregnancy that has a single placenta and no visible separating amniotic membrane. Polyhydroamnios occurs in as many of 50% conjoined twin pregnancies. The ultrasonographic findings in conjoined twins include inseparable fetal bodies and skin contours, and both fetal heads' persistent appearance at the same level (10). In the present case, there was a single placenta. Moreover, the fetus had two heads, two arms, and three legs. The name parapagus dicephalus implies completely separate two heads and two faces with single heart. Dibrachius dipus cases usually have one heart. However, the heart is duplicated or conjoined in most of the cases (11). The present twins had only one heart with undetectable abnormality. Probably, due to the presence of cardiac abnormality, nuchal translucency (NT) was detected increase. Lymphedemas such as hydrops, edema, cystic hygroma and increased NT are the most common anomalies associated with conjoined twins that are detected in the first trimester (12). Hemodynamic disturbance due to abnormalities of the heart may be partly associated with increased NT in conjoined twins (13,14).

The diagnosis of conjoined twins is suspected during the prenatal scan at the 12th week of gestation. It is important to define the anatomy of the union. Pregnancy termination would be recommended in the event of complex cardiac fusion or poor prognosis of separation surgery of conjoined twins, as in the case of parapagus. Delivery is always by cesarean section at 36-38 weeks' gestation. Harma et al. reported a parapagus dicephalus twins; stillborn conjoined twins case admitted to the clinic at the 38th gestational week without any previous follow-up and delivered vaginally with difficulty (15). In our case, we informed the family about the prognosis of the pregnancy and the family decided on termination. Then the pregnancy was terminated.

Fetal anomalies such as conjoined twins can be diagnosed as early as the first trimester. Parapagus dicephalus is a rare type of conjoined twins and it has severe structural and functional malformations. Early prenatal diagnosis of parapagus dicephalus twins would have a significant role in the management of pregnancy. After informing parents about the prognosis of fetal and neonatal survey, pregnancy can be terminated in some cases like gross malformation and especially heart abnormalities.

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