

Sefalotorakofagus : Olgu Sunumu

A Case Of Conjoined Twin's Cephalothoracopagus Janiceps Disymmetros

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ÖZET:

Çalışmamızda 15. haftada iki boyutlu ultrasonografi ile tanı konulmuş oldukça nadir görülen bir birleşik fetus vakası sunulmuştur. Bu olgu sunumunda zıt yönlere bakan belirgin ve simetrik iki yüzü olan, cinsiyeti erkek olan sefalotorakofagus olarak adlandırılan birleşik fetus vakasının öyküsü ve klinik özellikleri anlatılmıştır. Bu gibi yaşama şansı olmayan anomalili fetusların erken gebelik haftalarında tanınması hasta yakınlarına terminasyon seçeneğinin erken haftalarda sunulmasını sağlayarak anneleri gereksiz operasyonlardan koruyacaktır.

Anahtar Kelimeler: Çoğul gebelik, anormallikler, ultrasonografi, bileşik ikiz

ABSTRACT:

We report the prenatal imaging findings of a rare case of male cephalothoracopagus janiceps disymmetros diagnosed at 15 weeks' gestation by 2D ultrasound. Cephalothoracopagus janiceps refers to twins with two identical faces, on opposite sides of the large conjoined head, the two faces are identical and symmetrical, this is called cephalothoracopagus janiceps disymmetros. The early diagnosis of conjoined twinning with a high probability of mortality is very important and allows the termination of the pregnancy before 18-24 weeks. So the mother can be protected from a major surgical procedure.

Key words: Multiple pregnancy, abnormalities, ultrasonography, conjoined twins

INTRODUCTION

Conjoint twinning is a very rare obstetrical event. All conjoined twins are monozygotic, monochorionic and monoamniotic. Spontaneous twinning occurs in 1,6% of all human pregnancies, of which 1,2 % are dizygotic and 0,4 % are monozygotic (1). Monochorionic monoamniotic twins are less than 1% of monozygotic twins, and conjoint twins are even less common that is approximately 1 in 30.000-100.000 live births (2). Also, conjoined twinning is three times more common in female fetuses than in males(1). Conjoined twins are classified according to the most prominent side of fusion. Ventral unions occur 87% of conjoined twins and are classified as cephalopagus(11%), throcapagus(19%), omphalopagus(18%), ischiopagus(11%) and parapagus(28%). Dorsal unions occur in 13% of conjoined twins and are classified as; craniopagus(5%), rachiopagus(2%), and pygopagus(6%)(3). Cephalothoracopagus, a very rare form of conjoined twins, with a fused head and thorax

but two separate spines, limbs, and pelvis, occurs once in every 58 sets of conjoined twins or once in every 3 million births(4). We report the prenatal imaging findings of a rare case of conjoined twinning, a male cephalothoracopagus janiceps disymmetros diagnosed at 15 weeks' gestation.

A CASE REPORT OF CONJOINT TWIN

A 39- year old, multigravid woman (gravida 13, para 4, alive 4) visited the department of obstetrics and gynecology of Zeynep Kamil Hospital at the 15th gestational week. The patient was admitted to the hospital as a routine control for the first time. The patient's past medical history was unremarkable and she had not taken any medication. Two-dimensional (2D) transabdominal ultrasound scan revealed monocoryonic-monoamniotic twin with four legs, four arms, double vertebral colon each one on the lateral sides, union torax, abdomen,

and cranium at frontal sides, two urinary bladders, two cardiac activities, two cerebellum and two posterior fossa and the normal amount of amniotic fluid. According to these ultrasound findings, a cephalothoracopagus conjoined twin was diagnosed. The couple was informed that this was a surgically inseparable condition because of the gestational age and opted for pregnancy termination and **informed consent was obtained from the couple**. Medical abortion was performed after the day of admission to the hospital. There were two vertebral colon, 4 legs and 4 arms in the X-Ray image, too. After the abortion, the fetus was examined morphologically. It was 14,5 cms in length and the head circumference was 15 cm. It had got one head with two faces each face looking forward to the shoulders. Also the thorax and the abdomen were joined up to the umbilicus(**Figure1**).

Figure 1: Postmortem photograph demonstrating anterior aspect of cephalothoracopagus janiceps disymmetros.



From this level downwards fetuses were separated. Each one had two legs and two arms, one vertebral colon and had male external genitalia (**Figures 2 and 3**).

Figure2: Postmortem photograph demonstrating total length and male gender of twins.



Figure 3: Postmortem photograph demonstrating legs and arms of twins.



The extremities were morphologically normal. There was one umbilical cord with two arteries and one vein. In the postmortem autopsy examination, it was found that the cranium had two foreman magnum, two brain stem and two cerebellum. The cranium was not separated at the front. Furthermore the cerebral hemispheres were not separated. The brain had only one ventricul and there was no corpus callosum. Optic nerves were normal without chiazma opticum and other cranial nerves were not developed normally. As the conjoint thorax was opened each thorax had two lungs and two hearts and each had vertebral colon at the back. As the abdomen opened, macroscopically there were two livers, two intestines, two colons and two apendix. On the other hand there was only one osepagus, one stomach and one duodenum. Also, there were two spleens, two pairs of kidneys, adrenal glands, and ureters, two urinary bladders and two pairs of testis.

DISCUSSION

Conjoined twins are rare and complex complication of monozygotic twins. In our case which is male cephalothoracopagus janiceps disymmetros, the frequency is much more rare. Cephalothoracopagus janiceps refers to twins with two identical faces, on opposite sides of the large conjoined head, but in many cases (75%) one of the two faces is rudimentary. When the two faces are identical and symmetrical, this is called cephalothoracopagus janiceps disymmetros (4-8). The term janiceps is derived from Janus, the two-faced Roman god(9). Thorax and upper abdomen are always fused, the lower

abdomen and pelvis never. In addition, more than %70 of conjoined twins are female(1). The same ratio appears similar in cephalopagus cases. Spencer (10) studied 59 cases of typical cephalopagus of which 14 (24%) were male. Taruffi (11) collected 23 case histories of typical cephalopagus of which 4 (17%) were male. The present case is typical of male cephalothoracopagus janiceps disymmetros which was diagnosed at 15 weeks of gestation. A similar case which was a male cephalothoracopagus janiceps disymmetros at 30 weeks and 24 weeks was reported by Hovorakova et al(12) and Khanna et al.(13), respectively. Also, Chen et al.(14) reported a case which was a female cephalothoracopagus janiceps disymmetros at 28 weeks. However, Kuroda et al.(15) reported a case of cephalothoracopagus janiceps twinning visualised by 3D ultrasound at 13 weeks of gestation. Their case had a large conjoined head with two faces, one of the faces was partially developed while the other was rudimentary and had female genitourinary tracts. Similarly, Biswas et al.(16) detected a case of cephalothoracopagus janiceps twinning at 13 weeks gestation by 3D ultrasound with one of the faces rudimentary and unformed external genitalia. So, our case appears first male cephalothoracopagus janiceps disymmetros which was determined in early pregnancy.

Survival of conjoined twins depends largely on the side of conjoining and the organs involved. The prognosis for cephalothoracopagus twins is extremely poor because surgical separation is not an option. After 18-20 weeks of gestation transvaginal termination will be difficult and delivery could require a major surgical procedure that is hysterotomy or caesarean section(17). Thus, the early diagnosis of conjoined twinning with a high probability of mortality is very important and allows the termination of the pregnancy before 18-24 weeks. As the increasing use of ultrasound in the first trimester screening most of the conjoined twins are diagnosed early in pregnancy. 2D ultrasound gave the initial diagnosis of conjoined twins. Because of all conjoined twins are monochorionic and monoamniotic, during the first trimester

ultrasound in twin pregnancies presence of only one yolk sac and absence of intertwin membrane should be suspected of conjoined twin. But, a false-positive diagnosis of conjoined twins in the first trimester may occur because of the close proximity of the fetuses(18). If the parents opt for a conservative prenatal management or postnatal surgery, the diagnosis is confirmed by 3D ultrasound. 3D ultrasound add more anatomic information and is valuable to improve the classification. Also, other imaging techniques such as MRI will provide more accurate and valuable information for the patients who desire postnatal surgery(17). In our case conjoining sides and shared cerebral structures were so extended, the patient opted for termination and was delivered vaginally. So detailed diagnosing techniques were not used in this case. A similar case which was a cephalothoracopagus was reported by Kokcu et al. (19). It was diagnosed at 34 weeks of gestation when the patient was first admitted to the hospital. So the pregnancy was terminated by caesarean section and the neonate died twenty minutes following delivery. However, Kuroda et al.(15) and Biswas et al.(16) reported cases of cephalothoracopagus janiceps twinning visualised by 3D ultrasound at 13 weeks gestation.

As a result, we emphasize the importance of early prenatal diagnosis for counselling the parents about the termination or continuation of the pregnancy and about the potential delivery type and possible surgical solution. So the mother can be protected from a major surgical procedure.

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