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THORACOPAGUS DIAGNOSED AT 12 WEEKS OF GESTATION – CASE REPORT

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Conjoined twins is a rare congenital malformation of unknown etiology with severe mortality and morbidity.

The most common type of conjoined twins is thoracopagus. The ultrasound examination is important to diagnose this rarely seen form of twin gestation. Making an early diagnosis with ultrasound examination gives the patient the option for pregnancy termination. In this report we are representing a case of 12 weeks pregnant primigravida with ultrasound detected thoracopagus. The pregnancy was terminated. The ultrasound examination has an important role because a lot of congenital malformations can be diagnosed early by it.

Key words: thoracopagus, conjoined twins, ultrasound examination

TORAKOPAGUS DIJAGNOSTIKOVAN U 12. NEDELJI GESTACIJE – PRIKAZ SLUČAJA

Sijamski blizanci su retka urođena malformacija nepoznate etiologije sa teškim mortalitetom i morbiditetom.

Najčešći tip spojenih blizanaca je torakopag. Ultrazvučni pregled je važan za dijagnozu ovog retko viđenog oblika blizanačke gestacije. Rana dijagnoza ultrazvučnim pregledom daje pacijentkinju mogućnost prekida trudnoće. U ovom izveštaju predstavljamo slučaj primigravida u 12 nedelja trudnoće sa ultrazvučno otkrivenim torakopagusom. Trudnoća je prekinuta. Ultrazvučni pregled ima važnu ulogu jer se njime mogu rano dijagnostikovati mnoge urođene malformacije.

Ključne reči: torakopagus, sijamski blizanci, ultrazvučni pregled

Introduction

Conjoined twins are extremely rare congenital malformation. They are symmetrical monozygous twins with incidence of 1:50,000 and three times commoner in females (1) with 100% of placental, vascular anastomosis and high rate (70-90%) of perinatal mortality. Conjoined twins are also named Siamese twins after Chang and End Bunker brothers born in 1811 in Siam (Thailand) they lived 63 years and worked in a circus (2). Thoracopagus is the most common form of conjoined twins.

Thoracopagus twins share heart and usually have severe cardiac anomalies. This anomaly has high rate of mortality.

Ultrasound is the primary imaging model for the early diagnosis. After diagnosis of this rare condition parents are consulted about poor prognosis and termination option (3). Although surgical techniques have been improved survival and success after separation on cardiac fusion is still low.

Termination of pregnancy is the best treatment. We represent a case of conjoined twins diagnosed by prenatal ultrasound at 12 weeks of gestation.

Case report

A 21-year-old primigravid woman was admitted to the Clinic at an 12-week gestation

because of a conjoined twin (thoracopagus) diagnosed by ultrasonography. Her last menstrual date was 12.2 weeks ago. She had no personal or family history of twins. Her physical examination and laboratory testing were normal. Sonography was performed two dimensional ultrasound images presented one yolk and two fetuses with 2 arms, 4 legs, 2 heads, hygroma cysticum and omphalocele 28 x 20 mm in size. The BPD of first twin was 24 mm and HC 66 mm, the BPD of the second twin was 20 and HC was 68 mm. The twins were joined at the thorax and upper abdomen. There were observed a single umbilical cord, and only one fetal heart. The placenta was localized anterior wall and one artery and one vein were seen in the umbilical cord. On the basis of these findings, the diagnosis of thoracopagus, conjoined twins was made (Figure 1, Figure 2, Figure 3) and the patient were informed about the malformation and the twins' high chances of perinatal mortality. The patient desired to terminate the pregnancy. Patient wrote a letter of permission for the termination of pregnancy to the Medical Ethics Committee and it was approved. The expulsion of foetuses occurred after the end of one cycle of prostin E2, with both twins intact (Figure 4). After that curettage was performed. Foetuses were sent for further autopsy. Patient was discharged home day after. The pathology report confirmed the diagnosis of intact thoracopagus conjoined twins with a single heart.

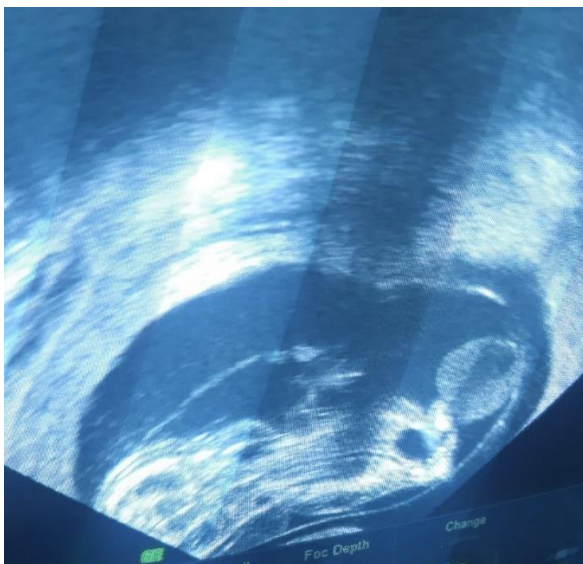


Figure 1.

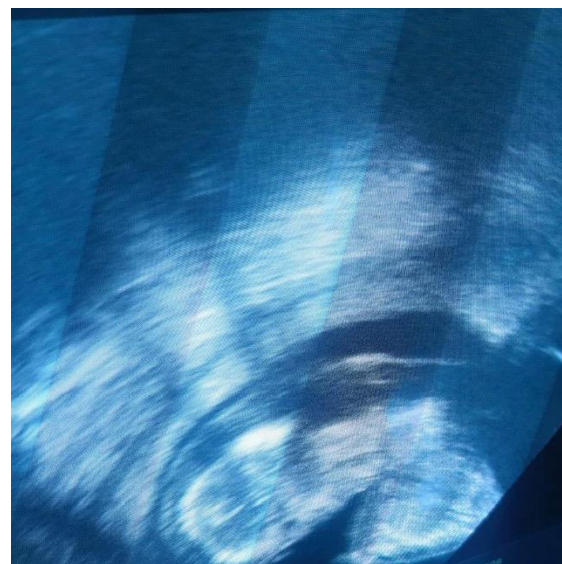


Figure 2.

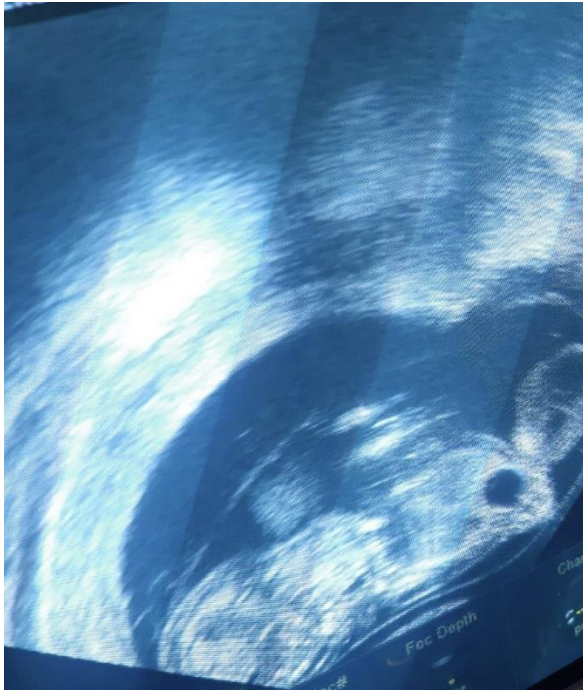


Figure 3.



Figure 4.

Discussion

Conjoined twins are monochorionic mono-amniotic twins. Its etiology is unknown. The precise mechanism of pathogenesis has not been established. There are different theories of pathogenesis, two theories are most such as incomplete separating of one embryo into two more than 13 days after fertilization - fission theory and other describes fusion of two embryonic plaques from a monozygotic pair into the other - fusion theory (4).

Conjoined twins are classified according to the most prominent site of conjunction which can be on any pole and characteristics depend on which body parts are shared. The suffix "pagus" is added which in Greek language means fixed (5). The fusion can be ventral and dorsal. Ventral can be rostral on: thorax (thoracopagus), abdomen (omphalopagus), cranium (cephalopagus); caudal pelvis (ischiopagus) and lateral- parapagus dispropus and parapagus dicephalus. Dorsal can be skull Craniopagus, sacrum (pygopagus), and back (rachipagus). Severely deformed connection is fatal (6).

Thoracopagus is the most common (19%) (7).

In thoracopagus shared parts are Sternum, diaphragm and anterior abdominal wall, 90% of

thoracopagus twins share one heart. Severity of forms depends on components of the hearts that are shared. The most severe form is a shared heart with one ventricle and the mildest form includes two hearts with single shared pericardium (8). It is estimated that up to 60% of cases die in utero, approximately 35% are stillborn or die within the first 24 hours after birth (9).

In the first trimester the ultrasound prenatal evaluation of conjoined twins due to the increased risk of perinatal morbidity and mortality but identification is usually in the second trimester, when ultrasound evaluation can confirm the diagnosis by describing detailed anatomy of the two fetuses and the parts shared. During examination fetal poles are associated without changing positions from one other. MR imaging can clarify shared organs.

Conclusion

Thoracopagus has low incidence but high rate of mortality. Termination of pregnancy is the wisest choice so ultrasound examination as a noninvasive method is helpful for early diagnosis because it permits the abortion to be performed at early stage of pregnancy.

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