CASE REPORT

Eastern Journal of Medicine
East J Med 25(2): 318-320, 2020
DOI: 10.5505/ejm.2020.24861

Thoracopagus: A Case of Conjoined Twins

Gökçe Naz Küçükbaş[1*], Gurcan Turkyilmaz[2], Güler Şahin[1]

[1]Van Yuzuncu Yil University, Obstetrics and Gynecology and Perinatology Department
[2]Van Region Education and Research Hospital, Perinatology Department

ABSTRACT

Conjoined twins originate from abnormal divergence of the embryonic plaque after 12th day of fertilization causing shared organ systems. The prevalence varies between 1/50.000 and 1/200.000 and diagnosis is possible by advanced ultrasonographic imaging techniques. Despite limited number of successful cases in the literature, conjoined twins have poor prognosis and management usually requires termination. In this study we present a 22 year-old, nulliparous woman in 14th gestational week without specific obstetric history diagnosed with thoracopagus.

Key Words: Thoracopagus, conjoined twins, monochorionic monoamniotic

Introduction

Conjoined twins (CT) occur when a monozygotic monochorionic monoamniotic embryonic plaque diverges after 12th day in utero. This leads to organ and tissue sharing (1). Incidence is reported between 1/50.000 and 1/250.000 (1-4). Female twins carry more risk than males (OR is 3:1) (2, 4). There are two distinct etiopathogenic theories of aberrant embriogenesis of CT: Fusion of two same embryonic plaques (fusion theory) and incomplete divergence of an embryonic plaque between 15 and 17th day after fertilization (fission theory). Fission theory is widely accepted rather than fusion theory (1, 4, 5). CT is cathegorized in 8 types by Spencer: Cephalopagus, Thoracopagus, Omphalopagus, Ischiopagus, Pigopagus, Parapagus, Craniopagus and Rachypagus (6). The most common type of CT is thoracopagus (40-60%) (5). ISternum, diaphragm and anterior abdominal wall is common structures in thoracopagus. 90% of thoracopagus twins share one heart and according to the components of the heart that is shared, thoracopagus is graded. The most severe form is a shared heart with one ventricle whereas the mildest form includes two hearts with single shared pericardium (3).

Stillbirth complicates half of the CTs and newborns have severe complicated malformations(5). The mild forms undergo separation surgeries and survival rate is 25% (1). These surgeries are accomplished with a multidisciplinary team consisting surgeons of pediatric, plastic, cardiovascular, ear nose throat divisions and newborns are followed in newborn intensive care units specialized for CT surgeries. Preoperative study starts with a 3D-printing technology of the CTs after postnatal imaging is obtained and multidisciplinary team approach surgery is performed (7).

Only rare cases undergo seperation surgery whereas majority of CT pregnancies are offered termination when diagnose was made within 1st trimester. This case presents a CT pregnancy diagnosed at 14 weeks of gestation and its management.

Case Report

22 year old, nulliparous woman was referred to our clinic at 14 weeks of gestation diagnosed with monochorionic monoamniotic twins. Parents were not consanguineous and medical histories were non-specific. Sonographic examination revealed conjoined twins, with separate extremities but a common thorax, one heart involving 2 atria and 2 ventricles. One of the twins had scoliosis (Figure 1). Fetal MRI confirmed conjoined twins with a common heart. Parents were informed about the anomaly and poor prognosis and termination of pregnancy. Postmortem examination revealed conjoined twins sharing one heart and liver within common thoracal cavity (Figure 2)

Discussion

Conjoined twins also known as Siamese Twins after Chang ve Eng Bunkler brothers born in 1811 in Thailand. They lived 63 years and worked in an international circus (6). Conjoined twins have various types and categorization is made by adding the suffix -pagus after common body part. Pagus means being

***Corresponding Author:** Gökçe Naz Küçükbaş, MD, Van Yuzuncu Yil University, Department of Obstetrics and Gynecology and Perinatology
E-mail: nazkuc@gmail.com, Phone: 0506 281 70 78
Received: 19.11.2019, **Accepted:** 11.03.2020

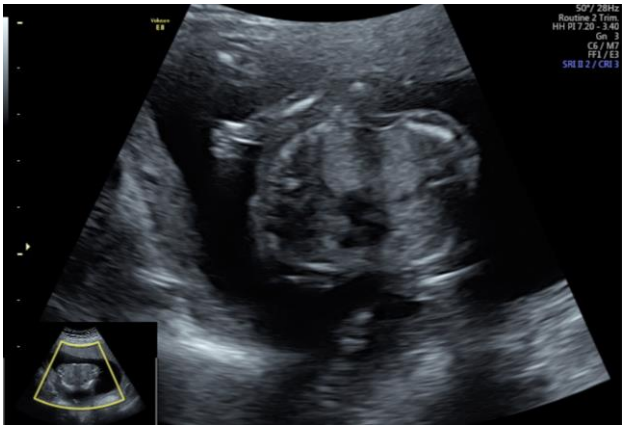

Fig. 1. A shared heart and thoracal cavtity were observed sonographically

joined in Latin (8). Thoracopagus describes conjoined twins sharing same thoracal cavity. CT joining occurs 87% ventrally and 13% dorsally (8).

Diagnosis of CT should be made prenatally because it is vital for maternal health. There is one CT case in Sierra Lione -a Subsaharan African country- which was not defined antenatally and resulted in increased maternal morbidity. This pregnancy was an unfollowed case, hospitalized for delivery and shoulder dystocia occurred during spontaneous vaginal birth. Although two shoulders were born after all maneuver birth could not be completed. Secondary vaginal examination revealed suspicion of another fetus and an emergency cesarean section was planned. Thoracopagus was diagnosed and the newborns did not survive (5). Undiagnosed conjoined twins have high risk of maternal and fetal morbidity and mortality.

Monochorionic monoamniotic twins constitute 1% of monozygotic twins and conjoined twins constitute 1% of monochorionic monoamniotic twins (8). Prenatal diagnosis is possible although this condition is rarely seen (1). Definite CT diagnosis is possible after 10 weeks of gestation as fetal movements are restricted below that gestational age (9).

Detection of associated fetal body parts, fixed positioning of both fetuses during several examinations are key features to diagnose CT under sonography. Joined fetal body parts define CT type. Fetal anomaly scan and fetal echocardiography examinations should be performed on ongoing CT. Organ systems that are separate may also contain anomaly and be examined with care (10). Magnetic resonance imaging (MRI) is superior in common tissue definition and not contraindicated in fetal period. Diagnosis confirmation and further investigation is preferably completed by fetal MRI (11). Karyotyping is not usually recommended as chromosomal anomaly risk is low in CT (10). Therefore in this case karyotyping was not performed.

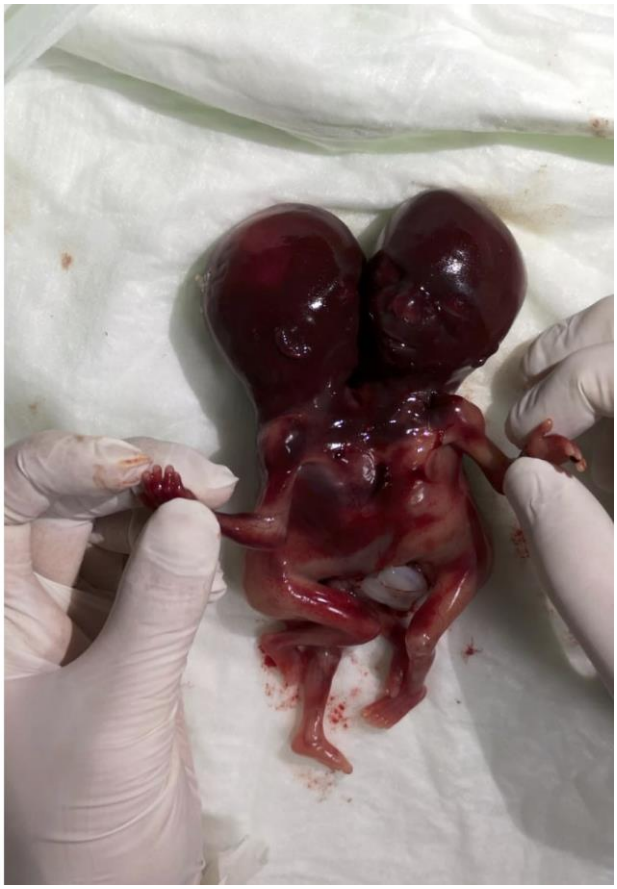

Fig. 2. Postmortem material of thoracopagus twin pregnancy

Half of the ongoing CT pregnancies end up with intrauterine demise and another 44% of CTs is lost in neonatal period (9). In a study combining 14 CT cases, intrauterine demise risk was 28%, postpartum death occurred in 54%, and 18% of CTs had separation surgery but half of them could not survive (10).

There are some multidisciplinary centres performing separation surgeries and utilizing specially designed intensive care units for CT. Survival rates are higher in CTs which do not share vital organs, however high risk of hypovolemic shock and sepsis (because of large skin defects) deteriorate survival (9, 12).

Malformations and high mortality and morbidity risks of CT surgery lowers survival rates. Parental consultation usually leads to decision of pregnancy termination (9). In this case, after detailed consultation, parents agreed on termination of the pregnancy.

Thoracopagus is a rare condition usually causing vital organ sharing -the heart- among twins. It is a rare condition and after diagnosis parents are consulted about poor prognosis and termination option. There is need of advanced approach in surgical procedures and specialized neonatal intensive care units in order to increase survival rates of conjoined twins.

References

1. Alkhateeb M, Mashaqbeh M, Megableh S, et al. Early prenatal diagnosis of thoracopagus twins by ultrasound. Acta Inform MEd 2015; 23: 60-62.
2. Goo HW, Park JJ, Kim EAR. et al. Cardiac fusion and complex congenital cardiac defects in thoracopagus twins: diagnostic value of cardiac CT. Pediatr Radiol 2014; 44: 1169.
3. Mathew RP, Francis S, Basti RS, et al. Conjoined twins - role of imaging and recent advances. J Ultrason 2017; 17: 259-266.
4. Mian A, Gabra NI, Sharma T. et al. Conjoined twins: From conception to seperation, a review. Clin Anat 2017; 30: 385-396.
5. Leigh MB, John-Cole V, Kamara M, et al. A Triple Obstetric Challenge of Thoracopagus-Type Conjoined Twins, Eclampsia, and Obstructed Labor: A Case Report from Sub-Saharan Africa. Case Rep Obstet Gynecol 2017; 2017: 6815748.
6. Kobylarz K. History of treatment of conjoined twins. Anaesthesiology Intensive Therapy 2014; 46: 116-123.
7. Fallon SC, Olutoye OO. The surgical principles of conjoined twin separation. Semin Perinatol 2018; 42: 386-392.
8. Graham GM, Gaddipati S. Diagnosis and management of obstetrical compliacations unique to multiple gestations. Seminars in Peinatology 2005; 29: 282-295.
9. Pajkrt E, Jauniaux E. First-trimester diagnosis of conjoined twins. Prenatal Diagnosis 2015; 25: 820-826.
10. MacKenzie TC, Crombleholme TM. The natural history of prenatally diagnosed conjoined twins. J Pediat Surg 2002; 303-309.
11. Norwitz ER, Hoyte LP, Jenkins KJ, et al. Separation of conjoined twins with the twin reversed-arterial-perfusion sequence after prenatal planning with three-dimensional modeling N Engl J Med 2000; 399-402.
12. Kobylarz K. Intensive care of conjoined twins. Anaesthesiology Intensive Therapy 2014; 46: 130-136.