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Case Report

Thoracopagus conjoined twins diagnosed by sonography

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Abstract: Objective: Conjoined twins are identical twins with congenital anomaly and severe mortality. The more common types include the thoraco-omphalopagus, fused at chest, and thoracopagus type, fused from the upper thorax to lower belly, at the chest. Here we are reporting one case of thoracopagus conjoined twins diagnosed by sonography. A 23-year-old woman with gravida 1 admitted to our clinic was diagnosed as thoracopagus conjoined twins shared heart with multi-malformations, intestinal dilatation, and the edge of umbilical cord inlet by ultrasonography at an 25-week gestation. Termination of the pregnancy and autopsy was performed. Conclusion: Prenatal sonography is important for discovering and identifying fetal malformations and enable parents to elect pregnancy termination.

Keywords: Thoracopagus conjoined twins, shared heart, fetal ultrasound

Introduction

Conjoined twining is one of the rare human malformations, with the incidence ranges from 1 in 45,000 to 1 in 200,000 live births and somewhat higher incidence in Southwest Asia, Africa and Brazil [1]. Approximately 75% of conjoined twins are female, and 70% are fused at the thorax (thoracopagus) or abdomen (omphalopagus) [2]. Considering that 40% of conjoined twins are not alive when they are delivered (stillborn) and 35% die within a day after they are born [3], early diagnosis and management of delivery is extremely important. The present study will present a case of conjoined twin with thoracopagus type and discuss the role of ultrasound in early diagnosis and management.

Case report

A 23-year-old woman, gravida 1, para 0, aborta 0, was referred to our university hospital in her 25-week of pregnancy because her prenatal sonography suggested a diagnosis of conjoined twin. She did not use prescribed medications, alcohol, tobacco, or illicit drugs. She was not involved in radiation or chemical contact. Her pregnancy was spontaneous, and her medical and family histories were unremarkable. No abdomen pain and vaginal bleeding was noticed. Neither she nor her husband had a family member with a history of twinning or congenital anomalies.

Sonography was performed and two fetuses with 4 arms, 4 legs, and 2 heads were visualized. The twins were joined from upper thorax to abdomen (**Figure 1A**). Only one fetal heart with multi-malformation was observed (**Figure 1B**). Yet each fetus has individual spinal (SP), liver, stomach, 2 kidneys and bladder. Intestinal expansion shows a maximum diameter of 22.7 mm (**Figure 1D**). The placenta was localized rear, and one artery and one vein were seen in the umbilical cord. Entrance on the edge of the umbilical cord from the placenta is approximately 1.2 cm. Based on these observations, the diagnosis of thoracopagus conjoined twins, fetal heart multi-malformations with intestinal dilatation, the edge of umbilical cord inlet (racket-shape placenta) were made.

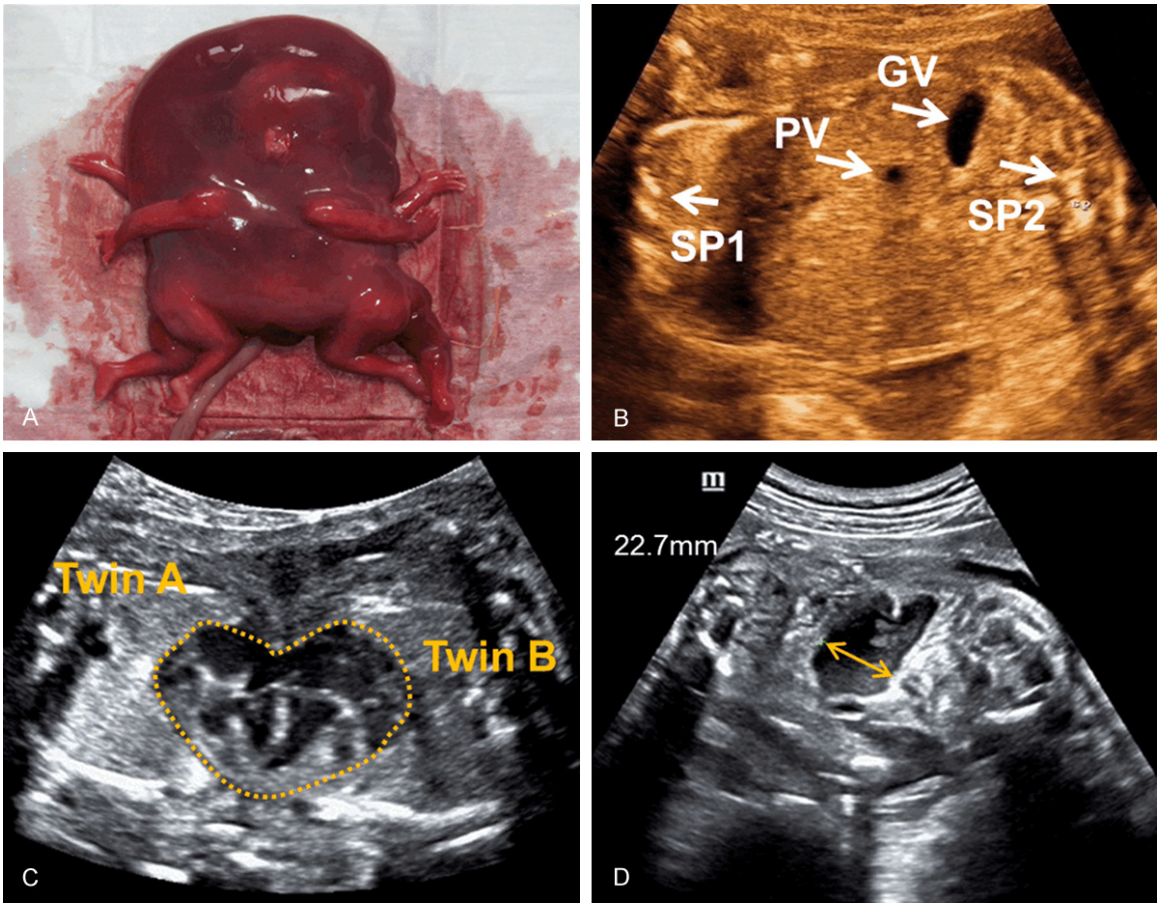

Figure 1. Image and sonography of the thoracopagus twin. A. Twins were joined from upper thorax to abdomen. B. SP Arrow: spinal; SB Arrow: stomach bubbles; PV Arrow: portal vein. C. The shared heart shows multi-malformation. D. Intestinal expansion shows a maximum diameter (Arrow) of 22.7 mm.

After diagnosis, the parents were informed about the situation and the poor chance for survival. They decided to terminate the pregnancy. A written informed consent was attained and the procedure was approved by the Medical Ethics Committee. Two days later, a vaginal delivery of the conjoined twins was achieved after induction of labor with prostaglandin. No complication was observed. Postmortem and autopsy examination revealed thoracopagus conjoined twins with chest and abdomen fusion, and heart and intestinal malformations (**Figure 1**).

Discussion

Conjoined twins are rare, and classified according to the most prominent site of conjunction: thorax (thoracopagus), abdomen (omphalopagus), sacrum (pygopagus), pelvis (ischiopagus), skull (cephalopagus), and back (rachipagus), among which thoracopagus (19%) ranks the most common type [4]. Conjoined twins only arise when the twinning event occurs at the primitive streak stage of development, approximately 13 to 14 days after fertilization [5]. However, its etiology is unknown.

Currently there are two contradicting theories to explain the origins of conjoined twins [6]. The traditional one is fission, in which the fertilized egg splits partially after day 12 of fertilization and the delayed separation of the embryonic mass form conjoined twins. The other one is fusion, in which the fertilized egg completely separates, yet stem cells seek stem cells-like on the other twin and fuse the twins together. Whether conjoined twinning is a result of fission or fusion is still much debated.

Fetal ultrasound has become a routine part of prenatal care in most of country, which due to

the fact that it is a low risk procedure that provides valuable information with relative ease [7]. General progress during a pregnancy can be efficiently evaluated using ultrasound, for example, placement of the placenta, multiple birth pregnancies, nuchal translucency screening, etc [8]. Early ultrasound assessment in twins allows planning of appropriate management of these pregnancies. Accurate determination of chorionicity is crucial to stratify the obstetric risk and to plan further scans [9]. Particularly, the number of placental in determining chorionicity in first-trimester diagnosis of chorionicity reported a sensitivity and specificity of 100% and 99.8%, respectively during 11 and 14 weeks of gestation [10]. Ultrasound also provides vital information about pregnancy dating and twin labeling [11]. Intriguingly, conjoined twin was previously reported as early as 11 week of gestation diagnosed by sonography [6]. In our case, the patient has no clinical syndrome and was not diagnosed until her first prenatal ultrasound checking at 25 weeks of gestation. This situation probably is mainly due to her lack of prenatal care knowledge and partially due to her social economic status. Nevertheless, the ultrasound diagnosis was effective and enabled her to determine the pregnancy outcome. Sadly, it's not uncommon for peasants living in rural area of China, which consist of approximately 50% population. These population tend to avoid prenatal ultrasound check until late pregnancy or unexpected syndrome shows up. That's why we presented this case to all clinical doctors. Even in late 25 weeks of gestation, sonography could bring benefit to patients. Though MRI offers more detailed assessment of fetal pathology in cases of twin pregnancies, including conjoined twins [12], in which sonographic evaluation is more difficult than in single cases, the method requires more expensive and delicate equipment.

In conclusion, prenatal sonography is important for discovering and identifying fetal malformations especially for conjoined twins which are associated with a high perinatal mortality. Early diagnosis with ultrasonographic examination of conjoined twins enables parents to elect pregnancy termination.

Disclosure of conflict of interest

None.

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References

[1] Denardin D, Telles JA, Betat Rda S, Fell PR, Cunha AC, Targa LV, Zen PR and Rosa RF. Imperfect twinning: a clinical and ethical dilemma. Rev Paul Pediatr 2013; 31: 384-391.

[2] Mankonen H, Seikkula J, Jarvenpaa T and Jokimaa V. A case of conjoined twins after a transfer of a multinuclear embryo. Clin Case Rep 2015; 3: 260-265.

[3] Alkhateeb M, Mashaqbeh M, Magableh S, Hadad R, Nseer Q and Alshboul A. Early prenatal diagnosis of thoracopagus twins by ultrasound. Acta Inform Med 2015; 23: 60-62.

[4] Mutchinick OM, Luna-Munoz L, Amar E, Bakker MK, Clementi M, Cocchi G, da Graca Dutra M, Feldkamp ML, Landau D, Leoncini E, Li Z, Lowry B, Marengo LK, Martinez-Frias ML, Mastroiacovo P, Metneki J, Morgan M, Pierini A, Rissman A, Ritvanen A, Scarano G, Siffel C, Szabova E and Arteaga-Vazquez J. Conjoined twins: a worldwide collaborative epidemiological study of the International Clearinghouse for Birth Defects Surveillance and Research. Am J Med Genet C Semin Med Genet 2011; 157C: 274-287.

[5] Wu Y, Lv Q, Xie MX, Wan LY, Ren PP, Ge A and Ge S. Fetal echocardiographic characteristics of fused heart in thoracopagus conjoined twins. Echocardiography 2014; 31: E218-221.

[6] Osmanagaoglu MA, Aran T, Guven S, Kart C, Ozdemir O and Bozkaya H. Thoracopagus conjoined twins: a case report. ISRN Obstet Gynecol 2011; 2011: 238360.

[7] Mahalingam S and Dighe M. Imaging concerns unique to twin pregnancy. Curr Probl Diagn Radiol 2014; 43: 317-330.

[8] Simpson LL. Ultrasound in twins: dichorionic and monochorionic. Semin Perinatol 2013; 37: 348-358.

[9] Carter EB, Bishop KC, Goetzinger KR, Tuuli MG and Cahill AG. The Impact of Chorionicity on Maternal Pregnancy Outcomes. Am J Obstet Gynecol 2015; 213: 390, e1-7.

[10] Blumenfeld YJ, Momirova V, Rouse DJ, Caritis SN, Sciscione A, Peaceman AM, Reddy UM, Varner MW, Malone FD, Iams JD, Mercer BM, Thorp JM Jr, Sorokin Y, Carpenter MW, Lo J, Ramin SM and Harper M. Accuracy of sonographic chorionicity classification in twin gestations. J Ultrasound Med 2014; 33: 2187-2192.

[11] Papageorghiou AT, Kennedy SH, Salomon LJ, Ohuma EO, Cheikh Ismail L, Barros FC, Lambert A, Carvalho M, Jaffer YA, Bertino E, Gravett MG, Altman DG, Purwar M, Noble JA, Pang R, Victora CG, Bhutta ZA and Villar J. International standards for early fetal size and pregnancy dating based on ultrasound measurement of crown-rump length in the first trimester of pregnancy. Ultrasound Obstet Gynecol 2014; 44: 641-648.

[12] Bekiesinska-Figatowska M, Herman-Sucharska I, Romaniuk-Doroszewska A, Jaczynska R, Furmanek M and Bragoszewska H. Diagnostic problems in case of twin pregnancies: US vs. MRI study. J Perinat Med 2013; 41: 535-541.