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Conjoined Twins Having Cranio-Thoraco-Omphalopagus along with Mild Polyhydramnios

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Abstract

Conjoined twins, sometimes known as Siamese twins, are a rare kind of congenital human malformation. Conjoined twins are an extremely rare and rarely encountered impediment to monozygotic twinning, occurring in 1 in 100,000 to 1 in 250,000 live births. Conjoined twins share the same genes and are of the same sex. Reports have indicated a 3:1 female predominance. A 17-year-old woman at 23 weeks of gestation was referred at her second trimester of pregnancy from a rural area. Ultrasonographic evaluation revealed the presence of a monochorionic, monoamniotic twin pregnancy. The twins conjoined with a single face, single thorax and abdomen with separate upper limb and lower limb. It was finally diagnosed as a case of cranio-thoraco-omphalopagus conjoined twins of about 23±1 weeks with mild polyhydramnios. A conjoined twin is a well-known rare entity. It is very important to make a diagnosis by ultrasound in time to prevent future complications. Thoraco-omphalopagus type is known to present several cardiac anomalies. Thus, early diagnosis of conjoint twins, details of organ sharing by imaging modalities will help to predict prognosis, aid in patient counseling and further management.

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Introduction

Conjoined twins, sometimes known as Siamese twins, are a rare kind of congenital human malformation. Conjoined twins are an extremely rare and rarely encountered impediment to monozygotic twinning, occurring in 1 in 100,000 to 1 in 250,000 live births. They are more common in populations that are not Caucasian.[1] Miscarriage or pregnancy termination is the cause of many of these pregnancies not achieving a viable gestational age.[2-4] Early discovery and treatment are essential because more than 70% of conjoined twins pass away within 48 hours of birth or have a catastrophic congenital defect. Because of the high risks of stillbirth and neonatal mortality, only 7-8 sets of conjoined twins make it to the point of surgical separation each year.[4,5]

The term "Siamese twins" describes a set of conjoined twins named after Siam, where they were born in 1811. The lifespan of both males was over sixty-two years.[6] Conjoined twins are unrelated to mother age or parity and do not exhibit any genetic predisposition. It is the outcome of monozygotic twins' incomplete

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division after twelve days of development. An ultrasound during week seven of pregnancy could provide a diagnosis.[7,8] Conjoined twins share the same genes and are of the same sex.[9] They develop from the same fertilized egg and share the same placenta and amniotic sac. There have been indications of an increased occurrence in certain parts of Africa and Southeast Asia, ranging from 1:14,000 to 1:25,000.[10] Evidence showed that 40–60% of stillbirths occur from conjoined twins, and only 35% of conjoined twin births who survive past the first 24 hours; it also indicated a 3:1 female predominance.[11] Based on the region of conjunction, conjoined twins can be categorized into seven well-known categories. The question of whether conjoined twins develop from the fusion or fission of monoamniotic twins is up for debate, particularly with regard to dorsal or ventral conjoining.[12] Classification is typically based on the most conspicuous point of attachment, which can be found in the thorax (thoracopagus; 30-40%), abdomen (omphalopagus; 25-30%), sacrum (pygopagus; 10-20%), pelvis (ischiopagus; 6-20%), head (craniopagus; 2-16%), face (cephalopagus), or back (rachipagus).[13-15] Conjoined twins have a high rate of morbidity and mortality, thus early detection is crucial. It is important to screen all monozygotic twins for the risk of conjointment. The two most common types of conjoined twins are thoracopagus and omphalopagus twins. Here we present a case of a pregnant woman having conjoined twins with cranio-thoraco-omphalopagus along with mild polyhydramnios.

Case History

A 17-year-old woman at 23 weeks of gestation was referred at her second trimester of pregnancy from a rural area. There was no family history of twinning on maternal side, and there was no history of medication or radiation exposure. Ultrasound was never done till date. Our initial assessment involved taking abdominal examination and ultrasonogram. On abdominal examination, fundal height was more than the period of amenorrhea corresponded to 23 weeks gestational age. Multiple fetal parts felt. On vaginal examination, cervical os was uneffaced and closed. Ultrasonographic evaluation revealed the presence of a monochorionic, monoamniotic twin pregnancy. The twins conjoined with a single face, single thorax and abdomen with separate upper limb and lower limb (Fig. 1). It was finally diagnosed as a case of cranio- thoraco-omphalopagus conjoined twins of about 23±1 weeks with mild polyhydramnios. An elective abortion was performed after counseling with the patient and her family (Fig. 2A & 2B).

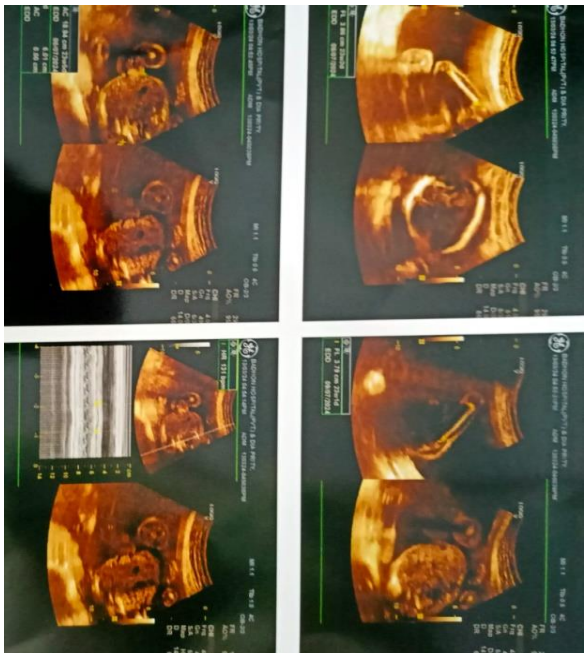

Fig. 1: Ultrasonogram of the uterus showing monochorionic, monoamniotic twin pregnancy with a single face, single thorax and abdomen with separate upper and lower limbs.

A B

Fig. 2A & 2B: Front and back images of the conjoined twins

Discussion

Conjoined twins are always of the same sex since they are always monozygotic, monochorionic, and monoamniotic.[13] A positive history of twin deliveries, ovulation induction medicine use, exposure to damaging radiation, and infertility therapy are risk factors that have been indicated to have a possible impact on the development of this unusual condition.[14] However, none of these risk factors were addressed by the history, family, or pregnancy history of our case. Moreover, there is no evidence linking the appearance of CTs to any demographic, genetic, environmental factor, or to the aging of mothers.[1] Even while most recorded occurrences of stillbirths in CTs were male fetuses, female fetuses are likely to experience CTs three times more frequently.[15]

There are several different types of conjoined twins, including thoraco-omphalopagus (joined at the chest, abdomen, or both), thoracopagus (joined at the thorax), omphalopagus (joined at the anterior abdominal wall), syncephalus (joined twins with one head), and ischiopagus (joined at the buttocks). Among all these, thoracopagus, omphalopagus, and thoraco-omphalopagus are the most common types, accounting for nearly 70% of conjoined twins.[16,17] The rarest type is omphalocaparus. The most precise method available for diagnosing it is ultrasonography. Premature fetal movements can lead to confusion between monochorionic twins and conjoined twins during the early weeks of pregnancy. Therefore, to confirm the diagnosis, a second scan should be performed in the eleventh and twelfth weeks of pregnancy.[18] Conjoined twins should be checked for further abnormalities in addition to neural tube problems, orofacial malformations, an imperforate anus, and a diaphragmatic hernia. The related congenital heart abnormalities are the primary determinants of survival.[19] Thoracopagus twins usually have a poor prognosis due to a higher prevalence of cardiac abnormalities and a more complex hepatic and biliary fusion These twins often share a heart and have composite cardiac defects, which compromises the outcome of disunion surgery.[20] 92% of conjoined twins have cardiac issues, seventy-five percent of conjoined hearts occur, and ninety percent of thoracopagus twins shared a pericardial sac. In 62% of cases, there were extracardiac abnormalities, including limb, facial, and abdominal wall deformities.[21]

In our case, the pregnant woman did not receive any antenatal visit or ultrasound scan before; hence, the diagnosis was made in the second trimester. Early diagnosis of CTs substantially aids in controlling a pregnancy and planning delivery techniques. When the parents in this case learned that the twins had a diagnosis of 23±1 week POG, they made the decision not to seek any form of care for them at birth. In our instance, neither twin survived to be given the opportunity to undergo additional imaging tests. Early diagnosis allows the parents of conjoined

twins to make informed choices. However, accurate data on the combined visceral involvement of cardiac structures might not become accessible until much later in pregnancy, which would have a substantial influence on counseling regarding remote viability. It seems that on careful transvaginal sonography and serial scanning, the anatomical parts of the fetuses are indistinguishable from one another. Magnetic resonance imaging (MRI), computed tomography (CT) scan, 2D- and 3-D ultrasonography – all are helpful to diagnostic procedure of conjoined twins as well as determining the type and severity of the associated abnormalities. Depending on the shared internal components and the attachment site, surgery to separate conjoined twins can be very easy or extremely complicated. Most circumstances involving separation are extremely risky and perhaps fatal. If the issue is detected early, as it usually is, pregnancy termination is advised because the neonates' long-term prognosis is not good. Ethical and social factors are taken into account when making this decision.[12]

Depending on the kind of union, overall survival rates can range from 20% to 25%. Conjoined twins do not increase the likelihood of a recurrence, according to available data. Conjoined twins of the cranio-thoracopagus are fortunately rare. The outlook for these twins is dire, even though they only have one brain and cannot have their brains surgically separated. The degree of dural venous sinus system sharing between partial and whole craniopagus twins can be determined. If there are no vital organs that the conjoined twins' thoraco-omphalopagus types are sharing, they might require difficult surgery to separate.[8] Conjoined twins should be delivered via cesarean section regardless of future plans for neonatal care, as this lowers the risk of intrapartum death, internal bleeding, and possible damage to the shared fetal organs.[22]

Conclusion

Conjoined twins are one well-known example of a rare event. To prevent complications and give parents the option of a safe, medically assisted abortion, it is imperative to use ultrasound technology as soon as possible to diagnose a pregnancy. It is well known that the Thaco-Omphalopagus type is prone to numerous cardiac problems. Thus, prognosis prediction and patient counseling will be supported by early twin diagnosis and knowledge regarding organ sharing from imaging modalities. MRI and ultrasound work well together to make a powerful diagnostic tool. Positive outcomes would be maximized by pediatric surgeons working in collaboration with neonatologists and obstetricians.

References

1. *Mutchinick OM, Luna-Muñoz L, Amar E, Bakker MK, Clementi M, Cocchi G, et al. 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(4):274-87.*
2. *Rode H, Fieggen AG, Brown RA, Cywes S, Davies MR, Hewitson JP, et al. Four decades of conjoined twins at Red Cross Children's Hospital--lessons learned. S Afr Med J. 2006;96(9 Pt 2):931-40.*
3. *Stone JL, Goodrich JT. The craniopagus malformation: classification and implications for surgical separation. Brain. 2006;129(5):1084-95.*
4. *O'Brien P, Nugent M, Khalil A. Prenatal diagnosis and obstetric management. InSeminars in pediatric surgery 2015;24(5):203-6.*

5. *Willobee BA, Mulder M, Perez EA, Hogan AR, Brady AC, Sola JE, et al. Predictors of in-hospital mortality in newborn conjoined twins. Surgery. 2019;166(5):854-60.*
6. *Martin JA, Hamilton BE, Sutton PD, Ventura SJ, Menacker F, Kirmeyer S, et al. Births: final data for 2005. Natl Vital Stat Rep. 2007;56(6):1-103.*
7. *Kaufman MH. The embryology of conjoined twins. Childs Nerv Syst 2004;20(8–9):508–25.*
8. *Kumari S, Singh V, Mishra HK. Thoraco-omphalopagus Conjoined Twins: A Rare Case Report. J Mahatma Gandhi Univ Med Sci Tech. 2018;3(3):102-4.*
9. *Quiroz VH, Sepulveda WH, Mercado M, Bermudez R, Fernandez R, Varela J. Prenatal ultrasound diagnosis of thoracophagus conjoined twins. J Perinat Med 1989;17:297-303.*
10. *Diaz JH, Furman EB. Perioperative management of conjoined twins. Anesthesiology 1987;67:965-73.*
11. *Edmonds LD, Layde PM. Conjoined twins in the United States, 1970-1977. Teratology 1982;25:301-8.*
12. *Bangal VB, Gavhane SP, Kharde PS, Singh RK. Caesarean Section for Thoraco-Omphalopagus Conjoined Twin – A Case Report. Int J Biomed Res. 2013;4(5):222.*
13. *Sinha A, Saxena R, Pathak M, Rodha MS. Conjoined thoracopagus twins - our experience of successful separation. J. Indian Assoc Pediatr Surg. 2021;26(5):354-7.*
14. *Kamalian N, Shirani S, Soleymanzadeh M. Thoraco-omphalo-ischiopagus tripus conjoined twins: report of a case. J Forensic Res. 2011;2(1):117.*
15. *Casta A. Cardiovascular evaluation in thoracopagus twins. Circulation. 1999;100(1):103.*
16. *Seo JW, Lee YS, Chi JG. Cross-sectional illustration on major types of conjoined twins. J Korean Med Sci. 1988;3(1):19-25.*
17. *Pajkrt E, Jauniaux E. First trimester diagnosis of conjoined twins. Prenat Diagn. 2005;25:820-6.*
18. *Chen CP. Thoraco-omphalopagus twins associated with omphalocoele and an umbilical cord cyst. Taiwan J Obstet Gynecol. 2007;46:183-4.*
19. *Villarreal JA, Yoeli D, Masand PM, Galvan NTN, Olutoye OO, Goss JA. Hepatic separation of conjoined twins: operative technique and review of threedimensional model utilization, J. Pediatr Surg. 2020;55(12):2828-35.*
20. *Seo JW, Shin SS, Chi JG. Cardiovascular system in conjoined twins: an analysis of 14 Korean cases. Teratology. 1985;32(2):151-61.*
21. *Kingston CA, McHugh K, Kumaradevan J, Kiely EM, Spitz L. Imaging in the preoperative assessment of conjoined twins. Radiographics. 2001;21(5):1187-1208.*
22. *Greco PS, Pitts D, Weadock WJ, Ladino-Torres M, Laventhal NT, Mychaliska G, et al. Conjoined twins: an obstetrician's guide to prenatal care and delivery management. J Perinatol. 2021;41(10):2424-31.*