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Case Report

Thoraco-omphalopagus conjoined twin: a rare case report

Reema Jain*, Nishi Makhija

Department of Obstetrics and Gynecology, Teerath Ram Shah Hospital, Delhi, India

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***Correspondence:**
Dr. Reema Jain,
E-mail: reemaforfb@gmail.com

ABSTRACT

Conjoined twins also known as Siamese twins, is a rarely seen congenital anomaly with an incidence as low as 1 in 50000 to 1 in 100 000 live births. Among various types of conjoined twin, commonest type is the thoracopagus type, where the fusion is anterior, at the chest, and involves the heart. There is a high rate of stillbirth and neonatal deaths resulting in very few cases surviving long enough for surgical separation. We reported one case of multi gravid woman, admitted to our hospital with an ultrasound report of conjoined thoracopagus twin at 19 weeks gestation. Surgical termination of pregnancy was done. Conclusion. Making an early diagnosis with ultrasonographic examination gives the parents a chance to elect pregnancy termination and avoid life threatening neonatal complications.

Keywords: Siamese twins, Thoraco-omphalopagus, Monozygotic twins

INTRODUCTION

Conjoined twins, popularly referred to as Siamese twins, represent one of the rarest forms of twin gestation. They occur in roughly 1 in every 200 monozygotic twin pregnancies and are always identical. The incidence ranges from 1 in 50,000 to 1 in 100,000 live births with a somewhat higher incidence in Southwest Asia and Africa.[1] Conjoined twins are monozygotic twins that do not completely separate from one another and are partially attached due to the incomplete division of one fertilised ovum resulting in monochorionic (sharing one placenta) and monoamniotic (sharing one amniotic sac) conjoined twins.[2]

Because this situation carries high risk, early diagnosis and management of delivery is extremely important. The role of ultrasound in early diagnosis and management is discussed. Due to miscarriage or termination of pregnancy, many of these pregnancies do not achieve a viable gestational age.[3] Many conjoined twins are not alive when born (stillborn) or die shortly after birth, advances in surgery and technology have improved survival rates. Some surviving conjoined twins can be surgically separated. The success of surgery depends on where the twins are joined and how many and which organs are shared. It also depends on the experience and skill of the surgical team. Making an early diagnosis with ultrasonographic examination gives the parents a chance to elect pregnancy termination and avoid life threatening neonatal complications.

CASE REPORT

A 28-year-old, G2P1, with a previous normal delivery, came to the hospital for her first antenatal visit at 6 weeks, during which an obstetrical ultrasound and other routine investigations were advised to her. She reported back after a few days with an ultrasound report showing twins, both live fetus, 6.5 weeks pregnancy with a huge dermoid cyst (which she knew before also) and all other normal routine investigations. All required ante-natal counselling was done to her. She then went to her village and reported back to the hospital at 19 weeks. On examination, her uterine height was more than expected., both the foetal heart sounds audible, an anomaly scan was advised to her. To the astonishment, her anomaly scan showed twin live pregnancy with thoracopagus conjoined twin. The twins were joined at the thorax and upper abdomen. The scan also showed a huge right sided dermoid cyst. The

unfortunate news was shared with the family, the parents were informed of the malformation and the likely outcome if the twins survived after delivery. The parents decided to terminate the pregnancy and refused further evaluation and investigations pertaining to the twin. The written informed consent was taken. Surgical termination was done along with removal of the dermoid cyst.

Figure 1: Thoracopagus conjoined twin.

DISCUSSION

Conjoined twins are classified according to the most prominent site of conjunction: thorax (thoracopagus), abdomen (omphalopagus), sacrum (pygopagus), pelvis (ischiopagus), skull (cephalopagus), and back (rachipagus). Depending on the aspect of the embryonic disc, the most common types are thoracopagus (19%).[4] Its etiology is unknown, but an incomplete division of the zygote between 13th and 15th days after fertilization is probably responsible.[5] The overall survival rate for conjoined twins is approximately 25%.[6] The condition is more frequently found among female foetus, with a ratio of 3:1.

Conjoined twins are always of the same sex as they are monozygotic, monochorionic, and monoamniotic at all times. Some risk factors like positive history of twin delivery, use of drugs for induction of ovulation, infertility treatment, and exposure to detrimental radiation were proposed to have a likely effect on the development of this rare condition.[7] However, none of these risk factors were covered by the past, family, and pregnancy history of our case.

Any of the following classical signs identified by USG may suggest the diagnosis: inability to separate foetal bodies after careful observation, both foetal heads in the same plane, unusual cervical spine flexion backwards, no change in the relative position following manual manipulations and movements by the mother.

Thoracopagus twins usually have an impoverished prognosis because of a greater incidence of cardiac anomalies and the more complex hepatic and biliary fusion.[8] These twins usually share hearts and have composite cardiac anomalies as well, which imperil the successfulness of disunion surgery.[9] The extent of the union of the hearts in thoracopagus twins varies but can include fusion of the large vessels, the atria, the atria and ventricles, and rarely, a single heart in one of the twins

Early diagnosis of conjoined twins was previously reported, but not before the 10th week of gestation.[10] This could be the reason of missing of conjoined twin pregnancy in our case.

Once conjoined twins have been diagnosed, characterization of the type and severity of the abnormality can be performed with ultrasound, three-dimensional ultrasound, computed tomography, or magnetic resonance imaging.

Choice for management of pregnancy includes, termination of pregnancy or the expectant management, should be rigorously discussed. Individualisation of each and every case is required as surgical separation of conjoined twins has been reported in the literature but with limited success only.

In the present study, the diagnosis has been performed in the second trimester, and because the family has chosen termination of this pregnancy, further diagnostic intervention was not considered.

CONCLUSION

In conclusion, conjoined twins are associated with a high perinatal mortality and morbidity; therefore, making an early diagnosis with ultrasonographic examination of conjoined twins gives the parents a chance to elect pregnancy termination. The obstetrician's role in timely prenatal diagnosis, counselling, and organization of interdisciplinary medical care is indispensable in cases of conjoined twin.

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REFERENCES

1. Rees EJ, Vujanic GM, W. M. Williams WM. Epidemic of conjoined twins in Cardi. Br J Obstetr Gynaecol. 1993;100(4):388-91.
2. Saxena R, Sinha A, Pathak M, Rathod KJ. Conjoined thoracopagus twins: a systematic review of the anomalies and outcome of surgical separation. Afr J Paediatr Surg. 2023;20(3):157-65.
3. Stone JL, Goodrich JT. The craniopagus malformation: classification and implications for surgical separation. Brain. 2006;129(5):1084-95.
4. Schnaufer L. Conjoined twins. Swenson's Pediatric Surgery, In: Raffensperger JG, eds. New York, NY, USA: Appleton Century-Crofts; 1980: 910-20.
5. Abossolo T, Dancoisne P, Tuaillon J, Orvain E, Sommer JC, Riviere JP. Early prenatal diagnosis of asymmetric cephalothoracopagus twins. J de Gyn Ecologie Obst Etrique Et Biologie De La Reproduct. 1994;23(1):79-84.
6. Stone JL, Goodrich JT. The craniopagus malformation: classification and implications for surgical separation. Brain. 2006;129(5):1084-95.
7. Kamalian N, Shirani S, Soleymanzadeh M. Thoraco-omphalo-ischiopagus tripus conjoined twins: report of a case. J Forensics Res. 2011;2(1):1000117.
8. Villarreal JA, Yoeli D, Masand PM, Galvan NTN, Olutoye OO, Goss JA. Hepatic separation of conjoined twins: operative technique and review of three-dimensional model utilization. J Pediatr Surg. 2020;55(12):2828-35.
9. Seo JW, Shin SS, Chi JG. Cardiovascular system in conjoined twins: an analysis of 14 Korean cases. Teratology. 1985;32(2):151-61.
10. Hubinont C, Kollmann P, Malvaux V, Donnez J, Bernard P. First-trimester diagnosis of conjoined twins. Fetal Diagn Ther. 1997;12(3):185-7.

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