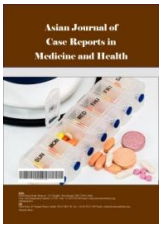

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Thoracoomphalopagus-the Twins Sharing Heart

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Authors' contributions

This work was carried out in collaboration among all authors. Author MS managed the case primarily, designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Author SC assisted in literature reviewing and final drafting of manuscript. Author PS managed the analyses of the study. Author Rajkiran managed the literature search and was involved in surgical management of the case. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Conjoined twin is one the rarest and most fascinating phenomena of twinning. The incidence ranges from 1 in 50 000 to 1 in 100 000 live births. Even though the degree and location of conjunction and the shared internal organs determine the prognosis of conjoined twins, they are associated with a high perinatal mortality rate. Early prenatal diagnosis and precise characterization of conjoined twins are essential for optimal obstetric, interventional and postnatal management as well as to reduce psychological trauma to the parents. Imaging by ultrasound and MRI plays a significant role in diagnosis and further management prediction. In this report, we present a case of multigravida pregnant woman who presented at 36+2 weeks gestation with thoraco-omphalopagus twins in advanced labour in a tertiary hospital. This case report not only reports a rare case but also emphasizes on the fact that even after so many advancements in the field of medicine, there still exists a gap in timely access to the healthcare facility, especially in developing countries.

Keywords: Conjoined twins; MRI; thoracoomphalopagus; pregnant woman.

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1. INTRODUCTION

Conjoined twin is one of the rarest and most fascinating subset of monochorionic monoamniotic twins. Conjoined twins result due to incomplete embryonic division on 13th to 15th day following conception, resulting in varying degrees of fusion between the two fetuses. Conjoined twinning occurs in about 1% of monochorionic twins with an estimated incidence ranging from 1:30,000 to 1:2,00,000 live births and 1 in 650 to 900 twin deliveries. An increased incidence of 1:14,000 to 1:25,000 is stated in various parts of South East Asia and Africa. Around 40% to 60% of conjoined twins are reported to be still born and approximately 35% of live born ones do not survive beyond first 24 hours. Prognosis depends on the gravity of the vital organ shared, the accompanying congenital anomaly and location and extent of fusion between the twins. There is female predominance with the ratio of male to female being 1:3, particularly in thoracopagus [1]. Spencer and colleagues classified conjoined twins according to the most prominent site of fusion into three major groups:

1. Twins fused ventrally -cephalopagus (head), thoracopagus (Thorax), omphalopagus (Abdomen) and Ischiopagus (pelvis);
2. Twins fused dorsally -pygopagus (sacrum), rachipagus (spine, back and craniopagus (cranium);
3. Twins fused laterally - parapagus.

With an incidence of 74-75%, thoracopagus is the most common variety. Omphalopagus is the least common variety with about 0.5% quoted incidence.

2. CASE REPORT

A 24 year old G4P3L2 woman presented to the gynae casualty of a tertiary hospital of central Delhi in advanced labour. The heteroanamnesis data collected from the patient and the closest relatives revealed that the pregnancy was confirmed by urine pregnancy test done at home after an amenorrhea of 2 months. However, the pregnancy was not registered at any health facility. First and second trimester pregnancy periods were uneventful. Fetal movements were perceived normally from the fifth month. First antenatal ultrasonography was done at 6 months gestation by referral of a local practitioner and was documented to have a single live intrauterine fetus of 24+2 weeks. Conjoined twin was diagnosed on a third trimester ultrasound done by a radiologist at 33+3 weeks and fetal MRI was advised for the same. MRI revealed thoraco-omphalopagus type of conjoined twin with single fused heart at the midline of thorax. However, due to excessive motion artefacts, exact morphology of fetal heart and liver could not be commented (Fig. 1).

There was no history of twinning in family. All three previous deliveries were home vaginal delivery at term conducted by dai. She came to the casualty in advanced labour. On examination, vitals were stable with normal general physical examination. On per abdomen examination, uterine contractions were moderate with head in the lower pole palpable 3/5th above the brim. Second head was palpated in the maternal right iliac fossa. Single fetal heart sound was localized in midline below umbilicus. A per vaginum examination revealed 8 cm dilated and 80% effaced cervix; the presenting part was high up. Pelvis was adequate. A decision for an emergency Lower segment caesarean section (LSCS) was taken after explaining the situation and obtaining consent. The same was discussed with senior neonatologist and the pediatric surgeons. Lower segment caesarean section was performed by the senior surgeon. Peroperatively, liquor was in excess amount and first twin was extracted as cephalic followed by second twin as breech. The twins cried immediately after birth. The twins were girl babies fused ventrally from sternal angle to below the level of umbilicus with single umbilical cord (Fig. 2).

Twins were handed over to the neonatologist. The combined weight was 4000 g. Placenta was extracted completely with membranes intact. It was a monochorionic placenta weighing 550 g (Fig. 3).

Uterus and abdomen were closed in layers after ensuring hemostasis.

Both the babies were hemodynamically stable and planned for follow up MRI after 3 months with pediatric surgeons after explaining all possibility of neonatal death. The mother was discharged on day 7 with satisfactory postoperative period.

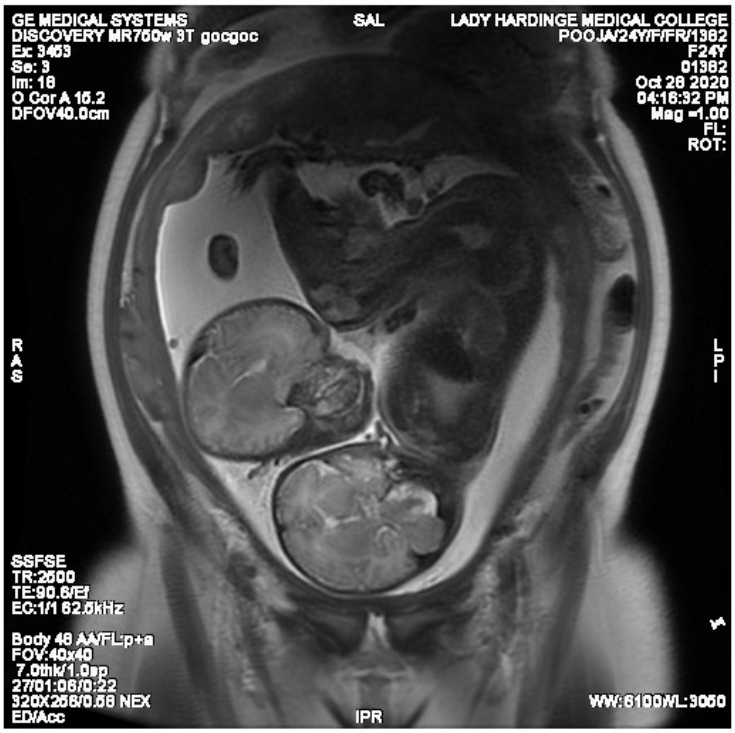

Fig. 1. MRI showing thoracoomphalopagus twins-the twins are joined ventrally with sharing of the heart

Fig. 2. The live twins attached ventrally from sternum to the level of umbilicus

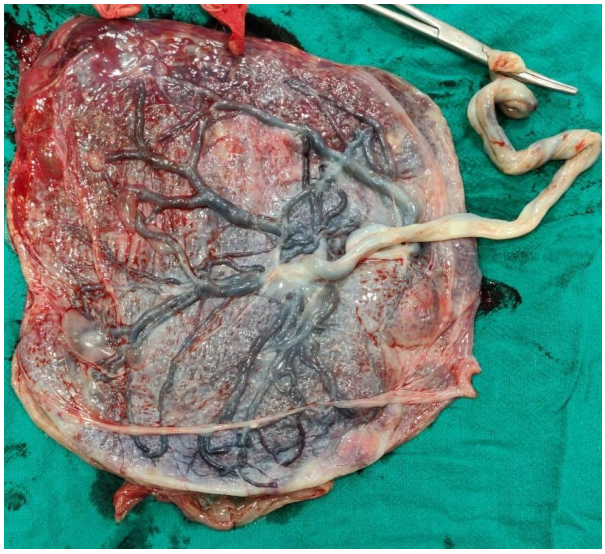

Fig. 3. Single placenta of monochorionic twins (thoracoomphalopagus)

3. DISCUSSION

Two contraindicating theories exist to explain the origins of conjoined twins. Theory of fission states that the fertilized egg splits partially and results in conjoined twins due to delayed separation of the embryonic mass after 12th day of fertilization. Another theory is one of fusion, in which a fertilized egg completely separates, but later fuses resulting in conjoint twinning [2].

The frequency of conjoined twins is independent of the maternal age and parity and is mostly sporadic. Polyhydramnios is frequently present and the same was seen in our case. There is a significant female predominance in thoracopagus conjoints. In accordance, our reported case had the same sex predominance [3].

Antenatal diagnosis of conjoined twins is usually uncommon and the fact that they are conjoined is usually not determined until late in gestation or during parturition. The diagnostic modalities available for antenatal diagnosis of conjoined twins includes ultrasonography and magnetic resonance imaging [4]. Gray and associates established a list of criterion for antepartum diagnosis of conjoined twins: the heads being at the same level and body plane; unusual proximity of the spines; spines are usually extended; no change in fetal position relative to each other after movement or manipulation [5].

A detailed survey of the vasculature of fused vital organs is very important in determining the prognosis of surgical separation and also owing to the high frequency of associated anomalies related to fusion, which includes neural tube defects, orofacial clefts and diaphragmatic hernia. Although, our case had no grossly evident fetal congenital anomaly, Mishra et al. [6] reports fetal craniosynostosis, limb aplasia and omphalocele in their case study of omphalopagus twins. Presence of congenital anomalies in monochorionic twins on antenatal ultrasound may raise alarm to suspect conjoined twins but situation may be difficult if no fetal anomalies are detected otherwise. There are other case studies who have reported detection of conjoined twins as early as 16 weeks [7].

The vital organs shared by the babies would strongly prognosticate the outcome in case surgical separation is possible postnatally. Case study by Omran et al. [8] reports successful neonatal outcome after surgical separation as the omphalopagus twins shared gastrointestinal organs and abdominal wall. Recently Bahodur et al. [9] reported two cases of successful surgical separation of thoraco-omphalopagus conjoined twins after tremendous efforts and surgical intricacies but these babies luckily had separate hearts making the surgical outcome a success. On the contrary, in our case babies shared a single heart posing a great surgical challenge and worst prognosis.

The possibility of difficulties during delivery should be taken into account, such difficulties can be due to the size of the conjoined twins and

their position in relation to each other, such pregnancies are usually delivered by caesarean section and have a risk of extension of uterine incision and traumatic postpartum hemorrhage.

Early prenatal diagnosis and precise characterization of conjoined twins are essential for timely and optimal obstetric intervention and postnatal management as well as to reduce psychological trauma to the parents.

This case report not only reports a rare case but also emphasizes on the fact that even after so many advancements in the field of medicine, there still exists a gap in timely access to the healthcare facility especially in developing countries. Early diagnosis of such cases may help to offer timely termination of pregnancy. When severe forms of conjoined twinning are diagnosed prior to 20 weeks, termination via vaginal delivery should be considered. Caesarian section is indicated when surgical separation and viability is possible to minimize complications to the fetus and the mother. In advanced gestation caesarian may be only resort to deliver these twins. This thereby emphasizes on the the need for early and accurate diagnosis to outline a particular management plan for the mother and the baby in order to mitigate potential complications.

4. CONCLUSION

The case reported by us adds to the available literature on this rare condition. Moreover, this highlights the inadequacy of health facilities in developing countries,where a casually done ultrasound missed an important condition which if diagnosed timely would have helped the patient to opt timely for medical termination of pregnancy .Ultrasound is the modality of choice for prenatal detection of conjoined twins and MRI offers for better characterization. Vaginal delivery is preferred for ealy gestations and for fetuses with poor prognosis based on MRI. Caesarian section is indicated in advanced gestations where vaginal delivery may not be possible due to non descent of presenting parts or when there is a possibility of nearly successful surgical separation of viable babies. The condition is associated with social and legal issues with late termination, influenced by cultural and religious beliefs. The low incidence and poor outcomes of this condition calls for a vigilant examination on part of the radiologist as well as the obstetrician to avoid problems during the advanced gestation of pregnancy.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Alkhateeb M, Mashaqbeh M, Magableh S, Hadad R, Nseer Q, Alshboul A. Early prenatal diagnosis of thoracopagus twins by ultrasound. Acta Inform Med. 2015; 23(1):60-2.
DOI:10.5455/aim.
2. Spencer R. Theoretical and analytical embryology of conjoined twins: part I: Embryogenesis. Clin Anat. 2000;13:36-53.
3. Mutchinick OM, Luna-Muñoz L, Amar E, 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): 274-287.
DOI:10.1002/ajmg.c.30321
4. Teixeira Castro P, Werner H, Matos AP, Daltro P, Araujo Júnior E. Symmetric and ventrally conjoined twins: Prenatal evaluation by ultrasound and magnetic resonance imaging and postnatal outcomes. The Journal of Maternal-fetal & Neonatal Medicine: The Official Journal of the European Association of Perinatal Medicine, The Federation of Asia and Oceania Perinatal Societies, The International Society of Perinatal Obstetricians. 2019:1-8.
DOI: 10.1080/14767058.2019.1651282
5. Apuzzio JJ, Ganesh V, Landau J, Pelosi M. Prenatal Diagnosis of Conjoined Twins. Am J Obstet Gynecol. 1984;148:343-4.
6. Mishra N,Rohilla M.Thoraco-omphalopagus Conjoint Twin:A case Report and Literature Review. Gynaecology and Obstetrics Case Report. 2015;1(1):3:1-3.

7. Sultan OM, Tawfeek AS. Conjoined twins—thoraco-omphalopagus (type A). BJR Case Rep. 2016;2:20150016.
8. Omran A, Nassif M, Salhab N, Abdo A, et al.Early separation of omphalopagus conjoined twins: a case report from Syria, Journal of Surgical Case Reports. 2020;1:rjz374.
Available:https://doi.org/10.1093/jscr/rjz374
9. Bahador A, Nikeghbalain S, Foroughi M, Beizavi Z, Sahmodini MA, Eghbal MH. Successful Surgical Separation of Thoraco-Omphalopagus Symmetrical Conjoined Twins in Iran: Two Case Reports. Iran J Med Sci. 2020;45(2):143-147.
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