Case Report

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Conjoint Twin-Thoraco-Omphalophagus: A Case Report and Literature Review

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Abstract

Conjoined twins is a rare entity experienced by any obstetrician. The management of the same requires early diagnostic work up antenatally and involvement of parental decision making. A 27-year-old woman of term gestation presented with conjoined twins-thoraco-omphalophagus and was taken up for elective LSCS and delivered. This article reports presentation of conjoined twins and the literature is reviewed.

Keywords: Conjoined twins; thoraco-omphalophagus.

Introduction

United or conjoined twins have been referred to as Siamese twins after Chang and Eng Bunker of Siam (Thailand), who were displayed worldwide by P.T Barnum in circus born in 1811.[1] The brothers lived for 63 years without considering separation and they married sisters and fathered 21 children.[9] Description of conjoined twins date back to ancient Egyptians but the first well documented case was born in 1100 in Kent, England. Conjoint twins are a rare phenomenon of a monochorionic monoamniotic twin. The incidence varies from 1 in 50000 to 1 in 100000 live births.[4] Most of them (2/3rd) are female foetuses. This is because mortality rates are lower in females and female zygotes have a greater tendency to divide.[1] Most of them result in miscarriages and stillbirths (60%); only 18% of all conjoined infants survive longer than 24 hours. Preterm delivery is the most common outcome. Around 35% of live births die within first 24 hours. Spencer et al. divided the twins into three major groups: twins with ventral union, a dorsal union and a lateral union. The first major group includes four types: craniophagus (head), thoracophagus (chest), omphalophagus (umbilicus) and ischiophagus (hip). The dorsal union includes three types: pyophagus (sacrum), rachiphagus (spine) and craniophagus (cranium).[2] The last major group includes just paraphagus(side) twins. Thoracophagus is the most common type (75%) and omphalophagus the least (0.5%). Incidence of thoraco-omphalophagus is 28%. Pyophagus twins have better prognosis than the omphalophagus and the thoracophagus.[2]

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A 27 years old woman, who is a gravida 5 para 4 living 3, from a poor socioeconomic background had a spontaneous conception and she wasn't aware of her pregnancy until 29 weeks of gestation when an ultrasound was done which revealed a conjoint twin (thoraco-omphalophagus). (Figure 1.1)

Fig. 1: Conj-twins.

Fig. 2: Fused abdomen.

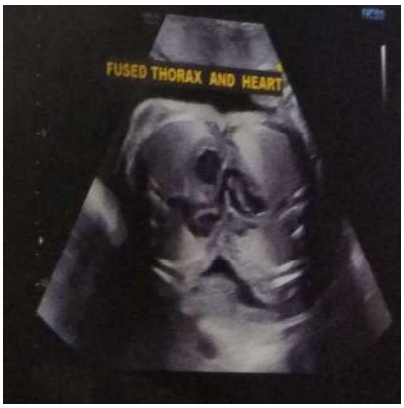

Fig. 3: Fused thorax.

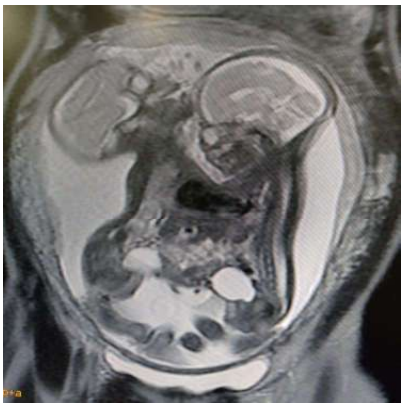

Fig. 4: Ultra-sonography.

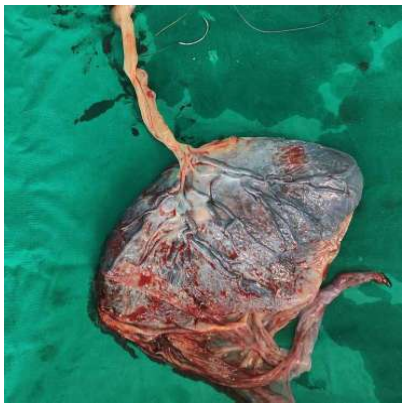

Fig. 5: MCMA placenta.

The MRI image showed the twins sharing a single heart, liver and part of the bowel. (Figure 1.2).

She came to Cheluvamba hospital, Mysore medical college and research institute at 38 weeks of gestation not in labour. On per abdomen examination, an unduly enlarged uterus with multiple fetal parts palpable and USG revealed a MCMA conjoint twin with a single placenta and umbilical cord, the thorax and upper abdomen of the foetuses are fused. She was planned for an elective caesarean section. Lower segment caesarean section was performed by the obstetrician and twins extracted by vertex of first twin followed by its breech and breech of the second twin followed by its vertex. Both twins were alive females, cried at the time of birth sharing the same thoracic cavity and abdomen with two heads and eight limbs. (Figure 2) Their combined weight was 4.5 kgs and APGAR was 6 and 8 at 1min and 5 min for twin 1 and 7 and 9 at 1 min and 5 min for twin 2. The twins were shifted to NICU. Supportive care was given and was referred to higher centre for further management.

Discussion

Conjoint twins are believed to be the outcomes of a faulty division of an embryo at 13-15 days of conception. Two contradicting theories exist to explain the conjoint twins -the fission and the fusion theory. Fission theory talks about the partial splitting of the fertilized egg at day 13 of gestation producing identical twins with shared anatomical structures. The fusion theory-more acceptable theory in which the fertilized egg completely separates but the stem cells which search for similar cells interact sometime between 13 and 15 days of gestation at sites where the ectoderm is not present or where it is disrupted, like in thoraco-omphalophagus at the site of septum transversum and heart. Oropharyngeal and cloacal membranes:

Cephalophagus, ischiophagus, paraphagus.[1] Unlike identical MCMA twins conjoined twins have remarkably different personalities. There is no association of conjoined twins with race, parity, maternal age or heredity. It is sporadic and recurrence risk is negligible. Ultra-sonography is the most accurate pre-natal diagnostic technique for this condition. In the early weeks of pregnancy, the monochorionic twins may be mistaken for conjoined twins. A repeat ultrasonographic examination should ideally be done at between the eleventh and twelfth weeks of gestation, to confirm the diagnosis and to avoid discovering a conjoint twin later as in the above case.[3] Many cases unsupervised without any antenatal check-up could end up in deadly complications such as rupture uterus requiring laparotomy, destructive surgeries for delivery of the fetuses or even hysterectomy.[6] Postnatally chest and abdomen X-ray, ultrasound: cranium, spinal cord, abdomen, echocardiography, CT and also contrast studies of the gastrointestinal and genitourinary tract are the investigations. The prognosis of conjoined twins is poor. The total survival rate is 7.5%. Management of conjoined twins has been described under three headings. Non-operative management in cases of complex cardiac union without chance of a reconstruction surgery, extensive cerebral fusion, parental refusal of surgery, almost 100% mortality. The second one is Emergency separation in case of dead or dying twin, threatening the survival of its sibling or with a correctable congenital anomaly. The chances of survival are 25%. The other option is that of a planned separation is when the twins are stable and thriving well and the surgery can be planned at 2-3 months of life.[5] Surgical separation of nearly complete conjoined twins may be successful when vital organs are not shared. The outcomes of surgical separation appear to be good in selected cases. If surgical separation is possible with expected survival of both twins, then it is strongly recommended. Identification of the anatomical structures is accomplished with postnatal imaging, preferably MRI. The surgical survival rate is around 66.7%. Improvement in survival rate for surgery of conjoined twins is due to advances in diagnostic techniques, especially computed tomography and magnetic resonance imaging, meticulous anaesthetic management with careful monitoring of fluid replacement intraoperatively, improved surgical techniques with special emphasis on restricting blood loss, such as the use of ultrasonic separation of fused livers, achieving body-wall closure, and, most importantly, the value of previous experience, and postoperative intensive care with accurate attention to potentially labile cardiovascular status.[7] In January 2021, 14 days old conjoined twins sharing the chest wall, liver and part of the intestines were planned for separation surgery. After a 6-hour long surgery the twins were separated successfully and were discharged after 5 weeks at Wadia hospital, Mumbai.[8] Surgical separation of conjoined twins is extremely challenging taking in the consideration the high perioperative risk. Also, surgical management would necessitate prolonged neonatal ICU requirement which may not be possible for many of the patients having a poor economic background.

Conclusion

Our case emphasizes on the need for a prenatal ultrasound diagnosis in early gestation. Moreover, this highlights the inadequacy of health facilities and the general awareness among the public. Emergency surgery may be required, but it is preferable to delay surgery to allow growth and the completion of investigations. Inevitably, the ultimate prognosis will depend on the state of the conjoined organs and the potential for successful separation. Tragically, in some cases separation will not be possible. Despite successful separation, some children are left crippled and disabled, requiring lifelong follow-up and care. Parental decision making plays its vital role in deciding the fate of the twins as it poses a burden physically, mentally and socioeconomically.

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