Conjoined Twins- Cephalo-Thoraco-Omphalopagus: A Case Report

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ABSTRACT

A thirty-one-years old woman with spontaneous conjoined twin pregnancy, Gravida 3 Para 2 with history of previous 2 cesarean sections was referred to the obstetrics and gynecology clinic in King Hamad University Hospital at 18 weeks of gestation for further management. She was diagnosed as cephalo-thoraco-omphalopagus female conjoined twins in private clinic, which was confirmed in our institute. Thus, surgical termination by hysterotomy was performed.

INTRODUCTION

Conjoined twins (CT) also known as Siamese twins are defined as monochorionic monoamniotic twins that are anatomically fused together in utero[1]. It is an extraordinary phenomenon of uncertain etiology that occurs due to an abnormality during embryological development. The incidence rate varies between 1 in 50,000 to 1 in 100,000 births[2] and it has a 1:3 male to female ratio with female fetuses being most commonly affected[3]. Several types of conjoined twins were described in the literature with thoracopagus being the most common type and omphalopagus the least common[4].

THE CASE

A thirty-one-years old Egyptian woman with spontaneous twin pregnancy, Gravida 3 Para 2 with history of previous 2 cesarean sections was referred to the obstetrics and gynecology clinic in King Hamad University Hospital as a tertiary center at 18 weeks of gestation seeking for a second opinion regarding an abnormal antenatal scan report from a private clinic. The patient did not have any family history of twins and no history of consanguinity. No medical history or allergies were reported.

The ultrasound that was performed at the private clinic at 17+weeks gestation revealed a diagnosis of conjoined viable female twins (cephalo-thoraco-omphalopagus) with 2 faces fused laterally and four eyes with two central globes lying adjacent to each other. Two brains were fused along the temporal region, which appeared grossly normal.

A single fused chest was noted along with fused abdomens. A single heart lying along the left side of the thoracic region and a possible atrioventricular canal defect were noted.

The lung volumes appeared reduced with a single stomach seen at the level of the heart. A single liver was seen, and the kidneys were not clearly visualized however 2 urinary bladders were present. Extremities appeared grossly intact with four arms and legs. Amniotic fluid seemed normal, and the placenta was located in the upper posterior uterine segment. The cervix appeared long and closed on transabdominal scan.

An ultrasound scan performed in our hospital confirmed a diagnosis of conjoined viable female twins (cephalo-thoraco-omphalopagus) with 2 faces fused laterally and four eyes with two central globes lying adjacent to each other. Two brains were fused along the temporal region, which appeared grossly normal (Figure 1).

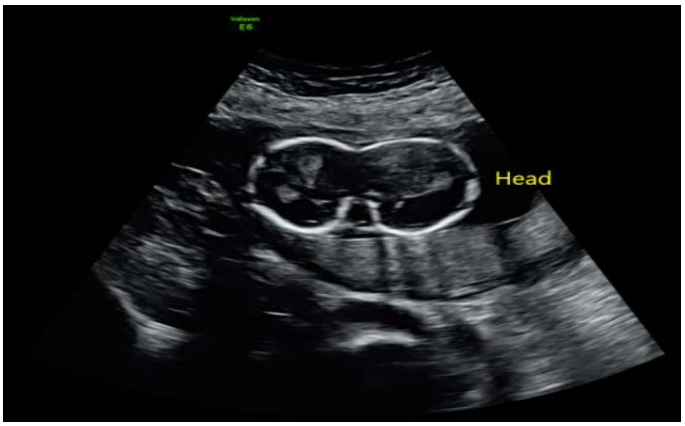

Figure 1: Trans-abdominal scan showing conjoined twin with two brains fused along the temporal region

Only one stomach and heart were seen. Ventricular septal defect was noted however the septum primum was not visualized, as it would be in a complete atrio-ventriculum canal defect. (Figure 2) Spine of fetus B appears scoliotic (Figure 3); Spine of fetus A appears grossly normal.

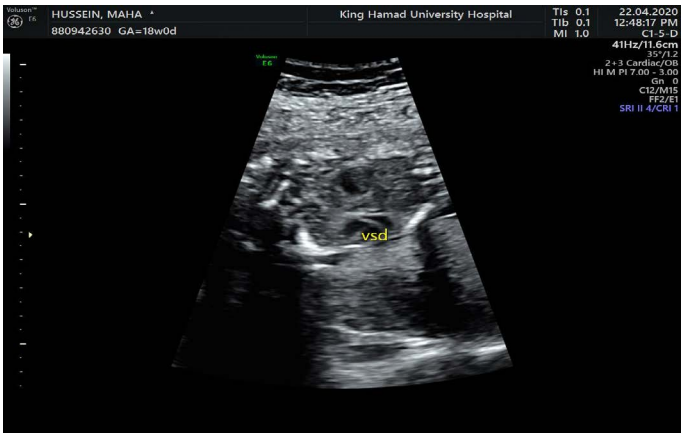

Figure 2: Trans-abdominal scan showing ventricular septal defect

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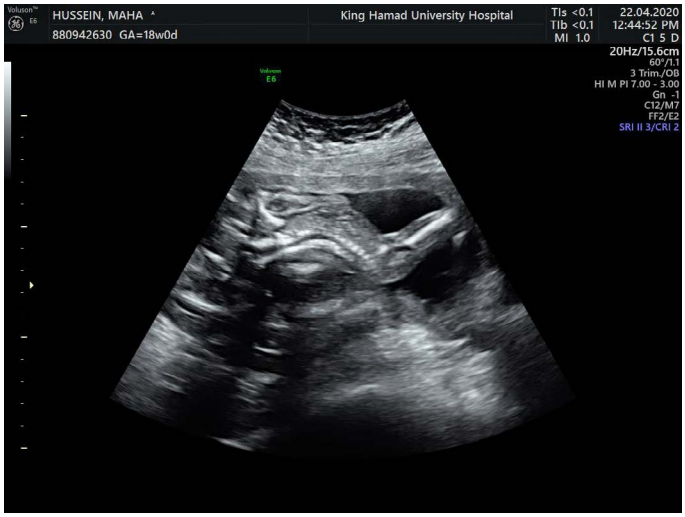

Figure 3: Spine of fetus B appears scoliotic

Both the patient and her husband were counseled in detail about the results of the ultrasound report and were advised for surgical termination, as both fetuses are incompatible with life. The couple agreed so consent was taken from the patient and signed by 3 consultants and the pediatrics team was informed about the case.

Following the discussion, the patient was booked for admission and hyesterotomy was performed at 18 weeks of gestation without any complications to the mother. The twins exhibited no signs of life at the time of delivery. Gross examination of the conjoined twins confirmed that both heads were fused laterally along with thorax and upper abdomen while the four upper limbs and 4 lower limbs appeared normal. Both twins were females weighting a total of 345 grams (Figure 4). Post-operative recovery was uneventful, and the mother was discharged in stable condition.

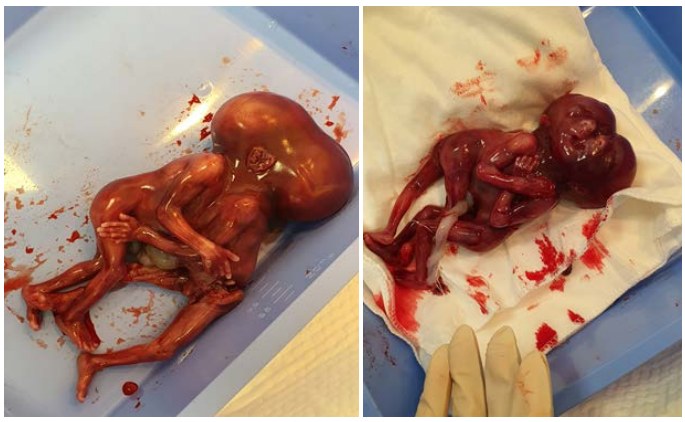

Figure 4: Conjoined twins post hysterotomy

DISCUSSION

Conjoint twins are thought to be the result of a faulty division of an embryo at 13-15 days of conception[5]. There are various theories that were proposed to explain this phenomenon. The most common theories behind the basis of development of conjoined twins are the "fission theory "and "fusion theory"[5]. The former suggests that an incomplete split of a single fertilized egg occurs, and the forming 2 embryos remain fused at the unseparated parts. Whereas the fusion theory suggests that there is complete separation of the fertilized egg but as they are close in proximity, an interaction between both twin cells may occur resulting in cell signaling and finally conjoined anomalies[6]. Most of the authors accepted the fusion theory as it can explain all types of conjoined twins[7].

The cause of conjoined twins is still not fully understood. There are some risk factors that were thought to have a possible effect on the development of this phenomenon such as a positive genetic history of twins, delivery abnormality, ovulation inducing medications, fertility treatment and radiation exposure. In our case we could not identify any obvious risk factors that may have caused this abnormality[8].

The prognosis of this condition is poor. Fourteen cases were included in a study of prenatally diagnosed conjoined twins. The study found that 28% of the cases died antenatally and 54% died immediately following delivery and18% survived; of which 50% died postoperatively [3,9,10].

There are different classification systems of conjoined twins. (Spencer et al 1996) 4 classified them into 8 main types based on the degree and site of fusion (Figure 5). Thoracophagus, omphalophagus and thoraco-omphalophagus are the most common types in this classification system accounting for about 56% of conjoined twins. The rarest type according to Spencer is Cephalophagus accounting for 11% of all cases. Fusion of the head, thorax and upper abdominal cavities are characteristics of Cephalophagus twins. Cephalopagus twins are further divided into two types: Janiceps (two faces are on either side of the head) or non-Janiceps (with a relatively normal head and face)[11]. To the best of our knowledge, the conjoined twins reported in our case are one of the rarest types of cephalophagus in which both heads are fused laterally (Janiceps type).

Embryonic aspect	Type	Incidence	Primordium	Extent of union	Separability
Ventral (87%)	—	—	—	—	—
Rostral (48%)	Cephalopagus	11%	Oropharyngeal membrane	Top of head to umbilicus	None
	Thoracopagus	19%	Heart	Thorax, upper abdomen, conjoined heart	Rare
	Omphalopagus	18%	Diaphragm	Thorax, upper abdomen, separate hearts	Likely 82% success
Caudal (11%)	Ischiopagus	11%	Cloacal membrane	Lower abdomen, genitourinary tract	Likely 63% success
Lateral	Parapagus	28%	Cloacal membrane (2 notochords?)	Pelvis, variable trunk, diprosopus 2 faces, dicephalus 2 heads	Rare
Dorsal (13%)	Craniopagus	5%	Cranial neuropore	Cranial vault	Unlikely without sequelae
	Rachipagus	2%	Neural tube (mid-portion)	Vertebral column	None reported
	Pygopagus	6%	Caudal neuropore	Sacrum	Likely 68% success

Figure 5: Classification of conjoined twins

Early diagnosis of conjoined twins with trans-abdominal or trans-vaginal ultrasonography has a vital role in the management so early termination of pregnancy can be done however usually it cannot be detected before 10 weeks of gestation[12].

Once conjoined twins are confirmed three-dimensional ultrasound, computed tomography, or magnetic resonance imaging are used to identify the type and the severity of the conjoined twins regarding anatomical anomalies[13]. This emphasized the important role of the radiologist and obstetricians in early detection of conjoined twins to avoid any problems in the later half of pregnancy and to determine the mode of delivery[3].

Since most conjoined twins are diagnosed antenatally, they are delivered electively by cesarean section, as it is the safest option for both mother and babies and to avoid complications such as stillbirth, uterine rupture, labour dystocia, shoulder dystocia, retained second twin and hysterectomy[14-16].

In cases where the gestational period is between 18-20 weeks, the pregnancy can be terminated medically and delivered vaginally using labor-inducting medication[17]. As there is an increased risk of uterine rupture and shoulder dystocia with vaginal delivery, especially with a history of multiple previous cesarean sections like in our case, we opted for surgical intervention after full counseling of the patient about the risks and benefits of each mode of delivery. The patient was booked for an elective hysterotomy at 18 weeks of gestation without any complications to the mother and was discharged in good condition.

CONCLUSION

To conclude, antenatal care, early ultrasound scan and multi-disciplinary team management plays a vital role in the early detection of conjoined twins in order to avoid any complications that result from undiagnosed conjoined twins at a later gestation and to involve multi- disciplinary team earlier on in the management plan to give optimal care to both the patient and babies.

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