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CASE REPORT

Conjoined Twins with Omphalocele and Unilateral Club Foot: A Case Report

SHARMA P *, GHIMIRE A **

ABSTRACT

Conjoined twins are a rare clinical entity. Various congenital defects like anencephaly, gastrointestinal anomaly, urogenital anomaly, cleft lip, cleft palate and lumbosacral meningomyelocele may be associated with conjoined twins. Here, we report a case of thoracoomphalopagus associated with omphalocele and club foot.

Key Message: Conjoined twins may be associated with congenital anomalies. Detailed ultrasonographical evaluation should be done for the proper evaluation of this anomaly.

Key Words: Conjoined twins, Thoracoomphalopagus, omphalocele, club foot.

* (MD), Lecturer, ** (MBBS), JR, Department of Radiology and Imaging, Nepalgunj Medical College Teaching Hospital, Banke, (Nepal) **Corresponding Author:** prakashshrm@yahoo.com

Introduction

Conjoined twins, more commonly known as Siamese twins, is a rare entity in an obstetrical Such cases occur in approximately practice. 1% of monozygotic twins or between 1 per 100000 and 1 per 250000 live births [1]. It results due to the incomplete division of the embryonic disc [2]. The most common form of conjoined twins is caused by the fusion of the anterior thorax and/ or abdomen, which is also called thoracopagus or omphalopagus and thoracoomphalopagus, constituting 70% of all conjoined twins [3]. Typical thoracopagus are conjoined from the upper thorax to the umbilicus, each twin with a normal foregut, but with a single complex multiventricular heart [4]. Thoracoomphalopagus associated with omphalocele and club foot is among the rarest in the conjoined twins. To the best of our knowledge, till date, no such case has been reported. USG is very good imaging modality in

the diagnosis of conjoined twins. In cases of complex anomaly, where USG can't define the anomaly sufficiently, MRI can be used as an adjunct.

Case Report

A 24 year old lady with second gravida, presented at nine months of pregnancy, with the complaint of pain in the lower abdomen. She had no history of multiple pregnancies or a child with any congenital abnormality. Her previous delivery was a term, normal vaginal delivery with no complication. After her examination in the obstetrical department, she was sent for an obstetric scan to assess the foetal well being and to rule out multiple pregnancies. Our ultrasound scan detected live conjoined twins which were joined from the thorax up to the level of abdomen. The chest and the liver were fused; however, the hearts were separate [Table/Fig 1]. The conjoined twins were also associated with ompahlocele. Unilatearl Club foot was also detected on one of the foetus. The gestational age by ultrasound was around 36 weeks. There was oligohydramnios with amniotic fluid volume of 4.5 cm. Termination of pregnancy was done by lower segment caesarean section after proper counseling, as the cardiotocography revealed foetal distress. Examination of the twins confirmed the prenatal ultrasound findings [Table/Fig 2]. Both were females. Both were alive during birth and died after two minutes of delivery.



(Table/Fig 1) Ultrasonography showing axial section at the level of abdomen showing fused liver.



(Table/Fig 2) Thoracoomphalopagus with ompahlocele and unilateral Club foot in one child

Discussion

Conjoined twins, a rare entity in obstetrical practice, is a fascinating congenital abnormality with devastating consequences, both for the twins and the family. This is caused due to the failure of the division of the embryonic disc until after day 13 from conception. Sixty percent of the conjoined twins are still born [5]. But in our case, the babies were alive at the time of birth and died after two minutes of birth. There are no defined aetiological factors for conjoined twins, although there have been reports of geographical clustering [6]. Conjoined twins are more common among females than males and in nonwhites than whites. No maternal age effect was found [7]. In our case

also, the twins were female. The classification of the conjoined twins is based on the fused anatomic regions, followed by the suffix pagus. For example, craniopagus refers to head to head fusion, thoracopagus- chest to chest fusion and omphalopagus- abdomen to abdomen fusion [8]. Twenty-eight percent of conjoined twins are classified as thoracoomphalopagus [7], [8]. [9]. In our case, the conjoined twins were thoracoomphalopagus and were associated with the omphalocele and the unilateral club foot in one. The occurrence of club foot may be due to mechanical compression. There was no history of club foot in the family. Alkalay AL et al [10] and Karnak I et al [11] had also reported a similar case of conjoined twins with omphalocele earlier. Similarly, a report of six cases of conjoined twins published by Oronoy A et al [12], showed various congenital defects like gastrointestinal anencephaly, anomaly. urogenital anomaly, cleft lip, cleft palate and lumbosacral meningomyelocele. The liver is shared in 81% of the omphalopagus twins [9]. Similarly, in our case, there was a fused liver. Conjoined twins are always monozygotic and are associated with one chorion and one amnion Ultrasound is helpful in diagnosing [13]. conjoined twins. In case of monochorionic and monoamniotic twins, careful assessment should be done to rule out the shared parts between the foetus. The index of suspicion of conjoined twinning should increase if the twins maintain a constant and often unusual relative position and move together and if the neck and head are constantly hyperextended [3]. Though our case was diagnosed at term due to late presentation to the hospital, conjoined twins can be diagnosed even at the embryonal stage with the help of Doppler studies [1]. In more complex congenital anomalies, ultrafast MRI can be used as an adjunct to detect these.

Cesarean section is indicated when surgical separation and viability is possible without any complications to the foetus and the mother.

Conclusion

In conclusion, Thoracopagus associated with omphalocele and club foot is among the rarest in the conjoined twins and ultrasonography helps in it's proper diagnosis.

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