

Cephalophagus Non Janiceps: An Extremely Rare Variant of Conjoined Twins: A Case Report

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Submission: October 11, 2017; **Published:** November 14, 2017

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Abstract

Conjoined twins are uncommonly seen in twin pregnancy. Among conjoined twins, “*cephalophagus*” is an extremely rare variant. “*Cephalopagus*” twins are fused with their head, thorax and upper abdominal cavities. These twins are of two types: Janiceps (two faces are on the either side of the head) or non Janiceps type (with a relatively normal head and face). We present a case of non janiceps type cephalophagus variant of conjoined twins. The twins had a single head, apparently normal face, four upper limbs and four lower limbs. The twins were joined with their head, thorax and upper abdominal cavity. On ultrasound (USG) examination (done after delivery), the twins had two separate lungs and heart. The genitourinary system was unremarkable. This malformation was not detected early in pregnancy as the patient had no prior antenatal visits. These twins died within one hour of birth. Parents refused for autopsy.

Cephalophagus twinning is an extremely rare event with limited cases reported till date. Though an early prenatal diagnosis can be made by USG at 12 weeks gestation, it was missed in index case. Early diagnosis aids in termination of pregnancy thereby minimizing the mental trauma to the mother & family. Surgery is not attempted in these cases as the heads are fused and there is abnormal arrangement of the central nervous system. Autopsy must be considered in such cases for the better understanding of this congenital anomaly which may further shed light on embryogenesis & understanding the exact etio-pathogenesis of twinning.

Keywords: Conjoined twins; Cephalophagus; Embryology; Monozygotic twins

Introduction

Conjoined twins, popularly known as Siamese twins, result from aberrant embryogenesis [1]. It is a rare presentation with an incidence of 1 in 50,000 births. Since 60% of these cases are still births, so the true incidence is estimated to be approximately 1 in 200,000 births [2-4]. This disorder is more common in females with female to male ratio of 3:1 [5]. Conjoined twins are classified based on their site of attachment with a suffix ‘pagus’ which is a Greek term meaning “*fixed*”. The main types of conjoined twins are omphalopagus (abdomen), thoracopagus (thorax), cephalopagus (ventrally - head to umbilicus), ischopagus (pelvis), parapagus (laterally - body side), craniopagus (head), pygopagus (sacrum) and rachipagus (vertebral column) [6]. Cephalophagus is an extremely rare variant of conjoined twins with an incidence of 11% among all cases. These types of twins are fused at head, thorax and upper abdominal cavity. They are pre-dominantly of two types: Janiceps (two faces are on the either side of the head) or non Janiceps type (normal single head

and face). We hereby report a case of non janiceps cephalopagus conjoined twin, which was diagnosed after delivery.

Case Report

A 29 year old multigravida with history of previous one normal delivery presented to labor room in advanced labor. Her period of gestation was 32 weeks 2 days. She never had any ante natal visit to any health institute. Fetal malformation was never assessed by any ultra sonogram (USG). There was no history of consanguinity. Her previous birth was normal home conducted delivery one year back. In this pregnancy due to onset of labor pains in eighth month of gestation, she presented to hospital. Within few hours of admission, she delivered conjoined twins vaginally. On external examination these twins revealed a single head and thorax (Figure 1). There were four upper limbs and four lower limbs. The lower abdominal wall appeared separate for both the twins (Figure 1). The gender of both the twins was male. On the posterior aspect, the twins had two separate

occipital protuberances. An emergency sonogram (USG) was done to assess for the status of vital organs including hearts and lungs. USG examination revealed two separate hearts & two separate lungs. However, unfortunately these twins expired within one hour of delivery. The parents were advised for an autopsy; however they refused citing religious reasons [7,8].



Figure 1: Cephalophagus variant of conjoined twins (fused at head, thorax & upper abdominal cavity).

Discussion

Conjoined twinning is an uncommon complication of monozygotic twinning. Of all the monozygotic twin pregnancies, it has an estimated incidence of 1%. The first documented case of conjoined twin was that of Biddenden maids in 1100 AD, who lived together for 34 years and were joined at hips and shoulders [9]. In 1689, In Basel, Johannes Fatio performed the first successful separation on conjoined twins [2]. There are several varieties of conjoined twins and these are classified based on different classification systems. The embryonic classification of conjoined twins by Spencer classifies them into eight types based on the extent and location of fusion. The most common type in this classification include: thoracophagus, omphalophagus and thoraco-omphalophagus; all accounting for 56% conjoined twins [6]. Cephalophagus is the rarest variety of conjoined twins, accounting for about 11% of all conjoined twins. The twins in this disorder are fused with their head, thoracic and upper abdominal cavities. Cephalophagus twins are further of two types: Janiceps (two faces are on the either side of the head) or non Janiceps (with a relatively normal head and face). We found two occipital protuberances in the occipital region [10].

The index case represents the non janiceps type cephalophagus variety of conjoined twins. The gender of both the twins was masculine. However, it is believed that this malformation occurs predominantly in females, with a ratio of 3:1. Cephalophagus conjoined twins usually have four upper and lower limbs which are normal in appearance, as seen in the index case. USG findings suggested that the arrangement of internal

organs in this case was similar to that described in literature. Spinal cord revealed fusion of upper cervical segments with visualization of two separate vertebral columns caudal to it. There were two separate hearts and two lungs in fused single thoracic cavity. Unfortunately due to religious reasons parents refused for autopsy. Autopsy would have been the gold standard for exact delineation of anatomy of internal organs.

The autopsy results of cephalophagus twins in literature explain the unique features present in such cases. It has been found that the twins have two hearts in a single thoracic cavity, one of them is well developed and the other is rudimentary. The digestive system of these twins consist of single pharynx, esophagus & stomach and two separate intestines with a normal pathway to the pelvic region [7]. The embryological etiology for cephalophagus conjoined twins still remains an enigma. There are two different theories for the development of this bizarre anomaly; fissure theory and fusion theory. According to the fissure theory, there is incomplete separation of embryonic discs in 15th to 17th days of gestation. This theory was contradicted by Spencer and other authors who suggested the fusion theory which suggests the union between two distinct embryos taking place in early embryonic stage [2-4]. Each theory has its own strengths and limitations. Reaching a conclusive answer to the pathogenesis of conjoined twins is hindered by rarity of this event and inability to carry out conclusive experiments.

Certain pregnancy features alert the obstetrician to the possibility of conjoined twin pregnancy. These features include: poly-hydraminos (seen in 50% of conjoined twin pregnancies), relatively unchanged position of the fetuses over a long time, twin fetus with one fetal death, inability to localize two separate heads or inseparable bodies [11]. It is important to diagnose such pregnancies at an earlier gestation so that termination can be performed and mother & family do not suffer from mental trauma. Additionally timely diagnosis is also an essential aspect in planning obstetric management of conjoined twins. Currently, a prenatal ultrasound is commonly used for the diagnosis of a conjoined twin pregnancy and can be performed as early as twelve weeks of gestation. A detailed scan at twenty weeks of gestation provides a reasonable evaluation of the extent of the fusion, and any other issues [12]. Cephalophagus conjoined twins are usually non-viable due to numerous malformations present. The brain and spinal cord are believed to be extremely abnormal. Based on these circumstances, most authorities do not recommend surgical intervention. In majority of cases, cephalophagus conjoined twins do not survive until birth or die shortly after birth (as seen with index case). Nonetheless, autopsy examination of these twins can help in detailed understanding of the embryology and etio-pathogenesis of twinning as such.

Conclusion

"Non- Janiceps type of Cephalophagus twins" are extremely rare variety of conjoined twins. These twins have fused head, thorax and upper abdominal wall. The lower abdomen and

pelvic cavities are free. There are four upper limbs and four lower limbs. This variety of conjoined twins is usually non-viable due to extreme malformations of the brain and the spinal cord. Hence, surgical intervention in such cases are not attempted. Nonetheless, autopsies of such fetal outcomes can be very helpful in understanding the unique anatomic arrangement found in these twins which can highlight the exact mechanisms of embryology and help in better understanding of etio-pathogenesis of this unique presentation.

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