Undiagnosed live derodidymus, an intrapartum horror: a case report

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ABSTRACT

Background: Conjoined twins have invariably been a subject of revulsion among many societies across the world despite the fact that they can be diagnosed early in pregnancy and optimal obstetric management instituted accordingly.

Case Presentation: We present a case of a prenatally undiagnosed derodidymus (dicephalous diauchenos) twins, an extremely unusual variant of conjoined twins. The case was such a petrifying unanticipated phenomenon to both the parents and medical staff.

Conclusion: Routine prenatal ultrasonograhy and careful prenatal screening must be strongly emphasized if we are to minimize such perinatal mysteries.

Keywords: Case report, conjoined twins, derodidymus, dicephalous diauchenos, Siamese twins.

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Background

Conjoined twins, also commonly known as Siamese twins, are a very rare variant of monozygotic twins with high perinatal mortality and maternal morbidity [1]. A varying incidence of between 1:50,000 and 1:100,000 live births has been reported [2]. In twin pregnancies, the incidence is from 1:900 to 1:650 [3]. This is generally higher in Africa, more so in sub-Saharan Africa, and south-west Asia [4,5]. Conjoined twins represent a horrific congenital condition with varying myths attached to them by the society. Fortunately, the condition can be diagnosed early in pregnancy and optimal obstetric management instituted accordingly. However, in situations where prenatal diagnosis is not made, sudden encounter in the peripartum period becomes inevitable. Such a phenomenon is not without serious psychological consequences to the medical personnel, parents and relatives.

Case Presentation

An 18-year old Gravida 1 para 0 + 0 at 37 weeks and 5 days was admitted at our facility as a referral in from a peripheral health unit due to footling breech. At time of arrival, mother complained of spontaneous onset labor like pains that had started about 8 hours prior to the admission. These were notably intense regular and progressively increasing in intensity and frequency crampy lower abdominal pains that radiated to the lower back, associated with a show. She reported normal fetal movements. She had two prenatal visits from a nearby health facility where she was told everything was okay; however, not sure of the dates when she had the two visits, and had no any medical documents. Besides the reported human immunodeficiency virus negative test done on her during her first visit, no any other Uganda's recommended routine prenatal care protocol workups [6] including obstetric ultrasonography were done throughout her prenatal period. There was no family history of twins noted. On arrival, she was in moderate labor pains, all vital signs unremarkable. Fundal height was 37 weeks, breech presentation with two palpable uterine contractions over a period of 10 minutes' period that lasted 30 and 34 seconds, respectively. Fetal heart rate was 116 beats per minute and repeated at 5 minutes as 114 beats per minute. Vaginal examination revealed a fully dilated cervix with footling breech. Membranes were noted ruptured with meconium stained liquor grade II. An emergency caesarean section was done in view of footling breech and non-reassuring fetal status. We performed the delivery under spinal anesthesia and the fetal outcome was a conjoined male twin who scored 3/10 at 1 minute and zero at 5 minutes, birth weight 4.8 kg. The twin grimaced for a short while, and succumbed. It was such a petrifying experience as it was not anticipated. We were not able to do autopsy because

the parents declined the consent. Mother was managed postoperatively with iv crystalloids, antibiotics, analgesia, and vitals monitoring among others. Counseling was done to the mother, her husband, and relatives amidst strong myths and revulsion. Mother was discharged on her fourth postoperative day in a fairly good condition.

Discussion

Conjoined twins are identical twins with fused bodies [7]. The anomaly is associated with a lot of stigma not only to the parents but also to the entire family. It has been a source of fascination for both the public and the health care providers since time immemorial, their birth initially viewed as an ominous sign of impending disaster [8]. The etiology remains uncertain [9] although differing theories to explain their development have been put forth. One theory describes an incomplete splitting of one embryo into two. The other one describes fusion of a portion of one embryo from a monozygotic pair onto the other [10]. From an embryologic point of view, their formation typically results either from failed separation of the embryonic plate between day 15 and day 17 of gestation, or, from secondary union of two separate embryonic discs at the dorsal neural tube or ventral yolk sac areas at week-3 to week-4 of gestation [5]. Inclined on the background that the embryonic disk starts to differentiate on day 13, a split occurring after this time results in sharing of the body parts in addition to sharing the chorion and amnion.

Conjoined twins are generally classified based on their point of fusion. Because the fusion is always at specific homologous sites [11], the clinical classification is based on the most prominent site. The suffix "pagus" is attached to the nomenclature [12]. Cunningham et al. [10] has described nine variants of conjoined twins; six with a ventral fusion, that is; omphalopagus; thoracopagus, cephalopagus, ischiopagus, parapagus diprosopus, parapagus dicephalus, and three with dorsal fusions; that is craniopagus, rachipagus, and pygopagus. Thoracopagus twins are the most common, involving about 74%-75% [2] of all conjoined twins. We have presented an extremely rare case of conjoined twins apparently not featured in all the literature we accessed. These, as shown in Figure 1, were twins born with two separate heads, and two separate necks both heads and necks sharing all the rest of the body; that is attached on the same trunk with two upper extremities and two lower extremities. This according to Stedman's medical dictionary translates into an extremely rare form of conjoined twins called derodidymus, also known as dicephalous diauchenos, defined as one with "two heads and two necks". The definition, however, does not take consideration of the points of fusion, nor the details of the different parts of the body, for example, the number of arms involved, number of legs and so on. It is possible that deficiencies in appropriate nomenclature to describe all the different possible variants of conjoined twins exist, and whereas conjoined twins typically exhibit a female preponderance with a male to female ratio of 1:3 [13], ours was phenotypically male.

Conjoined twins are frequently diagnosed by ultrasonography in the mid-pregnancy. Early diagnosis during the first trimester has also been noted [14]. We have observed that some authorities recommend a follow-up imaging by magnetic resonance imaging (MRI) and computed tomography (CT) performed to confirm the diagnosis and further information [7,10]. The question of use of CT scan given the effect of radiations on the co-existent pregnancy however remains a point of analysis. Our patient had had two prenatal checkups and interestingly

Figure 1. *Images of the dicephalous diauchenos twins.*

all had been reportedly normal. She reported normal fetal movements. The mother had no any ultrasound scan done throughout the pregnancy period despite having had the two prenatal visits. This consolidates the role of routine obstetric ultrasound scans in pregnancy which could have allowed timely recognition of the anomaly and possible counseling of the parents on discontinuation of the pregnancy. This would minimize unnecessary stress, stigma, and other psychosocial problems that are likely to occur after delivery at late weeks of gestational age. Psychological problems aside, the delivery of conjoined twins has been associated with several other obstetric complications [5,7]. Early diagnosis of such anomalies would, therefore, enable the health care providers to anticipate and prevent such complications.

The lack of diagnostic modalities that aid in confirming the true diagnosis of such anomalies remains a serious challenge among most health facilities in the resource constrained countries. For instance; whereas imaging techniques such as X-ray or MRI, autopsy, and karyotype analysis of the fetus and the newborn could have contributed to the value of this report, these could not be accessed at this facility. This emphasizes the need to increase access to such assessment modalities by all health facilities, especially in resource constrained countries so as to enable timely establishment of the pathology in an event that such a scenario has been encountered.

Conclusion

Routine prenatal ultrasonography and careful prenatal screening must be strongly emphasized if we are to minimize such perinatal mysteries.

What is new?

Conjoined twins, although rare, are known to occur all over the world. This report presents an extremely rare variant of conjoined twins apparently not featured in all the literature previously.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this case report.

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None

Consent for publication

Written informed consent was taken from the family of patient.

Ethical approval

Ethical approval is not required at our institution for publishing an anonymous case report

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Summary of the case

1	**Patient (gender, age)**	Newly born phenotypically male conjoined twins, born to an 18-year-old primegavida
2	**Final diagnosis**	Derodidymus (dicephalous diauchenos)
3	**Symptoms**	Not Applicable (N/A)
4	**Medications**	N/A
5	**Clinical procedure**	N/A
6	**Specialty**	Obstetrics and gynecology