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CASE REPORTS

Conjoined Twins: Unicephalus Non-Janiceps Tetrapus Tetrabrachius

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Abstract Conjoined twins are an uncommon type of monoamniotic twins with an incidence of about 1.5 per 100,000 births worldwide. Non-janiceps is the extremely rare sub-type of cephalopagus which is reported as 1 in 58 of all conjoined twins or 1 in 3 million births. We present a G6, P5 31-year-old pregnant who was diagnosed with a unicephalus non-janiceps conjoined twins. Due to its incompatibility with life, the pregnancy was electively terminated. The prognosis of conjoined twins depends on the location and extent of the conjoined area and the accompanying congenital anomalies of the fetuses. Early diagnosis is vital because it can help in assessing the type of conjoined twins and subsequently pre-postnatal decision making. Elective termination of non-janiceps type of conjoined twins which is incompatible with life is a justifiable possibility.

Keywords Cephalopagus · Conjoined twins · Tetrabrachius · Tetrapus

Introduction

Conjoined twins are an uncommon form of monoamniotic twins, predicted to occur once in 50,000–100,000 births [1]. Female twins are more common than males [2, 3]. They are classified according to the site of fusion followed by the Greek postfix "-pagus," meaning "fixed", as parapagus (28%), thoracopagus (19%), omphalopagus (18%), cephalopagus (11%), ischiopagus (11%), pygopagus (6%), craniopagus (5%) and rachipagus (< 1%) [3, 4].

It can be diagnosed in the 1st trimester or early 2nd trimester. Ultrasonographic findings are single placental mass, no intertwin membrane and contiguous skin covering the fetuses. Embryonic/fetal poles are closely related and do not change their position with respect to each other. Congenital anomalies are almost always present and polyhydramnios is discerned in half of conjoined twins in late pregnancy [5].

The prognosis is poor because most die in utero or within the first 24 h in the newborn period [6]. The decision whether to continue the pregnancy depends on the type of conjoined twins and it necessitates multidisciplinary team and family counseling. We present a rare 20 weeks unicephalus non-janiceps conjoined twins which were electively terminated.

Case Report

A 31-years old, spontaneous conception, gravida 6 pregnant woman with a consanguineous marriage was referred at the 19 weeks of gestation with a suspicion of conjoined twins. Ultrasonographic examination revealed male conjoined twins having ventriculomegaly, hyperextension of fused cervical spines, two cerebellums, joined thorax showing two discrete spines, two separately beating hearts with atrioventricular septal defects, four upper and lower extremities (Videoclip S1, S2). Color Doppler depicted one umbilical cord. The family opted to terminate the pregnancy after being informed about the poor outcome. After the approval and consent of Trakya University Perinatology

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Council (number: 2021/135), the pregnancy was terminated. Macroscopic examination of the 320 g conjoined twins showed fusion from head to umbilicus and the presence of an enlarged single head, one face, two ears and eyes, one nose, four arms-legs with five fingers (Fig. 1). Autopsy yielded one cranium/cerebrum, two cerebellums, 2 hearts, 2 stomachs, 1 shared liver, 2 spleens, 4 kidneys). The left and right upper and lower extremities appeared normal. (Fig. 2).

Discussion

Conjoined twins are rare developmental anomalies and they are reported to occur with a prevalence of 1:50,000 in utero to 1:250,000 live births worldwide. Data about conjoined twins are restricted to case reports and case series, but the literature is insufficient. It is reported that the incidence varies between 1/50,000 and 1/200,000 in the United States and the highest incidence was observed in India (1/2,800) [1]. There is a female predominance in conjoined twins (70% female) [2, 3], but our twins had male sex according to their external genitalia albeit we did not perform a genetic analysis. The pathophysiologic etiology of conjoined twins is still not fully understood, but two theories about its pathogenesis were suggested. One theory (fissure theory) suggests that there is incomplete cleavage of the embryonic disc after the 13th day post conception [2]. According to another theory (fusion theory) which is not commonly accepted, conjoined twins are the result of an interaction between two embryos from completely separated fertilized eggs and there is secondary fusion between initially seperate embryonic discs [7].

Conjoined twins are classified as cephalopagus (head to umbilicus), thoracopagus (chest), omphalopagus (abdomen), ischiopagus (lower abdomen), parapagus (lower abdomen/pelvis), craniopagus (head), rachipagus (vertebral column), and pygopagus (sacrum/perineum), based on the site of fusion [4]. Our case is cephalothroracopagus or cephalopagus which encompasses one head, one face with tetrapus, tetrabrachius and it is subtype of craniopagus [8]. Cephalopagus twins are classified into two types: janiceps (two faces are on the either side of the head); or an extremely rare variety called non-janiceps (with relatively normal one head and a single face) which was evident in our case [9].

Congenital anomalies are almost always present in conjoined twins. Willobee et al. [10] analyzed predictors of in-hospital mortality in a series of 240 newborn conjoined twins. They detected that associated congenital anomalies were significantly common in conjoined twins, cardiac anomalies (36%) being the most common, which also were observed in our both twins. In another study major congenital anomalies unrelated to the site of union were reviewed and the most common anomalies were the genitourinary tract (19%) and the central nervous system (18%) [1]. We

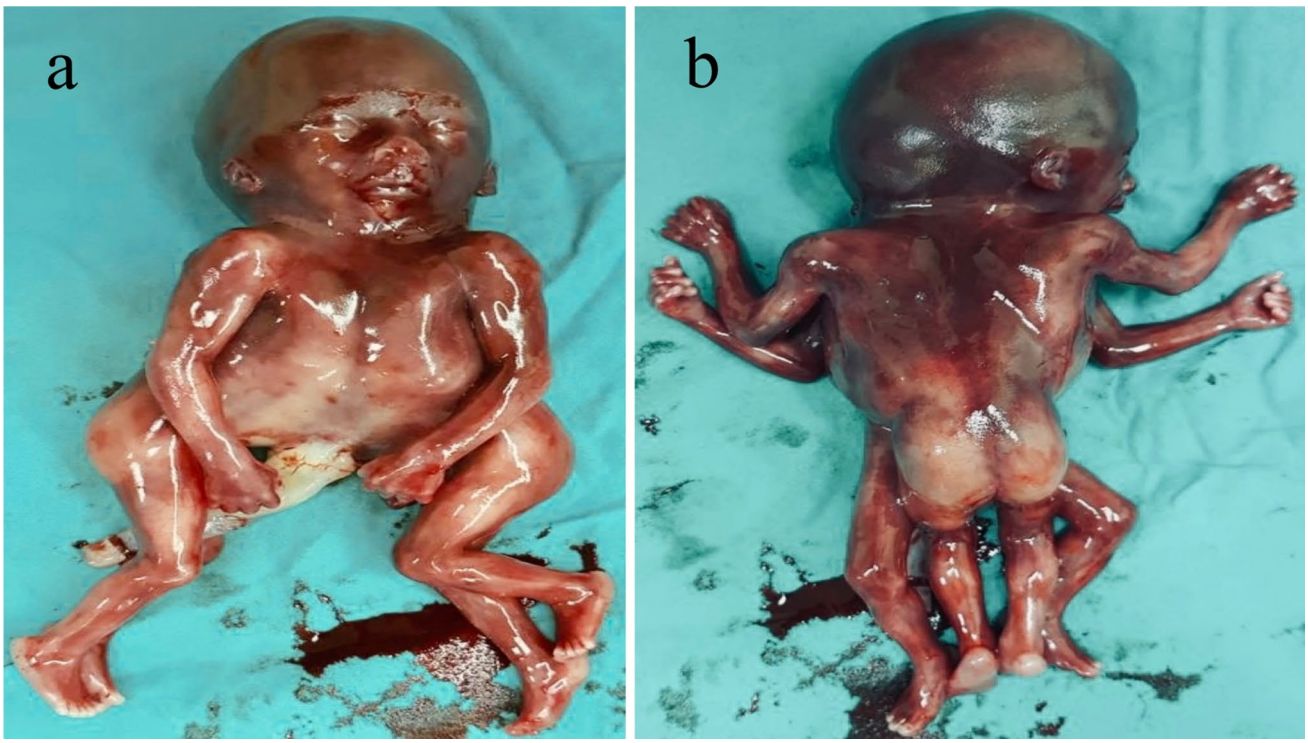

Fig. 1 Anterior (**a**) and posterior (**b**) macroscopic views of the conjoined twins

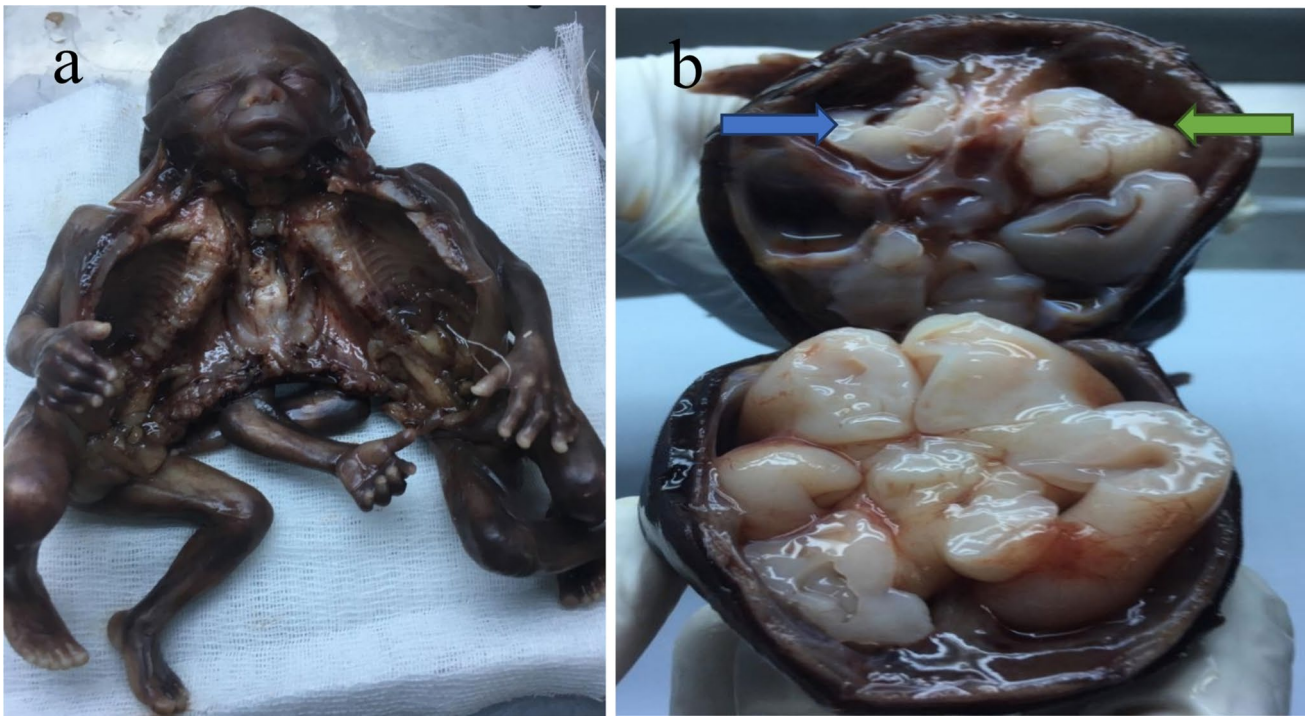

Fig. 2 **a** Autopsy appearance of the conjoined twins after removal of the internal organs. **b** Two different cerebellums are depicted with blue and green colored arrows

did not observe congenital anomaly unrelated to the site of union.

Conjoined twins are associated with a dismal outcome. It was predicted that 40% of them are stillborn while 35% die within the first 24 h [11]. Elective termination should be recommended for conjoined twins who have no chance of extrauterine survival or postnatal surgical separation. Site of fusion and accompanying malformations substantially affects the prognosis [12]. Thoracopagus type has the lowest survival rates because of the associated complex cardiac defects [2]. In our case, since the twins had a single cranium/face with shared brain which is incompatible with life, we offered elective termination. In the literature, successful separation of omphalopagus, ischiopagus, craniopagus or pyopagus types of conjoined twins have been reported. One center from Brazil studied outcomes of the management of 21 conjoined twins and reported 9 thoracopagus (only one could be separated), 8 ischiopagus (7 were separated), 3 omphalopagus (all were separated), and one separated craniopagus after sacrificing of the twin with microcephaly and sironemelia, with total surgical survival rate of 66.7% [13]. Separation of craniopagus twins is a complex procedure with ethical and technical considerations. A multidisciplinary team is essential to guide presurgical diagnostic evaluation in craniopagus twins [12]. A multimodal integrated imaging could provide comprehensive assessment of extracranial and intracranial anatomy before surgery [14]. Shared brain tissue, arteries and veins, and defects in the skull and dura cause surgery technically arduous [15].

In conclusion, prenatal diagnosis is indispensable to detect the type of the conjoined twins and decision to terminate the pregnancy. When there is a possibility of postnatal surgical separation, the decision to continue the pregnancy can be undertaken, yet multidisciplinary teamwork should be warranted. Non-janiceps type of cephalopagus twins are very rare, technically impossible to separate and incompatible with life. Hence, elective termination of the pregnancy should be contemplated.

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Declarations

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Consent for Publication The patient has given written informed consent to publish this case (including publication of images).

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