Case Report

Dicephalus dibrachius with anencephaly

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ABSTRACT

We present a case of inferior conjunction in a rare type of conjoined twins, dicephalus in a male fetus. The male fetus was born to a 24-year-old, gravida 2, and para 0, who had medical abortion at 15 weeks of gestation due to anencephaly with meningoencephalocele revealed by ultrasound examination. The fetus was born with 2 anencephalic heads with a bifurcation of the vertebral column and presence of 2 spinal cords. The other viscera and limbs were normal in number and location as for a male singleton. This case illustrates the relationship between conjoined twining, and neural tube defect (more particularly anencephaly) with a male zygote, which is an unusual presentation for this (type of zygote) gender.

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Conjoined twins have fascinated people for centuries. We increasingly accept them into our everyday lives as we grow to understand their unusual physical and emotional bonds and learn more about the science behind their development. Dicephalus dibrachius is an usual variation of conjoined twining. In this case, the fetus has 2 arms, 2 legs, and one trunk with 2 anencephaly heads. We should consider termination of the pregnancy, if antenatal diagnosis is made before 24 weeks of gestation. The aim of reporting this case is that it is one of the rare types of conjoined twining.

Case Report. A 24-year-old woman, gravida 2, para 0, abortion 1 was referred to our antenatal clinic due to presumptive diagnosis of anencephaly. The couple was a pair of non-consanguineous Indians with no significant family or medical history. Amniocentesis was performed at 15 weeks of gestation. Cultured amniocytes showed normal karyotype (46, XY) with a grossly elevated α-fetoprotein level. The 2-D ultrasound examination revealed the anencephalic male fetus with a large meningoencephalocele with abnormal shape of the spine. The heart, stomach, kidney, bladder, and insertion of the umbilical cord were normal in shape and position. Both upper and lower limbs were normal. After detailed prenatal and genetic counseling, parents decided on termination of preg-nancy as this is allowed for anencephaly in Bahrain. It was performed by oxytocic agents at 16 weeks of gestation. Macroscopic examination after termination of pregnancy revealed the following congenital anomalies; male fetus with 2 anenceph heads, single neck, and connected to a single broad chest (**Figure 1**). There was bifurcation of the vertebral column in which there were 2 spinal cords connected to the heads (**Figure 2).** The upper and lower limbs were normal for a singleton male fetus. Magnetic resonance imagining and skeletal survey confirmed the above diagnosis, and the parents declined post-mortem examination.

Discussion. Conjoined twins are a rare anomaly, with an estimated frequency of 1 in 50,000-100,000 births.[1] They develop from a single fertilized ovum and result when the embryonic disk completely divides more than 13 days after fertilization.[2] Dicephalus dipus dibrachius is an

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Figure 1 - Picture showing dicephalus conjoined twins with anencephaly.

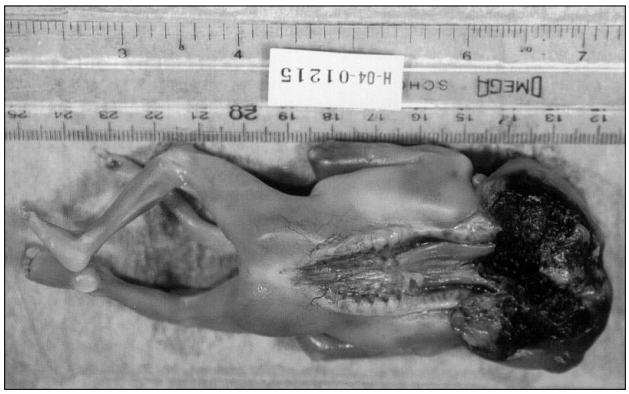

Figure 2 - Picture showing the fetus with bifurcation of the vertebral column.

extremely rare form of conjoined twin in which the infant has 2 arms, 2 legs, one trunk, but 2 heads. The infants are often stillborn or die shortly after birth.[3] Dicephalus with anencephaly is one of the rarest types. Dicephalus is considered an unusual variant of craniofacial duplication in conjoined twining. The phenotype comprises a wide spectrum and ranges from partial duplication (diprosopus) of a few facial structures to complete dicephalus.[4] Maggio et al[5] suggested criteria for ultrasonographic diagnosis of conjoined twins, which include inseparable fetal bodies and skin contours, single umbilical cords with more than 3 vessels, and a bifid appea-rance of the first trimester embryo should also raise suspicion for the diagnosis.

In our case the finding of dicephalus with encephalic heads makes it a very rare and unusual event in our hospital, with an annual delivery rate of around 6,000. Considering the incompatibility with life, early prenatal diagnosis and termination are of utmost important to save the patient from further psychological trauma.

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