Atypical Ischiopagus Conjoined Twins: Description of a Case

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Conjoined twinning is extremely rare congenital malformation occurring in a very low frequency. A 22-year-old woman presented with 33 weeks gestation to an obstetric clinic in active labor. The patient delivered conjoined male twins vaginally. External examination of the infants showed atypical ischiopagus twins. We emphasize that the precise knowledge of anatomic properties of both twins antenatally will help the obstetrician to decide the route of delivery and the pediatric surgeon to decide the surgical approach.

(Gynecol Obstet Reprod Med 2006; 12:000-000)

Key Words: Conjoined twins, Vaginal delivery

Conjoined twinning is a very rare condition with an occurrence rate of 1/50 000 to 1/100 000 deliveries. The types of conjoined twinning, in order of frequency, are thoracopagus, pygopagus, ischiopagus, craniopagus and, omphalopagus. Conjoined twins (CT) are generally diagnosed antenatally on the basis of certain radiological criteria and ultrasound findings since the wide use of ultrasonography prenatally. Here, we represent a rare type of conjoined twins.

Case Report

A 22-years old woman gravida 3, parite 2, admitted with 33 weeks gestation to our obstetric clinic in active labor. No history of maternal systemic illness and exposure to teratogenic agent or drug in pregnancy was noted. In her family history, there was no congenital abnormalities or twinning and she had never applied to an obstetrician during her pregnancy.

On physical examination, the vital signs of the patient were normal. Vaginal examination revealed a cervix completely dilated and effaced. The presenting part was a soft tissue. At that time no cardiac activity could be detected. After 30 minutes, the level of presenting part was not changed. So, we pulled the soft tissue down manually and had an extremity attached to that soft tissue that was pulled down. Then the patient delivered conjoined male twins vaginally with unexpected ease after that maneuver. Birth weight of the conjoined twins was 2180 g. length 45 cm and the only cephalic circumference was 32 cm. We inspected omphaloischio-pagus twins. One of the twins had an incomplete cranial development. Both had normal male external genitalia. There were four upper limbs and four lower limbs. One of the lower extr head with extremities was like a hoof. The dominant fetus had a well-developed normal appearance although the other fetus had no cranial development. There was also a large omphalosele in the abdominal region (Figure 1). The placenta was normal. There were two umbilical cords with one artery and one vein for each histologically. The postpartum
The course was uneventful so the mother was discharged on the second day.

The atretic vertebral column and the costae of the second twin could be seen beneath the dominant twin. The radiographic characteristics of conjoined twins are seen in Figure 2. The postmortem autopsy could not be done because the parents did not give permission for this examination.

Discussion

Conjoined twins have always fascinated physicians and people. The etiology is unknown, but the most accepted theory suggests that an incomplete cleavage of the embryo at approximately 2 weeks gestation is responsible for these malformations. They are therefore monozygotic and monochorionic, with the same chromosomal pattern and same sex. The prognosis is related to the type and extent of union. Female infants predominate with a ratio of 3:1. But in our case the infants were male. Conjoined twins were classified as typical and atypical also known as heteropagus. In heteropagus cases, one of the twins (parasite) is smaller and dependent on the other (autosite). Our interpretation about this case is that it is not a heteropagus twinning because the extremities of the second twin are well developed and the second baby was anencephalic. In our opinion cranial maldevelopment is due to the possible vascular accident that resulted in anencephaly. It is reported that early diagnosis and precise delineation of the shared organs of conjoined twins are essential for optimal obstetrical and postnatal management.

But there was no prenatal diagnosis in our case, so we could not planned better management of the pregnancy and the delivery, which might be essential to have twins alive. In late twin pregnancies, if there is an antenatal diagnosis the decision regarding vaginal delivery vs. cesarean section is based on the size of the fetuses and likelihood of survival. In our case as there was no cardiac activity and being unaware of the conjoined twinning we preferred vaginal delivery and the labor was uneventful as the patient was at 33 weeks gestation and the twins were not too big. Even though the twins were alive, it was impossible to separate them surgically because of the complicated conjunction.

We emphasize that the antenatal precise knowledge of anatomic properties of the twins such as the size of the fetuses and type of conjunction, could help the obstetrician to manage the pregnancy and to decide the route of delivery. This adequate prenatal and delivery management would help the pediatric surgeon to have a good start for successful twin separation procedure.

References