Management of Rachipagus Parasitic Twin.

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الخلاصة

حدوث التوأم المتحدين اللامتماثل والطفيليين نادر, وفي الوقت الحاضر يتم تصنيف هذه الحالات بشكل قياسي وفقا للالتصاقات بين التوأم الطفيلية المتماثلة ,سجلت حالة ولادة بعملية قيصرية للام في الاسبوع الرابع والثلاثين للحمل وكانت لرضيع ,ذكر يزن ثلاثة كيلوغرام وستمائة غرام مع وجود توأم طفيلي ملتصق من جهة أعلى منطقة الظهر في منطقة الفقرات الصدرية والقطنية ,تضمنت اجزاء الطفيلي طرف أعلى ناقص ,عمود فقري وحبل شوكي ,كبد ,أمعاء دقيقة, نسيج رئة , وغدة كظرية. اعتبرنا هذه الحالة نادرة لوجود الطفيلي الشاذ في مثل حالات التوامة هذه وهي أول حالة سجلت في المستشفى,وقد اجريت عملية لفصل التوأم الطفيلي في مستشفى النسائية والأطفال التعليمي , وحدة جراحة الأطفال و تكللت بالنجاح .

Abstract

Occurrence of asymmetrical or parasitic conjoined twins (CT) is rare, and currently they are classified analogically to the common unions of symmetrical CT. We report on an infant with a parasitic third limb attached to the back at thoracolumber area , in whom male genitalia was found . Parasite parts included incomplete upper limb, hemipelvis, thoracolumber vertebral column, spinal cord, and one kidney with ureter ,small bowel, lung tissue and adrenal gland. Auto site anomalies comprised evidence of thoracolumbar meningomyelocele. We considered this case to be a rare atypical parasitic rachiopagus CT. The differential diagnosis of the type of twining and other entities with caudal duplications is analyzed briefly . We report a case of rachipagus parasitic twin diagnosed delivered a live baby at 34 weeks of gestation to 30-year-old primgravida woman with history of 10 years secondary infertility .

Introduction

Asymmetric and parasitic conjoined twins are rarer anomalies of monochorionic monoamniotic twins, consisting of an incomplete twin attached to the fully developed body of the co-twin. Prenatal diagnosis of conjoined twins with B-mode ultrasound (US) [1 , 2], computed tomography (CT) [3], threedimensional US [4 , 5], and magnetic resonance imaging (MRI) [6] has been reported. Parasitic conjoined twin diagnosed in utero was presented in few cases previously [7, 8].

But literature review failed to identify a case of pygopagus tetrapus parasitic twin detected in utero. A fetus-in-fetu is an encapsulated, pedunculated vertebrate tumor. It represents a malformed monozygotic, monochorionic diamniotic parasitic twin included in a host (or auto site) twin. Characteristically the fetus-in-fetu complex will be composed of a fibrous membrane (equivalent to the chorioamniotic complex) that contains some fluid (equivalent to the amniotic fluid) and a fetus suspended by a cord or pedicle. The presence of a rudimentary spinal architecture is used to differentiate a fetus-in-fetu from a teratoma, since teratomas are not supposed to develop through the primitive streak stage (12-15 days). The prevalence is unknown. About 70 cases have been reported but this number varies according to how strictly identification criteria are used. Several cases have been formally recognized by some as fetus-in-fetu, while categorically rejected by others. Seven cases have been detected in utero. An estimated frequency of 0.02:10,000 is commonly reported in the literature, but this number is based on the unsubstantiated assumption that fetus-in-fetu represents 5% of conjoined twins. Further, the current trend is to consider that fetus-in-fetu does not represent a form of conjoined twins. The male-to-female ratio in the 39 reports that we reviewed was M1.3:F1. This is in contrast with conjoined twin, which occurs predominantly in girls [9, 10, 11, 12, 13, 14, 15].

Case presentation

The baby was born at a gestational age of 34 weeks to 30-year-old primgravida woman with history of 10 years secondary infertility, on clomid tablets, healthy, nonconsanguineous parents, without maternal illness, drug intake, or familial congenital abnormalities or twinning. Previous ultrasonographic examination reported polyhydramnios. Birth done by caesarean section, and 2 days later the patient was admitted to the Matrnity and Child Teaching Hospital –pediatric sugery unit. Birth weight was 3.600 g, length was 45 cm, and cephalic circumference was 32 cm. Apgar scores were 7 and 8 at one and 5 minutes, respectively. Characteristics of the membranes and placenta were unavailable. Upper trunk external appearance was normal. There were 3 upper limbs (one with three fingers attached to the parasitic mass in thoracolumber at the back) and 2 lower limbs. The parasitic vertebral column showed a well-differentiated spinal cord and, beside it, a third morphologic left kidney with adrenal gland and ureter of indefinable destination (U\S finding). Parasite structures were vascularized via major anastomosis. And regarding the precise history of the mother, a 30-year-old multigravid woman referred to maternal unit for targeted ultrasonography due to polyhydramnios at 28th week

of gestation. The woman had no family history of congenital anomalies and had taken clomid tab. during her pregnancy. Her obstetric background consisted of two first trimester abortions. Screening for gestational diabetes was negative. Sonographic examination initially revealed a single fetus and polyhydramnios with amniotic fluid index 30 cm. The fetus appeared normal except a mass at sacrum. Further evaluation with U\S identified that the fetus has normal apparent single head, spine, thorax, abdomen, two upper and two lower normal limbs, and two relatively well developed rudimentary upper limbs at thoracolumber area . Detailed anomaly scanning of the autosite with U\S revealed normal findings except the parasite at thoracolumber area region. Upper and lower limbs of the auto site were moving freely but no movement was detected at the parasite. Short and deformed long bones were also present in the parasitic limbs. The parents were counseled and were informed by the gynecologist. A Cesarean section was performed at 34th week of gestation and a live male infant weighing 3600 g was delivered. The placenta was single and normal. The infant appeared to be normal except the parasitic co-twin (Figure 1, Figure 2 and figure 3). The infant was further evaluated with US and MRI that confirmed that parasitic conjoined twin had relation with the spinal cord at T 7 to L 2 thracolumber vertebrea. One week after birth, the infant underwent surgery. The parasitic upper limbs were totally excised with whole the parasitic tissues [figure 4, figure 5]. Post-operative period was uneventful and the newborn was discharged as healthy. Post-natal and post operative follows-up were normal at three-month-old. Pathologic examination demonstrated skin covered parasitic body , Parasite parts included incomplete upper limb, hemipelvis, thoracolumber vertebral column, spinal cord, and one kidney with ureter ,small bowel, lung tissue and adrenal gland. At three month of age the infant developed hydrocephaly the C-T scan revealed evidence of ventriclomegaly the parents were counseled again by pediatric surgeon and neurosurgeon for V-P shunt which was done at Al-Najaf Teaching Hospital \ neurosurgical unit , post operative follows-up were normal at 1.5 year-old.

Conclusions

Conjoined twinning is a fascinating congenital abnormality with devastating consequences for the twins and the family. Conjoined twins develop from a single fertilized ovum and result from failure of the division of the embryonic disk until after day 13 from conception. Seventy percent of conjoined twins are female, and 40% are stillborn [16]. Conjoined twins are categorized based on the region of connection. In one attempt to universalize the current nomenclature of

conjoint twins, Spencer [17] proposed a simple and logical new classification based on the theoretical site of union. Parasitic twins are rare form of conjoined twins and are consisting of an incomplete twin (parasite) attached to the fully developed body of the co-twin (auto site). It is classified [18] as (1) an externally attached parasitic twin, (2) an enclosed fetus in fetu, (3) an internal teratoma, or (4) an acardiac connected via the placenta. Prenatal recognition of conjoined twins and precise characterization of the malformations are required for optimal obstetric management. Currently, sonographic diagnosis of conjoined twins may be straightforward if fusion of fetal parts is obvious. In utero diagnosis of conjoined twins was reported previously [19-22]. But prenatal diagnosis of parasitic conjoined twins is relatively difficult becasue of changing fetal positions and the rarity of these abnormalities as with any rare medical entity. Parasitic conjoined twins identified at birth were reported [23-26], but so far two cases diagnosed *in utero* with US and MRI were presented in the literature [27, 28]. Polyhydramnios is a secondary finding that occurs in 50% of conjoined twin pregnancies [29, 30]. Hence the site and extent of twin fusion are variable; a careful evaluation with US may help to identify parasitic conjoined twins *in utero*. Differential diagnosis includes parasitic twin, fetus in fetu (internal parasite) and teratoma. Presence of rudimentary lower limbs with foot in our case suggested the parasitic conjoined twin. The prognosis depends on the site and extent of twin fusion.In our case, during routine prenatal examination, the woman had three US scanning and had no abnormal findings noted until she was referred for polyhydramnios at 34 weeks' gestation, suggesting that prenatal diagnosis is relatively difficult. Rarity of the condition may lead to misdiagnosis or undiagnosed. Although our case was referred for detailed U\S due to polyhydramnios, its etiology is not clear and parasitic site might have vascular malformation that may lead to increased volume of circulation, like in the cases having teratoma and arteriovenous malformations. Hyperdynamic cardiac activity and increased cardiac output might casued polyhydramnios in our case. Even though it is rare, a parasitic conjoined twin should be considered in the differential diagnosis when polyhydramnios is present. Prenatal diagnosis in our case helped in counseling with the parents and in management of the case and delivery.Rachipagus parasites (most consisting only of supernumerary limbs) show vertebral union above the sacrum with neural or bony connections to the auto site [31, 32]. Monitoring must be continued postoperatively in the ICU. Because surgery is prolonged. Fluid and electrolyte balance should be closely monitored. Sepsis is a major cause of morbidity and mortality, and precautions must be exercised, particularly when large

skin defects are present.In conclusion, rachipagus parasitic twin is a rare form of conjoined twin with a favorable outcome. Obstetrician should be aware of the existence of a parasitic twin during prenatal examinations and the importance of the differential diagnosis of parasite and teratoma. Delivery at a tertiary center is highly suggested for optimal neonatal intensive care and pediatric surgical intervention.

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Figure 1 preoperative.

Figure 2 per operative view.

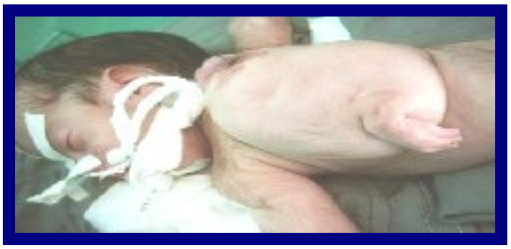

Figure 3 per operative view.

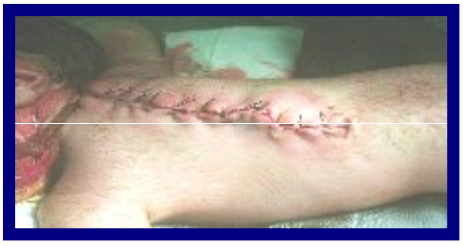

Figure 4 postoperative, sites Of excision of the parasitic mass.

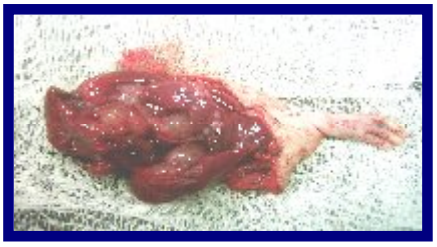

Figure 5 postoperative, complete excision of the mass, limb with tree digits, tissues of liver with small gallbladder, small bowel and spleen.