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CASE REPORT

In utero and postnatal imaging findings of parasitic conjoined twins (ischiopagus parasiticus tetrapus)

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Abstract Conjoined twins are a rare developmental anomaly with a reported prevalence of 1.47 per 100,000 births. We present an uncommon case of a parasitic ischiopagus tetrapus with a parasitic ischiopagus partial twin joined to the complete fetus at the level of the ischium diagnosed in utero by fetal MRI. The correct prenatal diagnosis led to birth by caesarean section. Prenatal MRI findings are presented and corroborated by postnatal imaging delineating the full extent and associated anomalies of this rare malformation. Differential diagnosis of duplicated lower extremities is discussed.

Keywords Conjoined twins · Prenatal diagnosis · Magnetic resonance imaging · Computed tomography · Fetus · Neonate

Introduction

Conjoined twins are a rare developmental anomaly with a reported prevalence of 1.47 per 100,000 births; parasitic conjoined twins are even rarer with a prevalence of 0.057 per 100,000 births in a recent worldwide multicenter study that reviewed more than 26 million births [1].

There are a variety of classifications for conjoined twins, but most are based on the site of union (i.e. craniopagus, pygopagus, thoracopagus, cephalopagus, parapagus, ischiopagus, omphalopagus, rachipagus, parasitic twins and fetus in fetu) [2].

Parasitic conjoined twins occur when monozygotic twins do not separate completely and one embryo's development prevails over the other. It is called parasitic rather than conjoined because one embryo stays incomplete or is entirely dependent on the body functions of the complete fetus [1, 3, 4].

We report a case of an ischiopagus parasiticus tetrapus with a parasitic twin joined to the complete fetus at the level of the ischium diagnosed by fetal MRI.

Case report

A 28-year-old woman, gravida 2, para 2 was referred for fetal MRI in the 22nd week of gestation, with sonographic diagnosis of fetal hernia into the cord, fetal clubfoot and suspicion of caudal regression because one out of two sonographically visualized legs was malpositioned with the foot lateral to the abdominal wall and did not show any motion. The parents were non-related, the family history was negative for congenital malformations and no teratogenic exposure during pregnancy was known.

Fetal MRI was performed on a 1.5-Tesla scanner (Discovery 450; GE, Milwaukee, WI, USA) with a multiarray coil covering the mother's abdomen. T2-weighted single-shot fast spin-echo (SSFSE) images were acquired in three orthogonal planes aligned to the mother's pelvis (TR: 1,268 ms, TE: 100 ms, slice thickness: 5 mm), fetal head (TR: 5,100 ms, TE: 80 ms, slice thickness: 3.5 mm) and body (TR: 5,100 ms, TE: 80 ms, slice thickness: 4.0 mm). The study was completed by sagittal dynamic 2-D fast imaging employing steady-state acquisition (FIESTA, flip angle: 45°, TR: 3.3 ms, TE: 1.1 ms, slice thickness: 14 mm) and axial T1-weighted fast spoiled

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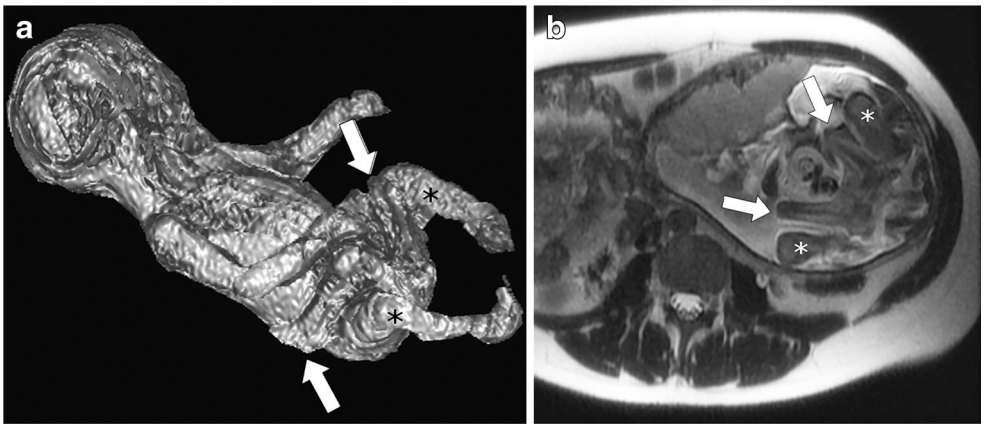

Fig. 1 Prenatal MRI of a fetus in the 22nd week of gestation shows ischiopagus parasiticus tetrapus. *White arrows* in 3-D volume-rendering of an SSFSE stack (**a**) and in axial SSFSE image (**b**) show cranially oriented, hypoplastic additional legs. The normally oriented legs are marked with *asterisks*

gradient echo (FSPGR, flip angle: 80°, TR: 100 ms, TE: 4.2 ms, slice thickness: 3.5 mm) sequences.

An ischiopagus parasiticus tetrapus was diagnosed based on the following MRI findings (Fig. 1). A single fetus was seen with normally developed neuroaxis and without abnormalities of the upper limbs. Chest and upper abdomen were normal, except for a small hernia into the cord, i.e. suspected omphalocele. There were four lower extremities with two legs normally articulating to the pelvis, showing spontaneous movements and feet held in a supinated position consistent with bilateral clubfoot. Two additional, complete but hypoplastic legs were found inversely oriented and crossing the normally orientated legs, originating ventral and caudal from a second hypoplastic pelvis and extending cranially with the knees facing anteriorly in 90° flexion and the feet at the sides of the fetus. These additional legs showed no movements.

The 3,375-g boy was born at 37 weeks' gestation by caesarean section with normal primary adaption after delivery. Clinical inspection confirmed an accessory, ventrally and cranially oriented pair of hypoplastic legs, the omphalocele and a right-side clubfoot. Additional findings were high anal atresia without fistula and duplicated scrotum with one testicle on each side and two penises with hypospadia (Fig. 2).

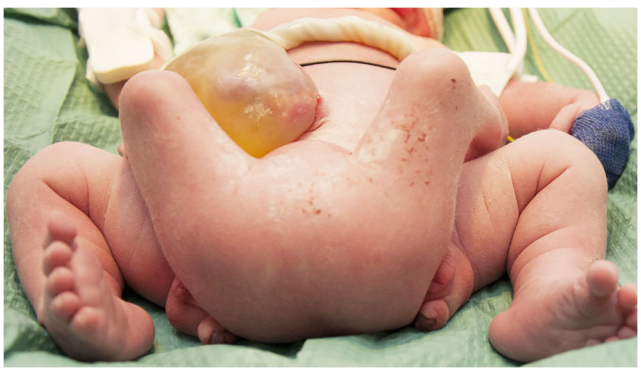

Fig. 2 Photograph of ischiopagus parasiticus tetrapus postpartum shows the duplicated penises, the parasitic partial twin and the omphalocele

Imaging work-up of the neonate by radiographs (Fig. 3), low-dose multidetector helical CT of the lower extremities (20 mAs, 80 kV, reconstructed slice thickness: 0.9 mm, size-specific dose estimate: 23 mGy) (Fig. 4), MRI (Fig. 5) and voiding cystourethrography (VCUG) confirmed a normal thoracic and upper abdominal situs with two orthotopic kidneys, normal thoracic cage and spine. The omphalocele contained blind-ending, T2-intense fluid-filled intestinal structures surrounded by a membrane (duplicated intestinal structures). The dilated and blind-ending rectum was filled with T1-intense meconium (Fig. 5). The pelvis was enlarged, with duplicated and fused pelvic bones anteriorly and anteriorly articulating with the accessory legs (Figs. 3, 4). In the pelvis, there was a third kidney (Fig. 5). All three kidneys drained separately into a single bladder. As the patient was urinating through both penises, both urethras were catheterized and VCUG confirmed their origin from the single bladder.

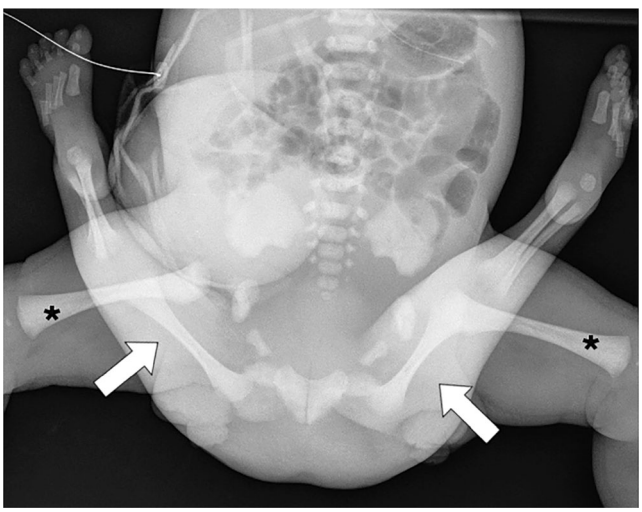

Fig. 3 Radiograph of the neonatal ischiopagus parasiticus tetrapus. *White arrows* show cranially oriented, hypoplastic additional legs. The normally oriented legs are marked with *asterisks*

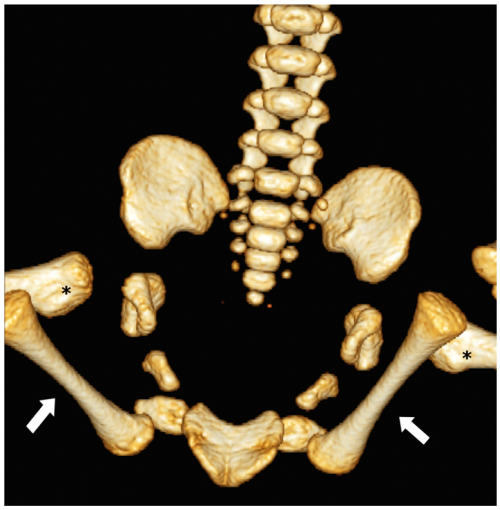

Fig. 4 Volume-rendered CT image of the neonatal ischiopagus parasiticus tetrapus. *White arrows* show cranially oriented, hypoplastic additional legs articulating with an inverted, rudimentary and anteriorly fused additional pelvis. The normally oriented legs are marked with *asterisks*

For assessing the blood supply of the parasitic ischiopagus twin, contrast-enhanced magnetic resonance angiography was performed after intravenous injection of 1.2 ml gadolinium-based contrast agent (Dotarem; Guerbet AG, Zürich, Switzerland) in a cubital vein and with automated bolus detection in the abdominal aorta. A single artery originating from the right internal iliac artery was found supplying the contents of the omphalocele, the accessory kidney and legs. Molecular genetic analysis proved the twins to be monozygotic.

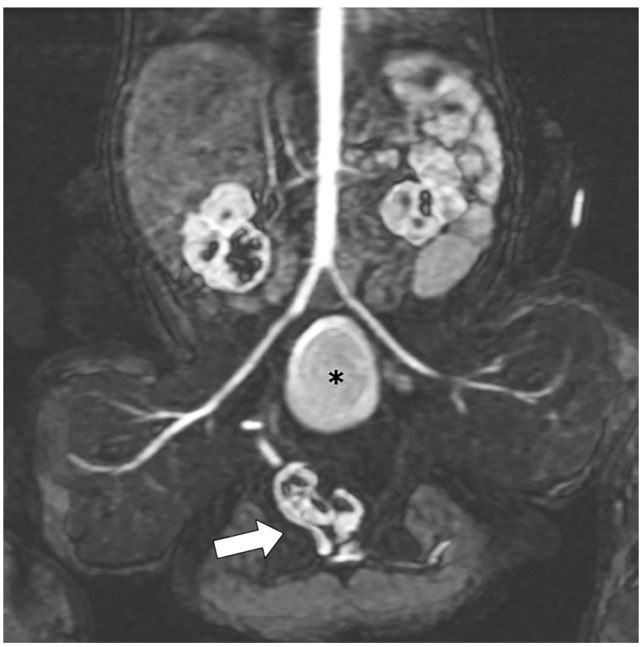

Fig. 5 Coronal contrast-enhanced MRI of the abdomen of the neonatal ischiopagus parasiticus tetrapus. Arrow points at additional pelvic kidney. Asterisk marks the meconium-filled rectum

Following the imaging evaluation, the neonate underwent emergent resection of the duplicated bowel in the omphalocele with primary abdominal wall closure. At the same time, anal atresia was palliated with a dismembered colostomy. At the age of 4 months, the infant is developing normally. Orthopedic and genitourinary reconstruction by a multidisciplinary team is planned at the age of 1 year to minimize the operational risk. Reconstruction of the anal tract is planned at a later stage.

Discussion

With the presence of four legs and associated intestinal and genitourinary anomalies, several diagnoses have to be considered, e.g., dipygus, congenital duplication of lower extremity human disorganization syndrome (HDS) and all types of conjoined twins except parapagus.

In our case, the cranial and ventral orientation of the additional legs originating from a ventrally duplicated rudimentary pelvis were a clue to the diagnosis of ischiopagus parasiticus tetrapus, as this is the only type of conjoined twins presenting with legs oriented in opposite directions. Dipygus would be a rare malformation in humans with a fetus with completely duplicated legs all oriented in the same direction [5]. Congenital duplication of parts of the lower extremities is more common with fewer then 40 cases reported in recent literature [6]. In pygopagus and rachipagus conjoined twins, the two pairs of legs would be located back to back but both oriented caudally. HDS is a very rare malformation syndrome including disfigurations triggered by disorganization of morphogenetic induction in the first weeks of gestation, which could result in supernumerary limb-like structures anywhere in the body [7].

Although one would expect to see the four legs on fetal sonography, in our case the ischiopagus twins were mistaken for caudal regression even by a senior obstetrician of a tertiary centre with vast experience in fetal sonography, probably because the normally developed legs were hidden behind the caudal part of the fetus. The correct diagnosis was only made with the less operator-dependent fetal MRI, which was important for deciding on delivery by caesarean section in a tertiary hospital. The fact that anal atresia, the pelvic kidney and complex duplication of the genitourinary tract were not evident on fetal MRI underlines the need for comprehensive postnatal imaging including CT, MRI with angiography and VCUG for fully delineating the bony, visceral, genitourinary and vascular anomalies in conjoined twins, which is essential for planning the staged surgical treatment [8].

Conflicts of interest None

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