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Short Clinical Report

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Heteropagus parasitic twins – a case of omphalopagus with major omphalocele and congenital heart disease

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CASE PRESENTATION

A 3250g, female second twin, known antenatally to have an omphalocele and interventricular communication, was delivered by elective cesarean section (pelvic 1st twin) to a 33-year-old, G1P1, mother at 36 weeks' gestation, at the referral tertiary level maternity of a university hospital center. It was a monochorionic diamniotic pregnancy through a donor in vitro fertilization. No obvious exposure to teratogens was documented during gestation. Both fetuses had negative karyotype for T21 and T13 and normal microarray.

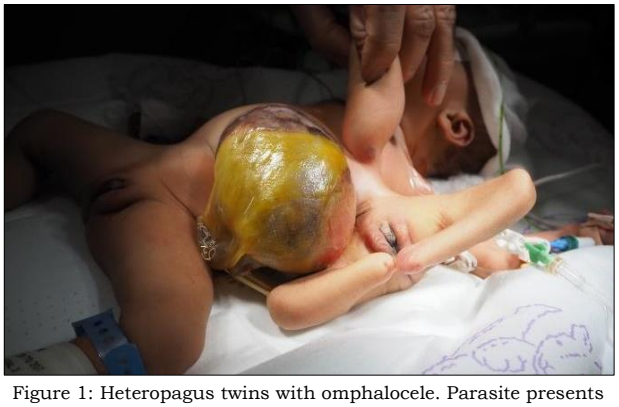

Figure 1: Heteropagus twins with omphalocele. Parasite presents with dysmorphic legs and arms, pelvis, buttocks, and genitalia.

Both newborns were hemodynamically stable. Examination of the second, conjoined twin found an acardiac acephalic parasitic twin with dysmorphic legs and arms, pelvis, buttocks, genitalia, and a soft tissue pedicle emanating from the xiphoid region of the primary twin. The parasite's urethra showed some urine flow. The autosite twin also presented an omphalocele with an intact sac (Fig.1). Postnatal echocardiography revealed perimembranous ventricular septal defect, muscular ventricular septal defect, and patent foramen ovale. Three-dimensional magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) revealed that both intestine and liver were present in the omphalocele and found the blood supply to the parasitic twin originating from an aberrant branch of the left iliac artery of the autosite twin.

On the 3rd postnatal day, the twins were separated with dissection of the pedicle, which consisted of an articular structure that connected one of the parasite's limbs to the autosite twin's sternum. The parasite had an atrophic otherwise normal kidney with renal vessels (one artery and vein), no adrenal glands, and an umbilical vessel and cord that also connected both twins (Fig.2). Ligation and excision of the structures allowed total separation between the autosite and the parasite. All abdominal organs of the primary twin were revised, and no anomaly was found. Primary closure of the omphalocele was successfully achieved.

The patient received vancomycin and fluconazole for 10 days, due to the wound infection. The postoperative period was uneventful, allowing rapid weaning from the respirator and fentanyl. The patient remained hemodynamically stable until the 13th postoperative day when she showed signs of congestive heart failure. Furosemide and fluid restriction were initiated. An echocardiogram at this point confirmed the perimembranous and muscular ventricular septal defects and the closure of the foramen ovale. The patient was transferred to the Pediatric Cardiology Unit at 34-day-old, weighing 2730g, and was discharged home 11 days after weighting 3000g on furosemide, captopril, digoxin, and oral iron supplementation. On the last Pediatric Cardiology assessment, at 13 months of age, she was growing well and uneventfully, an echocardiogram showed improvement of both ventricular septal defects and she progressively began to reduce captopril and digoxin.

DISCUSSION

The authors describe a case of female heteropagus twins, from a diamniotic, monochorionic placenta,

after medically assisted reproduction (sperm donation, embryo transfer), with a typical chromosomal pattern identified on amniocentesis. There were no identifiable risk factors. In the heteropagus form, a fusion occurs between a smaller incomplete parasite and a complete autosite. In this case, the junction is above the umbilicus, and the connection between them was a tubular structure formed by a rudimentary articulation, umbilical vessel, and cord and so it is referred to as omphalopagus twins. [1]

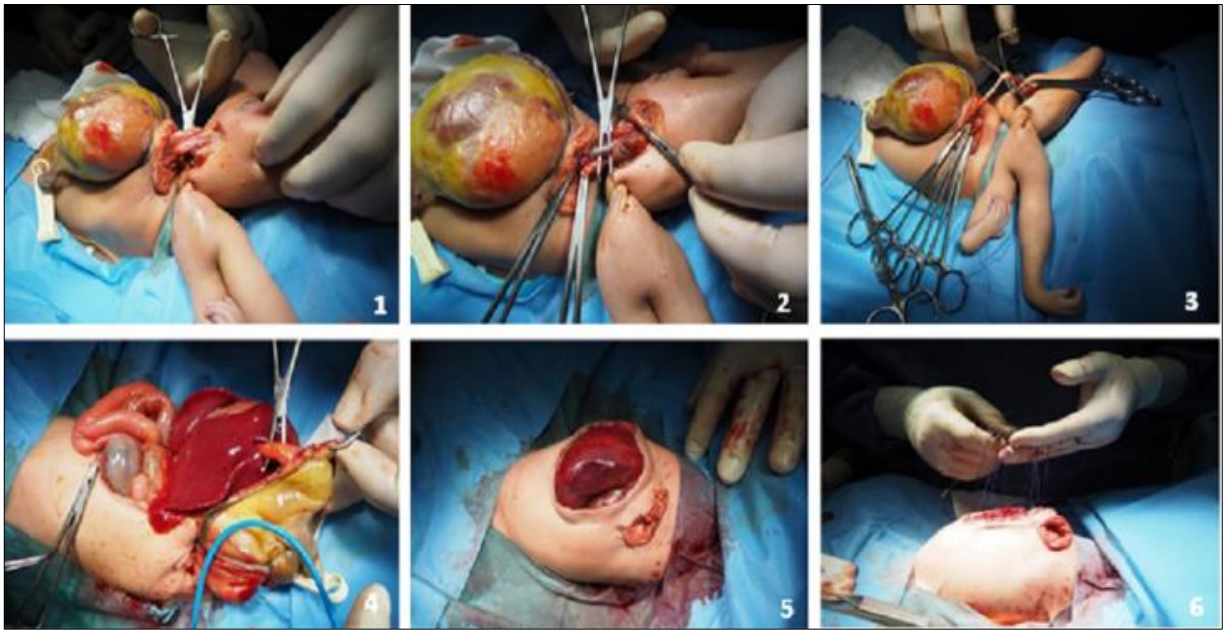

Figure 2: Twin's separation with isolation and ligation of the pedicle and omphalocele's primary closure.

Omphalocele is the most associated anomaly in the autosite, followed by cardiac anomalies. [2-4] In this reported case, both the omphalocele and cardiac anomalies were present in the autosite. Most parasitic twins are diagnosed with prenatal ultrasound as in this case. Since their prognosis is favorable, there is no need to end the pregnancy. [5] Prenatal echocardiography has a role in detecting cardiac defects as there is a high prevalence of congenital heart disease in autosite, particularly in the omphalopagus. Congenital heart disease has been described in about half of the reported cases as in this case, in which prenatal findings were confirmed postnatally. [2,6-8]

As there is great anatomical variation among cases, preoperative planning and subsequent operative approach must be tailored to the unique presentation of the patients. Preoperative imaging can involve ultrasound, CT, or MRI. In this case, MRA was performed, disclosing the origin of the blood supply to the omphalopagus.

Different surgical techniques are available, depending on the sharing of organs. In this case, surgery was simple and uncomplicated, as there were no bony and/or visceral communications between twins. Repairing omphalocele and closing the defect created by the removal of the parasite was also straightforward without using flaps or tissue expanders. Postoperative was uneventful except for wound infection, which is one the most common complications in this procedure, followed by an incisional hernia and teratoma formation.[7] The patient got complicated later with mild congestive heart failure, a main prognostic factor. [2,6].

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