Case Report

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Thoracic heteropagus conjoined twins associated to an omphalocele: Report of a case and complete review of the literature

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ABSTRACT

Heteropagus twins are an extremely rare event with an incidence of 1–2 million live births that represents, sometimes, a real challenge for paediatric surgeons. The majority of cases have complete or partial duplication of the pelvis and/or lower extremities. Thoracic heteropagus conjoined twins is a rare condition in which a grossly defective foetus (the parasite) is attached to the thorax of the main foetus (the autosite). We describe a case of a parasitic heteropagus attached at the chest wall with a rare presentation of giant exomphalos on the autosite. In this situation, the separation procedure was simple after a well-done anatomic study using X-ray, ultrasonography, magnetic resonance and echocardiogram are performed.

Key words: Asymmetric conjoined twins, heteropagus twins, incomplete conjoined twins, omphalocele, omphalopagus parasite, parasitic twins

INTRODUCTION

Heteropagus twin is an extremely rare event with an incidence of 1–2 million live births.[1] Heteropagus twin refers to a type of conjoined twin in which an incomplete smaller (parasitic) twin is attached to and dependent on an otherwise normal host twin (termed the autosite).[2]

The aetiopathogenesis of conjoined twins has yet to be determined. It is postulated that heteropagus twin occurs as a result of selective ischaemic damage *in utero*, thereby resulting in death and partial resorption of one of the twins and eventuates in an incomplete, parasitic twin attached to a fully developed twin.[3]

The parasite is totally dependent for growth on the host, usually acardiac, anencephalic, rarely contains thoracic organs, usually demonstrates lower limbs, trunk and sometimes dysplastic upper extremities with little or no movement on them, rudimental intraabdominal organs is a possible finding as well. In asymmetrical twin sets, the most common presentation is omphalopagus joined at either hypogastric or suprapubic region, whereas epigastric attachment is sparse.[4]

Being such a complex and rare occurrence, the surgical approach always poses a challenge for the surgical team, especially when facing the need for closure of a large defect. We present a case of successful surgical treatment of a parasitic heteropagus attached at the chest wall with a rare presentation of giant exomphalos on the autosite and no pleural involvement.[2]

CASE REPORT

A male infant was born at the hospital.

His birth history consisted of a normal spontaneous vaginal delivery after 40 weeks of uncomplicated gestation. He had a given weight 4400 with Apgar scores of 7 and 9 at 1 and 5 min, respectively. Birth weight was including both autosite and parasite.

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The mother was 33 years old (gravida 3, para 1). She had been submitted to ultrasonography (US), and the malformation had not been identified. No congenital abnormality had been documented in the patient's family history. No history of maternal medication use or exposure to radiation during pregnancy was reported.

The malformation consisted of a well-developed autosite, onto who's from the upper left part of the thorax, the parasite was attached above a giant omphalocele. The parasite twin was attached to the thorax at the top of the nipples. The autosite was active and appeared normal except for the omphalocele.

The parasite consisted of two hypoplastic arms and thorax, a misshapen trunk, a small abdomen, a pelvis, and two well-formed legs. Examination of the perineum showed genitalia: Small well-developed penis, and empty scrotum and no an anal opening. There was a total lack of movement in its extremities [Figure 1].

Radiologic studies, US and magnetic resonance imaging (MRI) of the autosite showed no significant anomaly. The result of echocardiography of the autosite was normal.

However, the parasite showed no musculature in the arms and legs (only skin, fat and bone), bone malformation (where the hypoplastic arms were attached).

After preparation, the newborn was operated on. Surgical separation of the twins was performed on the 7th day after birth. The parasite was attached to the host by skin, subcutaneous tissue, muscle, and fascial layers. The incision was extended cranially and reached the sternum. The omphalocele sac was excised circulary. The abdominal cavity was exposed. The vascular pedicle of the parasite arose from the pedicule intercostale of the autosite. Two kidneys, the ureters, the bladder of the parasite and segments of atresic bowel were identified easily. There were no intrathoracic organs in the parasite.

Both ends of the parasite's bowel were blind, without any connection to the autosite's gut. The autosite's intraperitoneal organs were completely normal in appearance. The autosite had a vesicular bifidity [Figure 2]. The parasite was separated by ligating the vessels, removing the amniotic sac and hypoplastic intestines without difficulty.

The thoraco-abdominal wall defect and the omphalocele were closed without difficulty [Figure 3].

The post-operative evolution had no complication. The patient was able to eat on the 7th day after surgery, and the thorax was stable with a satisfactory oximetry.

The patient was discharged from hospital 20 days post-operatively.

DISCUSSION

As a consequence of their infrequency, the heterogeneous terminology, and underreporting of abnormalities, epidemiologic data regarding heteropagus twins have been limited.[1,5,6]

Conjoined twins are mostly monochorionic-monoamniotic twins and categorised as symmetrical (diplopagus) and asymmetrical (heteropagus) forms on the basis of the sizes of twins attached at any part of their body. The exact aetiopathogenesis is unknown, but most authors consider it to be a result of an incomplete division of the embryonic disk during the first trimester.

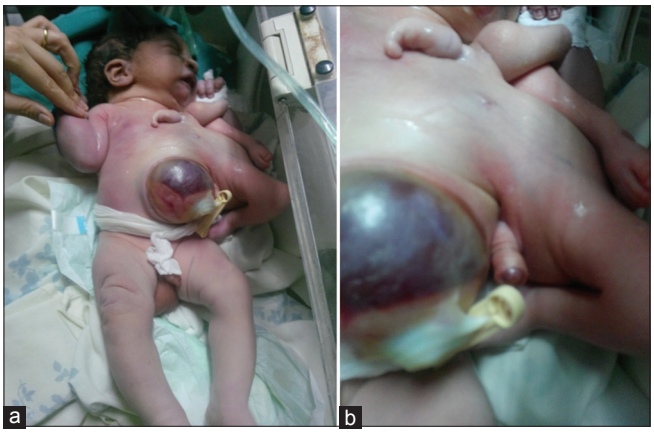

Figure 1: (a and b) Preoperative aspect of thoracopagus: The parasite is attached to the chest wall of the autosite associated with omphalocele

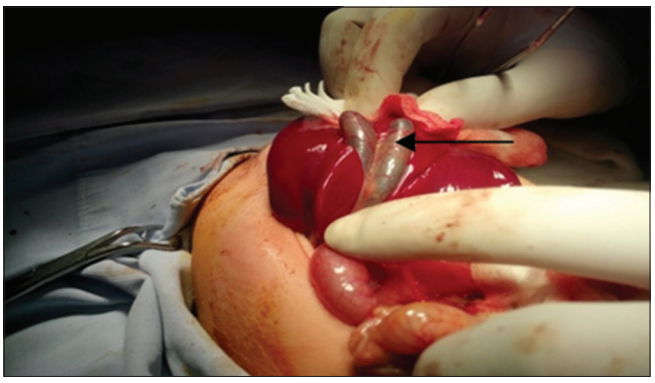

Figure 2: Pre-operative aspect of a vesicular duplicity

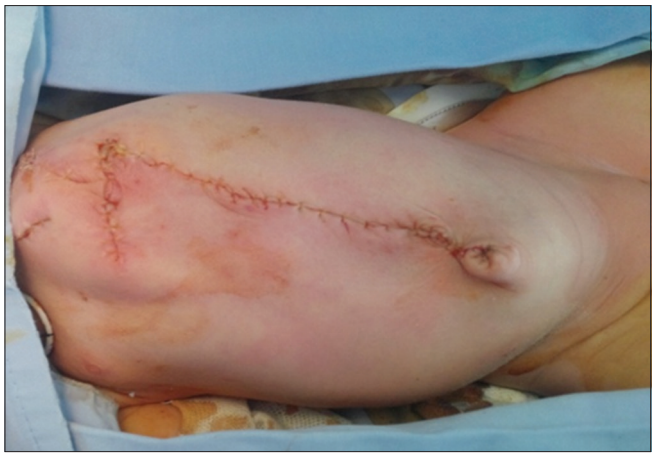

Figure 3: Post-operative aspect

In heteropagus form, a smaller incomplete parasite is fused to a complete autosite, usually in the hypogastric or suprapubic region of the host. In some extremely rare cases, the junction is above the umbilicus, which is named as epigastric heteropagus twins.[7-9]

Joined twins at the site of either abdomen or thorax result from the union at the extreme rostral aspect of the early embryonic disc, primarily involving only the septum transversum in omphalopagus, but both the septum and the cardiac primordium in thoracopagus. The latter comprises a small portion of all reported heteropagus twins because a defective heart of the autosite is unable to support the viability of both foetuses.[4,10] Albeit by the definition, thoracopagus parasiticus must involve abnormal heart (duplication of cardiac primordium in symmetrical twins), some authors have reported heteropagus parasiticus without heart involvement in the autosite and classified this as thoracopagus according to the site of attachment only.[11,12] Other authors have classified cases of asymmetric doubling as epigastric heteropagus when the parasite was connected at the chest wall because of giant omphalocele.[13,14]

The criteria for thoracopagus parasiticus in recondite cases are obscure since they include only the site of union and heart involvement.

In the latter case, the autosite was diagnosed with major congenital heart anomalies and the irrigation of the parasite was sustained by the vascular pedicle originating from the artery of the thoracic wall of the autosite, as there was no pleural cavity involvement in the communication between foetuses.

Most infants with heteropagus are male.

Conjoined twins have been classified as cephalopagus, thoracopagus, omphalopagus, ischiopagus, parapagus, craniopagus, pygopagus and Rachipagus.[15]

Embryonic death of parasitic twin leaves behind body parts vascularised by the autosite in heteropagus. Although the aetiology of embryonic death is yet unknown, it is thought to be caused by ischaemic atrophy and/or insufficient cardiac function of the parasite or vascular steal from the autosite. The parasite is usually acardiac except in a few cases and is supplied by the vessels arising from different regions of the autosite, including liver, left internal mammary artery, epigastric artery, umbilical vessels and falciform ligament.[7]

Antepartum diagnosis of parasitic twinning plays an important role in management. Outcomes are generally favourable. Therefore, heteropagus twins should be distinguished from more severe congenital malformations to avoid unnecessary termination.

At least, three examples exist in which the authors explicitly note the failure of prenatal sonographic assessment to detect heteropagus twins.[1,16-18]

Prenatal diagnosis can be established by ultrasonographic examination or MRI and is becoming increasingly frequent.[2]

Mode of delivery of heteropagus twins is only described in 34 articles from 1984 to the present. Eighteen pairs (53%) of twins were delivered vaginally and 10 (29%) by caesarean delivery.[1]

Pre-operative imaging before the separation of symmetric conjoined twins can be quite extensive. Because sharing of organs and vascular connections between autosite and parasite of heteropagus pairs tend to be less extensive, pre-operative imaging used has been more limited. Computed tomography (CT), ultrasound and MRI are the most common initial studies in all types of heteropagus twins.

In addition to CT and MRI, echocardiography is often added for thoracopagus and omphalopagus heteropagus.

Several groups have used Doppler ultrasound[17,19-21] or magnetic resonance angiography[16,22] to identify the vascular pedicle sustaining the parasite before surgical separation. Vascular connections are often determined intraoperatively. In general, regional CT or MRI as well as echocardiography are advisable pre-operative imaging modalities.

The parasites have been reported to be nourished via the vessels of falciform ligament, intercostal arteries, mesenteric arteries, epigastric arteries, left subclavian artery and brachiocephalic trunk.[4]

Omphalocele was present in approximately half of epigastric heteropagus, but major omphalocele was seen only in a few cases in thoracopagus. A common ventricle has been described in only three more reports in the reviewed literature.[4]

Options for treatment of such twins are dependent largely on the anatomy of the cardiovascular system. The use of sonography followed by MRI in evaluating the anatomy of conjoined twins is a valuable assessment sequence that plays a significant role in predicting prognosis for survival, and making decisions for treatment and follow-up.[7,23]

Separation is usually performed in a single operation.

Non-lethal complications ranged from an incisional hernia to teratoma formation at the site of retained parasite tissue.[1]

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Conflicts of interest
There are no conflicts of interest.

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