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Epigastric heteropagus twinning with dextrocardia: a case report

Ayesha Siddiqua[1*], Noor-ul-Sabah[1] and Muhammad Ramzan Bhutta[2]

Abstract

Background Heteropagus twinning is a rare birth defect in 1 in million live births. Epigastric heteropagus is the most frequent type, in which a smaller dependent (parasite twin) is attached to the body of the host (autosite twin) to the thorax and umbilicus. Several congenital anomalies have been previously described in the host twin. We describe the first case of epigastric heteropagus twinning with dextrocardia and situs inversus in which a successful surgical separation is performed.

Case presentation A 48-h-old female sex twins were presented to Bahawal Victoria Hospital. A careful anatomic study using contrast X-ray, magnetic resonance imaging, and echocardiogram showed epigastric heteropagus twinning and situs inversus in autosite twin. A successful surgical separation was performed, and the parasite twin was removed from the body of the autosite twin.

Conclusions Antepartum screening detects congenital malformations; however, several pregnancies are unsupervised in underdeveloped countries. In the case of heteropagus twinning, careful surgical separation in the absence of complications leads to the favorable survival of the autosite twin.

Keywords Heteropagus twins, Conjoined twins, Dextrocardia, Situs inversus, Epigastric twin, Omphalopagus, Parasites, Umbilicus, Pregnancy

Background

Heteropagus twinning is a type of conjoined twinning in which a partially formed twin (parasite twin) is attached and dependent on a fully developed twin (autosite twin). The literature on heteropagus twinning mainly comprises of isolated case reports. It is an extremely rare condition with an estimated incidence of 1 per 1 million live births [1]. "Epigastric" or "ompahalopagus" heteropagus twinning is the most common anatomical variant comprising 59% of the cases in which the primary area of attachment is at the level of umbilicus and thorax, without sharing the heart [1, 2]. A male predominance is seen in cases of epigastric heteropagus, and congenital heart defects in autosite are seen in half of the cases described [1, 3].

We present a case of female epigastric heteropagus twins with dextrocardia, having successful surgical outcomes. Only two cases of epigastric twinning with dextrocardia have been previously described in the medical literature.

Case presentation

A female infant weighing 4 kg was received on day 2 of life in the outpatient pediatric surgery department with heteropagus twinning. The infant was born after a full-term pregnancy, to an 18-year-old mother (G1, P1), from a consanguineous marriage. The pregnancy was unsupervised, and the infant was delivered via C-section.

The parasite was attached to the autosite at the thoracoepigastric region. The parasite consisted of a head with macrocrania, facial dysmorphia, a misshapen trunk, one

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ear, one immobile rudimentary leg, and no perineum (Fig. 1A).

The autosite was healthy and active. The physical examination revealed no abnormalities, and all neonatal reflexes were normal. The hematological and biochemical workup was also within normal limits.

The barium contrast enema (Fig. 1B) revealed mid-abdominal bowel loops predominantly in the healthy twin, and a single loop exhibited in the parasite twin. MRI showed that the brain and spine were unremarkable in the healthy twin. An abnormal corpus callosum, hypoplastic brain parenchyma, and hydrocephalus

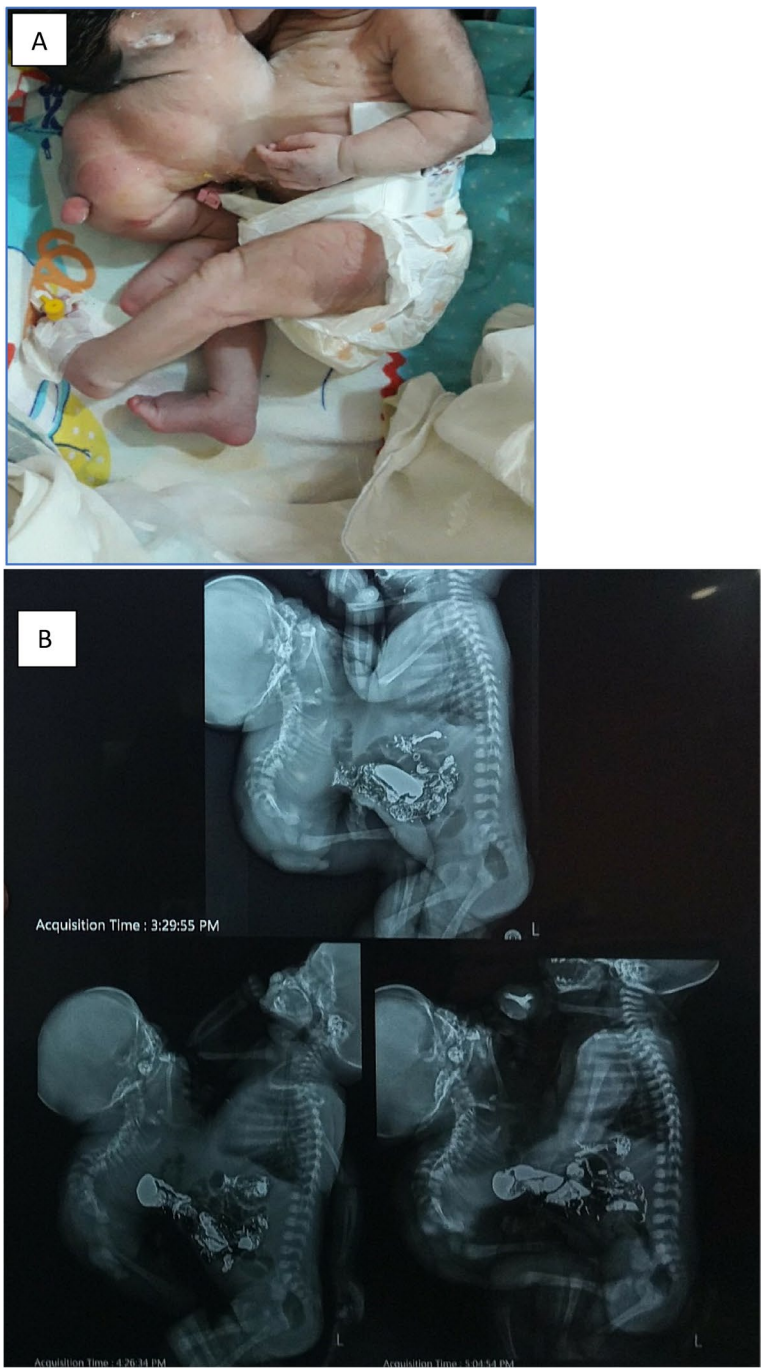

Fig. 1 **A** Preoperative view, showing parasite attached to the chest wall of the healthy baby. **B** Barium contrast enema, X-ray showing shared bowel loops

were seen in the parasite twin. The twins were fused anteriorly at the chest and abdominal cavity. A single large heart was noted in the healthy neonate. The single shared liver was present in the midline, with greater portion in the autosite twin. A single stomach was seen in the parasite twin, while majority of gut loops were in the healthy neonate. There was no connection except vessels connecting the neonates.

2-D color Doppler echocardiography showed dextrocardia with situs inversus. AV concordance was there. Atrial and ventricular septal walls were intact, and the heart showed good biventricular function.

On the 16th day of life, a comprehensive team surgery was performed, including pediatric, vascular, and plastic surgeons. The twins had anatomical connections spanning the skin layers and subcutaneous tissue. The vascular link was through the umbilical vessel. There was no bony connection between the twins. The twins did not share any connection at the chest wall. A release incision was given to separate the nonviable infant, vessels were ligated, and hemostasis was secured. The stomach and gut loops present in the abdominal wall of the parasite baby (Fig. 2) were preserved and transferred back to the healthy neonate. A reverse technique was employed to close the incision on healthy neonate.

The postoperative recovery was uneventful. The baby recovered and was discharged on the 8th postoperative day. A follow-up was done at 3 months over a phone call, according to which the baby was healthy and gaining weight. A later follow-up conducted revealed that the baby passed away due to chronic constipation at 6 months of life.

Conclusions

Conjoined twinning is a rare congenital anomaly, and "asymmetric" or "heteropagus" conjoined twinning is even more infrequent. The exact cause is unknown, but there are different theories to explain the embryological origin of these cases.

The "fission" theory suggests that incomplete fission of blastocyst inner cell mass during the primitive streak stage leads to the development of 2 centers of axial growth instead of one. The "fusion" theory suggests coalescing two distinct inner cell masses at a later stage. Asymmetry of parasite twins is thought to occur due to vascular compromise of parasite twins leading to dependence on autosite by collaterals [1]. Vascular connections arise by the liver, internal mammary artery, umbilical artery, epigastric vessels, and falciform ligament [3]. Selective ischemia leads to the development of various abnormal organs and limbs in the parasite twin [4].

Preoperative imaging is usually done by ultrasound, CT, and MRI. Echocardiography is recommended in thoracopagus and omphalopagus twins to assess heart defects [1, 5]. Angiography is usually not done as vascular connections are determined intraoperatively [5, 6].

The most common congenital cardiac defect seen is ventral septal defects in one-third of cases, and others include atrial septal defects, patent ductus arteriosus, coarctation of the aorta, and single ventricle [1]. Dextrocardia with right axis deviation and situs inversus in epigastric heteropagus was first described in prenatal diagnosis through three-dimensional ultrasound by MacKenzie et al. [7]. The pregnancy was medically terminated owing to anomalous fetuses. The second case is described by Mittal et al., where the autosite had dextrocardia with situs inversus and a single ventricle [4]. Both

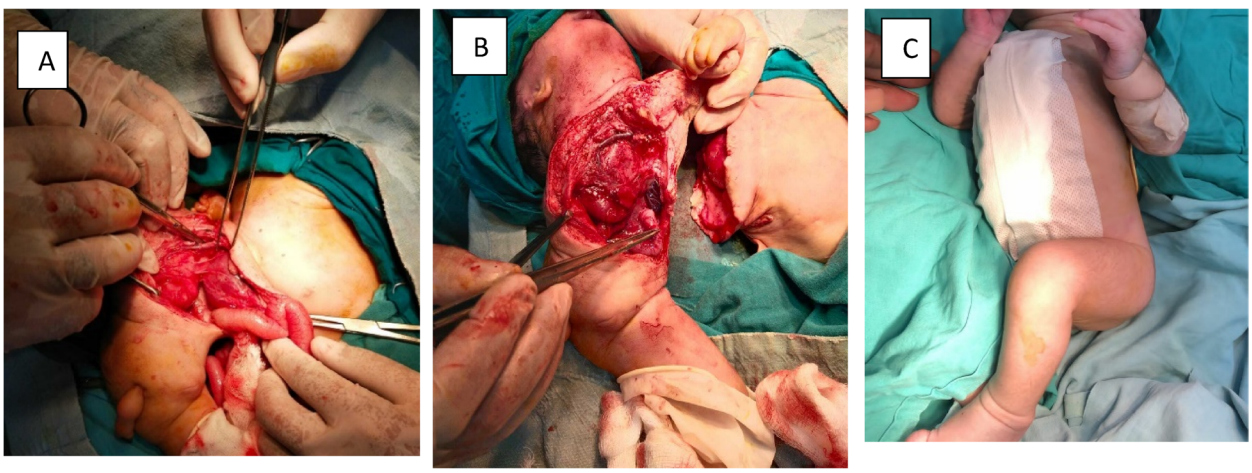

Fig. 2 **A** and **B** Intraoperative details showing shared gut loops between twins. **C** Postoperative healthy twin

aorta and pulmonary artery arose from the same ventricle. The child expired on 4th day of life owing to respiratory distress.

Direct communication between autosite and parasite gastrointestinal and hepatobiliary structures is rare; however, conjoined livers appear to be common [3]. Barium X-ray helped determine the sharing of gut loops between the twins in this case.

Although several complications may arise, postsurgical survival of heteropagus twins is better than conjoined twins due to less extensive visceral and vascular connections in autosite and parasite. Cardiorespiratory failure associated with congenital cardiac defects in autosite is the most commonly cited cause of death [1]. Antepartum assessment is crucial to determine the cases of more severe malformations, such as a single ventricle to avoid needless terminations of heteropagus twins [1].

This case is the first to our knowledge of epigastric heteropagus twins with dextrocardia having successful surgical outcomes. Positive outcomes for the autosite twin can be achieved through meticulous aseptic surgical interventions in instances of nonlethal congenital heart defects observed in epigastric heteropagus twins.

Abbreviations

MRI Magnetic resonance imaging
CT Computerized tomography
AV Atrioventricular

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Availability of data and materials
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Declarations

Ethics approval and consent to participate
Written informed consent is obtained from parents for publishing. The consent form is available as supplementary file. This report does not contain any personal information that could lead to the identification of the patient.

Competing interests
The authors declare that they have no competing interests.

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