Epigastric Heteropagus Twins : A Report of Four Cases

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Abstract

Epigastric heteropagus twins (EHT) is a rare form of conjoined twins. It refers to unequal and asymmetric conjoined twins in which the dependent part (parasite) is attached to the epigastrium of the dominant part (autosite). The authors herein report four cases of EHT. Omphalocele was present in 3 patients. Surgical excision of the parasite and repair of the abdominal wall defect were successful in three cases. Three infants had associated cardiac anomalies and one of them died during surgery from cardiopulmonary failure.

Key word : Conjoined Twins, Epigastric Heteropagus, Parasitic Twinning

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Conjoined twins are an extremely rare congenital anomaly, occurring in about 1 in 50,000 to 100,000 livebirths[1]. This entity is classified as symmetrical or asymmetrical. The asymmetrical form is known as heteropagus. The dependent part, so-called parasite is attached in a nonduplicated fashion to any portion of the body, or even within the body as a fetus-in-fetu[2]. The parasite is smaller than the dominant part (autosite). Epigastric heteropagus refers to conjoined twins in which the parasite is attached to the epigastrium of the autosite. The authors, herein, report four cases of epigastric heteropagus twin treated at our institution from 1986 to 2002.

CASE REPORTS

Case 1

A male baby, weighing 2,750 g, was born by vaginal delivery at Rajavithi Hospital on 5 January,

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1986. Antenatal course was uneventful and there was no history of prenatal ultrasound examination. No history of drug exposure was obtained. The baby had a parasitic leg connected at the epigastrium craniad to an omphalocele of 2 cm in diameter. This leg could not be moved passively. Surgical removal was performed at one day of age. The parasitic leg was resected without difficulty and the abdominal wall defect was closed completely. Post-operative course was uneventful. The baby did well with normal development at the six-month follow-up.

Case 2

A male baby, weighing 2,710 g, was born by vaginal delivery at Phra Phutthabat Hospital, Saraburi province on 24 August, 1989. Antenatal course was uneventful and there was no history of prenatal ultrasound examination. No history of drug exposure was obtained. The baby had a parasitic appendage of a lower trunk at the epigastrium craniad to an omphalocele 6 cm in diameter. Echocardiography of the autosite showed tetralogy of Fallot (TOF) and patent ductus arteriosus (PDA). Surgical removal was performed at one day of age. The parasite was attached to the autosite by skin, subcutaneous tissue, muscle and fascial layers. Intraabdominal organs of the autosite were completely normal in appearance. After excision of the parasite, the abdominal wall defect and the omphalocele were closed without difficulty. Post-operative course was satisfactory. Correction of the TOF and ligation of the PDA were performed at 2 years of age. The baby did well and had developed normally at the three-year follow-up.

Case 3

A male infant, weighing 3,400 g, was born by normal vaginal delivery at Lop Buri Hospital on 24 October, 1997. Antenatal course was uneventful and there was no history of prenatal ultrasound examination. His mother had no history of any drug use during the early period of pregnancy. The parasite was attached to the epigastrium of the autosite and consisted of a lower trunk, a pelvic girdle, lower extremities, and a rudimentary structure resembling an upper trunk with one arm. This monster was anencephalic. (Fig. 1) There was no bony connection between the parasite and the autosite. Echocardiography of the autosite showed a single ventricle and severe congenital cardiac anomalies. Surgical removal of the parasite was performed at tens day of age. The parasite was attached to the autosite by skin, subcutaneous tissue, muscle and fascial layers. A peritoneal communication and conjoined bladder were also noted. The patient succumbed to cardiopulmonary failure

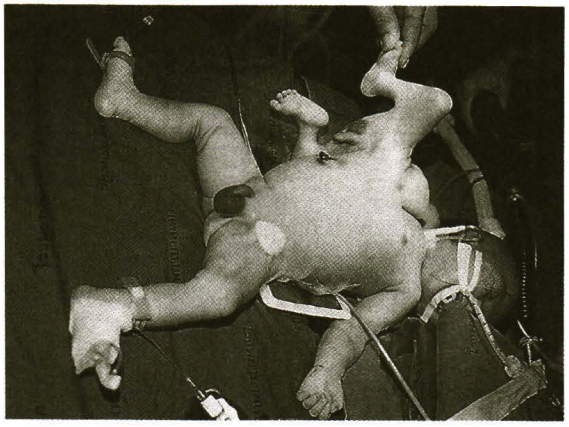

Fig. 1. The parasitic twin attached to the host's epigastrium. The parasite had a lower truck, a pelvic girdle, and lower extremities. (Case 3)

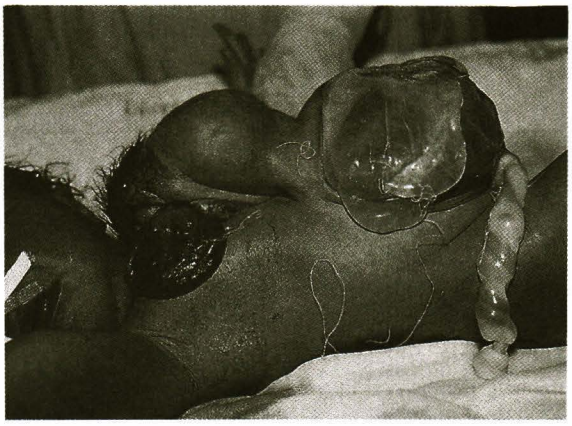

Fig. 2. The parasitic twin was attached to the host's epigastriu craniad to an omplalocele. The parasite had a rudimentary structure resembling an upper trunk with two rudimentary upper limbs. (Case 4)

Table 1. Clinical data of heteropagus patients.

Case	Year	Sex	Omphalocele	Bowel connection	Cardiac defect	Parasitic twin
1.	1986	M	+	-	-	One well developed leg
2.	1989	M	+	-	TOF, PDA	Rudimentary lower trunk
3.	1997	M	-	-	Single ventricle	Well developed lower limbs and pelvis, rudimentary trunk and upper limbs
4.	2002	F	+	+	PDA	Rudimentary upper trunk and limbs well developed liver, stomach and gall bladder, rudimentary intestine

Note : TOF = Tetralogy of Fallot, PDA = Patent ductus arteriosus

from his severe congenital cardiac anomalies during surgery.

Case 4

A female infant 3,500 g in birth weight was delivered by cesarean section for fetal distress at Rajavithi Hospital on 7 October, 2002. The mother was 34 years old (gravida 2, para 1). Prenatal ultrasonography was done at 22 weeks' gestation and an omphalocele was detected. No history of maternal drug use was obtained and her older sibling had no congenital anomalies. The parasite was attached to the epigastrium craniad to a 7 cm in diameter omphalocele of the autosite and consisted of a rudimentary structure resembling an upper trunk with two rudimentary upper limbs. (Fig. 2) Echocardiography of the autosite showed a PDA. Surgical removal was performed on the second day of admission. The parasite was attached to the host by skin, subcutaneous tissue, muscle and fascial layers. The peritoneal cavity of the parasite was connected to that of the autosite with fusion of two sets of livers. There were two stomachs and two gall bladders. A segment of the small intestine of the parasite was connected to the jejunum of the autosite. The parasitic bowel was resected but the liver of the parasite was left in the autosite. The abdominal wall defect and the omphalocele were closed without difficulty. Post-operative course was uneventful. The PDA was treated conservatively and had spontaneous closure at six months of age. The baby was healthy at six month follow-up.

DISCUSSION

The pathogenesis of conjoined twins is still uncertain. It is believed to be due to incomplete cleavage of the inner cell mass of the blastocyte at approximately 2 weeks of gestation[3-6]. It is accepted that conjoined twins derive from a single blastocyte that represent a form of monozygotic twinning. The finding of the same sex in both components of conjoined twins supports this theory[7]. Conjoined twins are classified into symmetrical and asymmetrical forms. The symmetrical form consists of thoracopagus, omphalopagus, pygopagus, ischiopagus and craniopagus[8]. The asymmetrical form, namely heteropagus, develops when one component is better placed so that it monopolizes the placental blood flow to the detriment of the other member (parasite)[9], and ischemic atrophy of one part of a monozygotic conjoined twins in early gestational life has been proposed as the pathogenesis of heteropagus[2,7].

There is a predominant incidence of male over female with a ratio of 5 : 1 in heteropagus. This is in contrast to female predominace encountered in symmetrical conjoined twins[10]. In the present study the male to female ratio was 3 : 1.

Nasta et al[11] reported a case with a connection of the parasite's bowel with the Meckel's diverticulum of the host. No bony connection was seen between host and parasite. An omphalocele was often present[9]. Three of our four patients had an associated omphalocele. The vascular supply to the parasite may arise from the liver, left internal mammary, epigastric, umbilical, falciform ligament, left subclavian or and local systemic arteries[9,12]. In all of the presented cases, the vascular supply to the parasite arose from the local systemic artery.

Cardiac anomalies are common in autosites and should be evaluated by echocardiography[7,11].

Three of our patients had associated cardiac anomalies : TOF and/or PDA in two cases, and a single ventricle in the other. (Table 1)

Surgical treatment with excision of the parasite was satisfactory. The abdominal wall defects were closed without post-operative ventral hernia. In the authors' experience, survival of the patient was adversely influenced by associated cardiac anomalies. Without a severe cardiac condition, the result was excellent.

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แฝดติดกันไม่สมบูรณ์ แบบเฮเตโรเพกัส : รายงานผู้ป่วย 4 ราย

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ภาวะ epigastric heteropagus twins (EHT) เป็นความผิดปกติรูปหนึ่งของแฝดติดกัน (conjoined twins) โดยมีความไม่สมดุลย์ของ conjoined twins ส่วนที่ไม่สมบูรณ์ (parasite) จะเกาะติดอยู่กับตัวเด็กที่สมบูรณ์ (autosite) บริเวณลิ้นปี่ คณะผู้รายงานได้รายงานผู้ป่วยเด็ก 4 ราย ที่มีภาวะ EHT โดยที่ผู้ป่วย 3 รายมี EHT ร่วมกับความผิดปกติของผนังหน้าท้อง (omphalocele) และอีก 1 รายมีภาวะ EHT โดยไม่มีความผิดปกติของผนังหน้าท้อง ผู้ป่วย 3 รายได้รับการผ่าตัด โดยตัดเอาส่วนที่ไม่สมบูรณ์ออก การรักษาได้ผลเป็นที่น่าพอใจ ไม่พบว่ามีภาวะแทรกซ้อนหลังการผ่าตัด ความผิดปกติแต่กำเนิดของหัวใจเป็นความผิดปกติร่วมที่พบได้บ่อย โดยพบในผู้ป่วย 3 ราย มี 1 รายเสียชีวิตขณะผ่าตัดจากภาวะหัวใจล้มเหลว

คำสำคัญ : แฝดติดกัน, แฝดติดกันไม่สมบูรณ์ แบบเฮเตโรเพกัส, แฝดไม่สมบูรณ์

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