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Epigastric heteropagus associated with an omphalocele and double outlet right ventricle

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ABSTRACT

Incomplete or asymmetrical conjoined twins are extremely rare congenital anomalies. We report a case of epigastric heteropagus associated with an omphalocele and double outlet right ventricle. The cystic legion of the epigastrium was detected in our patient by an ultrasound scan at 28 weeks' gestation. He was born at 37 weeks' gestation by scheduled caesarean section. A parasite with an incomplete head and lower limb was attached to the epigastrium of the autosite. Surgical separation of the parasite and silo placement for an omphalocele was successfully performed on the 4th day of life. He underwent secondary surgical closure of the omphalocele on the 10th day. For treatment of the cardiac anomaly, he underwent an operation of Blalock–Taussig shunt because of pulmonary artery stenosis at the age of 3 months and correction of double outlet right ventricle at the age of 10 months. At the 20-month follow-up, he was alive and showed a normal growth pattern.

Epigastric heteropagus (EH) refers to asymmetrical conjoined twins in whom an incomplete parasite is attached to the epigastrium of the autosite. Because of the rare incidence of EH, its manifestation and therapeutic outcomes remain unclear. Anatomical variability may jeopardize a successful surgical separation. In this case report, we describe antenatal ultrasonography and magnetic resonance imaging (MRI) for diagnosis. Furthermore, the vascular supply from the autosite to the parasite was assessed after birth by computed tomography (CT) angiography.

1. Case report

A 31-year-old healthy woman, gravida 5, para 2, presented at 28 weeks' gestation for ultrasound obstetric care. There was no history of twinning, family congenital anomalies, maternal illness, or drug intake during pregnancy. The antenatal course was uneventful. Ultrasonography revealed an omphalocele and a cystic lesion at the epigastrium. Therefore, she was referred to our gynecological department. Furthermore, the congenital cardiac anomaly of double outlet right ventricle (DORV) was identified. MRI was performed for a detailed examination at 30 weeks' gestation. The cystic lesion measured 57 × 44 × 43 mm and it extended to 73 × 56 × 49 mm at 36 weeks' gestation (Fig. 1). One rudimentary limb was also identified. Therefore, we suspected incomplete conjoined twins. A scheduled caesarean section was performed at 37 weeks' gestation. Birth weight was 2316 g. A parasite was observed consisting of a rudimentary head with hear and a lower limb was attached above an omphalocele to the epigastrium of the autosite (Fig. 2). He was immediately intubated after birth because of weakness of breathing, muscle tone reflexes, and systemic cyanosis (Apgar score of 5). The omphalocele was temporarily covered by a waterproof film. CT angiography showed a vascular supply from the left intrathoracic artery of the autosite to the parasite (Fig. 3). No open link between the autosite and parasite was observed. Herniated organs from the omphalocele were part of the liver and small bowel. Surgical separation of the parasite was performed on the 4th day of life. A circular incision was made at the union of the bodies. The pelvic bone of the parasite was connected to the xiphoid of the autosite by a tract of cartilage. The left intrathoracic artery of the autosite was identified in the left side of this connection. The parasite was acardius acormus and it weighed 216 g. After the separation, the body wall defect at the right side of the rectus abdominis muscle was covered by the opposite side of the rectus abdominis muscle. Simple closure of the skin was performed. Silo placement for the omphalocele using the Alexis® wound retractor (Applied Medical Resources Corporation, CA) was also performed (Fig. 4). On the 10th day, secondary surgical closure of the omphalocele was performed. He was extubated 2 weeks after the secondary operation, but

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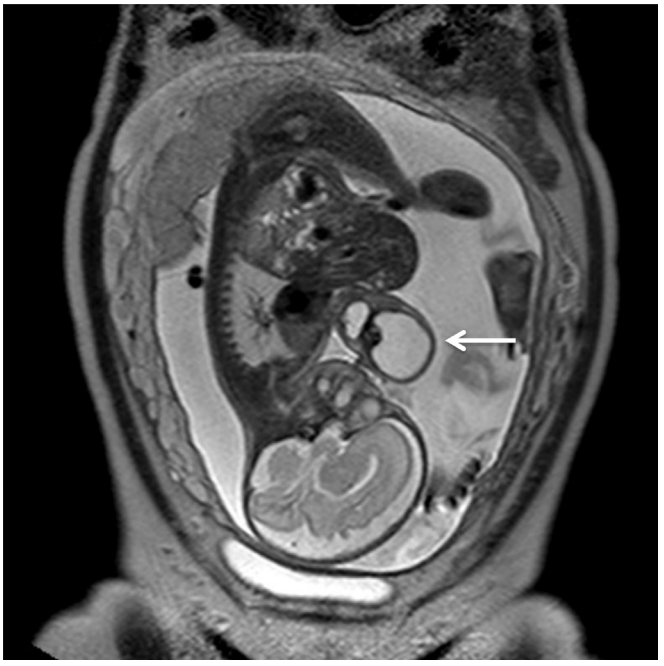

Fig. 1. MRI shows a cystic lesion (arrow) at the epigastrium at 30 weeks' gestation.

oxygen saturation of the peripheral artery was approximately 90% due to progression of pulmonary artery stenosis. At 3 months of age, he underwent an operation of Blalock–Taussig shunt. Subsequently, he underwent correction of DORV at 10 months of age. At the 20-month follow-up, he was alive and shows a normal growth pattern. Plastic surgery for protrusion of the xiphoid process and mild ventral hernia is under consideration.

2. Discussion

Twins who share their vital organs and body parts are referred to as conjoined twins. The precise etiology of conjoined twins is unknown, but formation of these anomalies is currently believed to be related to incomplete cleavage of the embryo at approximately 2 weeks' gestation [1]. Conjoined twins also represent a form of monozygotic twins [2]. Conjoined twins are classified by the site of union and their symmetry/asymmetry. The incidence of conjoined twins is considered to be 1 in 50,000 to 100,000 births [3] and heteropagus constitutes only 1–2% of all conjoined twins [4]. Furthermore, the majority of heteropagus cases have been cases of dypigus (caudal duplication), representing attachment of an accessory pelvis and one or more rudimentary lower limbs in the hypogastric or perineal region of the autosite [1].

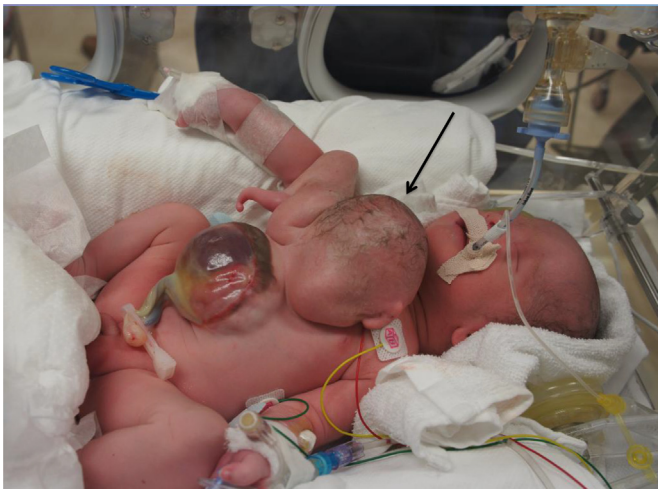

Fig. 2. A parasite consisting of a rudimentary head (arrow) and a lower limb was attached above the omphalocele to the epigastrium of the autosite.

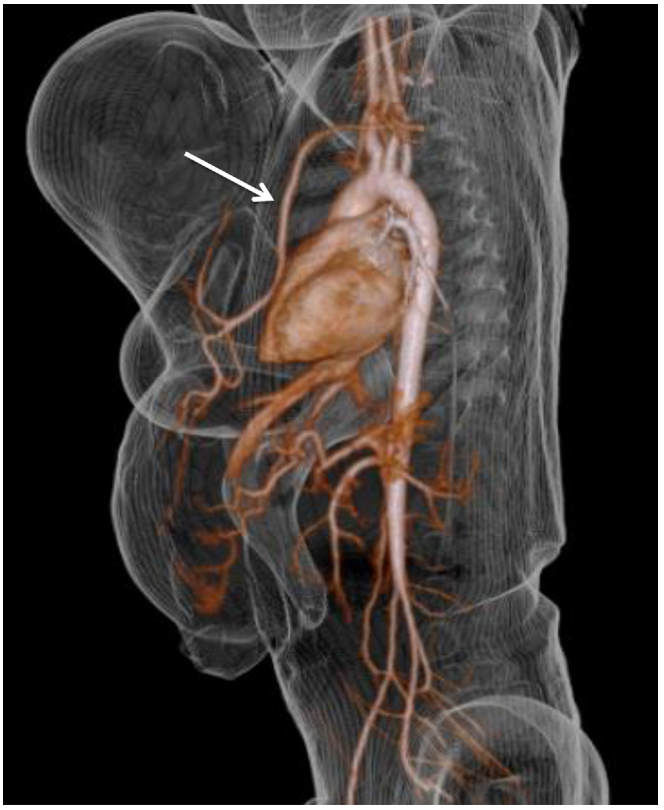

Fig. 3. CT angiography shows a vascular supply from the left intrathoracic artery (arrow) of the autosite to the parasite.

Karnac et al. [5] made several important observations on EH as follows: (i) there is a predominance of males, which is in contrast to female predominance that is encountered in general conjoined twins, (ii) a hereditary tendency was found in three cases [6–8], (iii) the host's and parasite's bowel does not appear to have any connection, (iv) omphalocele is the most common associated abnormality, (v) there is no sternal attachment of the rectus muscles in some cases, and (vi) a local systemic artery of the host supplies blood to the parasite. A bowel connection between the host and parasite was found in three cases [9–11]. Hager et al. [12] emphasized that options for treatment of EH largely depend on the pathological anatomy of the cardiovascular system. Another important point is the absence of innervation and skeletal muscle in the parasite's extremities. To the best of our knowledge, 32 cases of EH have been reported in the English literature from 1946 to 2013. Thirty-three cases, including our case, were reviewed (Table 1) [3,4,6–29]. Twenty-seven cases were male and six were female. Omphalocele coexisted in 16 cases, and cardiac disorder in 11 cases. Vascular supply to the parasite was reported as follows: falciform ligament, left internal mammary artery, left subclavian artery, left intrathoracic artery, and epigastric artery. Results of surgical separation were mostly successful, except in three patients [11,14,16] who died a few days after the operation because of circulatory failure, pericardial tamponade, respiratory failure, and sepsis. Prenatal diagnosis was made in three cases, including our case. These