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CASE REPORT

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Epigastric heteropagus twins – a report of four cases

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Abstract Asymmetrical conjoined twinning is an extremely rare congenital anomaly constituting only 1%–2% of all conjoined twins. In epigastric heteropagus twins (EHT), the dependent portion (parasite) is smaller than the host (autosite). The embryopathy is related to incomplete cleavage of the embryo at 2 weeks of gestation. However, some form of ischaemic insult in early gestation leads to selective atrophy of the cranial part of one of the monozygous twins. We report our experience with four cases of EHT and a review of the literature with a discussion of possible embryopathy.

Keywords Epigastric heteropagus · Parasitic twinning

Introduction

Parasitic twinning is an extremely rare congenital anomaly that occurs in 1 in 50,000 to 100,000 live births [8]. Although it appears grotesque, proper evaluation leads to successful separation of the autosite from the parasite. To our knowledge, there are only 12 similar cases reported in the English literature [1–12]. It is interesting to note that 4 of these case reports have been from India. We report four more cases, managed successfully at our institution.

Materials and methods

During the period 1991–2000, we encountered four cases of epigastric heteropagus (EHT). The age at presentation, clinical details, associated anomalies, and findings of angiographic and the DNA studies are shown in Table 1. All the cases were successfully separated.

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Discussion

Of the 12 cases of EHT reported so far in the English literature, four have been from India. Four more cases of EHT are added, and to the best of the authors' knowledge this is the largest single series reported from any center. Whether it is more common in this part of the world remains a speculation. EHT are predominantly males, unlike conjoined twins, which are predominantly females. It is presumed that the female zygote once formed is more likely to be viable compared to a male zygote. All our cases were males. In cases 1 and 2 the DNA study proved a male genotype in both the autosite and the parasite.

An omphalocoele was present in 7 of the 12 (60%) cases reported [5]. Two of our cases also had an associated omphalocoele. Unlike conjoined twins, EHT do not share bowel or other organs. Nasta et al. reported a case with a connection of the parasite's bowel with a Meckel's diverticulum of the autosite [7]. There are also no bony connections between the parasite and the autosite. However, in our case 2 there was a hinge joint between the upper limb bones of the parasite and the xiphisternum of the autosite.

The vascular supply to the parasite has been reported to arise from the liver, left internal mammary artery, epigastric vessel, umbilical vessels, and falciform ligament [5]. Angiography performed in two of our patients showed the blood supply to the parasite arising from the epigastric artery in case 1 and the left subclavian artery in case 2 (Fig. 1). EHT usually derive their blood supply from a local systemic artery; the subclavian artery has not as yet been mentioned in the literature. Angiography delineates the precise anatomy of the vascular connections and may help in not only better understanding the embryopathy of heteropagus twinning, but in planning surgical separation. DNA studies (cases 1, 2) showed a similar chromosomal makeup of the parasite and autosite, emphasising the monozygotic origin of the twins.

Table 1 Summary of the epigastric heteropagus twins (present series) (*VSD* Ventriculoseptal defect; *PDA* patent ductus arteriosus; *ASD* atrial septal defect; *A* artery)

Case no.	Age, sex	Year	Omphalocoele	Bony connection	Bowel connection	Cardiac defects	Vascular connection (angiography)	DNA study
1	1/12 M	2000	+	–	–	–	Epigastric A.	Monozygotic
2	2/12 M	1998	–	+	–	VSD + PDA	Left Subclavian A.	Monozygotic
3	15/365 M	1994	–	–	–	ASD	Not done	Not done
4	3/12 M	1991	+	–	–	–	Not done	Not done

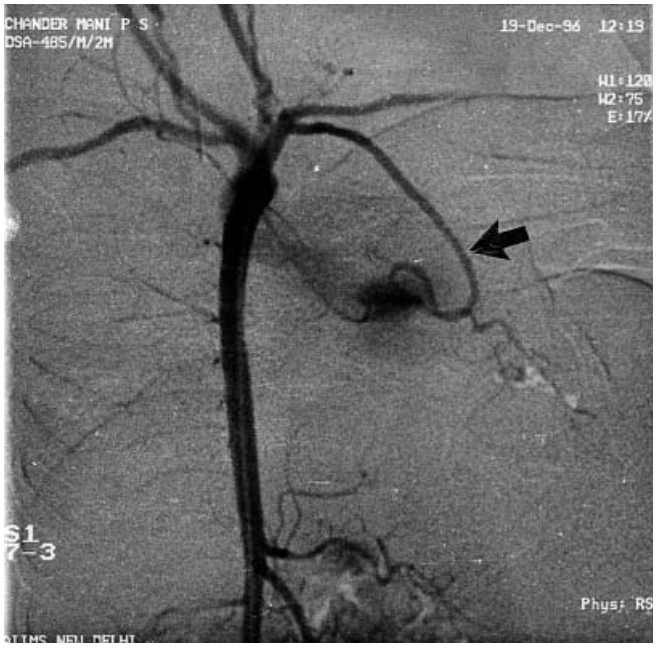

Fig. 1 Angiogram showing host left subclavian artery supplying parasite aorta (*arrow*)

Associated cardiac anomalies are common in a majority of the autosites, and should be evaluated by echocardiography [4, 7]. Two of our patients had an associated ventriculoseptal defect and a patent ductus arteriosus in one and an atrial septal defect in the other. They are on regular follow-up in paediatric cardiology for further management.

The exact aetiology of heteropagus twinning is still uncertain. Incomplete cleavage of the embryo at about the 2nd week of gestation is considered the most probable cause [13]. An ischaemic insult during early gestation may be the cause of selective atrophy of the cranial part of one of the conjoined twins. The factors that cause this selective atrophy are not discernible, however, this selective atrophy of the brain, heart, and lungs might be explained by oversensitivity of these tissues to ischaemia.

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