Heteropagus Twins—A Tale of Two Cases

Sukanta Das · Dipak Ghosh · Siddhartha Biswas · Sarbani Chattopadhyay ·
Nilanjan Mitra · Sourav Chakravartty · MD. Zahid Hossain · Veena Kumari

Received: 1 March 2009 / Accepted: 5 July 2009 / Published online: 26 January 2011
© Association of Surgeons of India 2011

Abstract Heteropagus twinning is a rare occurrence. Parasitic and asymmetric conjoined twins are rarer anomalies of monochorionic monoamniotic twins; which consist of an incomplete twin attached to the fully developed body of the co-twin. We present here two such cases of Heteropagus twinning.

Keywords Heteropagus · Twins · Omphalocele

Introduction

The term heteropagus twin was given by Potter and Craig [1]. The term Parasitic united twins was coined by Willis [2]. Heteropagus twinning is an extremely rare phenomenon. We present here two such cases that were successfully treated.

Case Report I

A full-term 2.1 kg male baby presented to the Paediatric Surgery OPD after being referred from a district hospital. He was the first child of a 23 year old mother. The

pregnancy was uneventful & the baby was delivered normally.

On the anterior abdominal wall of the child there was a spherical mass covered with normal skin. It had one well developed limb on its left side. There were two distinctly developed auricles, hairs resembling scalp hair and a pit below the hairs with small mucosal protrusions on its cephalic part resembling a face like structure (Fig. 1). There were also three small nodular structures on its surface resembling rudimentary limbs. The entire mass had a variegated consistency with feeling of bony structures as well as a soft cystic consistency. The patient also had an omphalocele which was separated from the mass by a normal bridge of skin.

Haematology & biochemical work-ups were within normal limits. Straight x-ray lateral view revealed vertebrae and bones of two limbs.

Fig. 1 The Heteropagus twin is attached to the anterior abdominal wall

S. Biswas (✉) · S. Chattopadhyay
Nil Ratan Sircar Medical College & Hospital,
138 A.J.C. Bose Road,
Kolkata 700014, India
e-mail: drjoy26@yahoo.co.in

S. Das · D. Ghosh · N. Mitra · S. Chakravartty · M. Z. Hossain ·
V. Kumari
Medical College & Hospital,
88, College Street,
Kolkata 700073, India

Fig. 2 Barium meal follow through showing no communication of the gut with the parasitic twin

Barium meal follow-through (Fig. 2) showed a normal study without any extension of the gastrointestinal tract into the swelling. USG examination showed liver being extended into the swelling and presence of cystic areas. No other organs like kidney or heart could be detected. C.T.Scan also revealed the same findings. Echocardiogram and E.C.G. of the child were also within normal limits.

The child was operated at 2 months of age. On exploration a portion of the liver was found extending into the swelling corroborating with the ultrasound findings. Besides this; a cystic tubular intestine like structure was found which was completely separated

Fig. 3 The resected specimen of the Heteropagus twin showing several viscera in it - see text

Fig. 4 The cystic tubular structure in the heteropagus twin has been cut opened to reveal a gut like structure with jejunal characteristics

from the intestines of the fully developed co-twin. The vascular connections of the parasite came from the anterior abdominal wall. The entire swelling was excised en-bloc after excising the protruded portion of the liver (Fig. 3)

The cystic tubular structure was cut opened and it revealed green bilious fluid (Fig. 4). Normal jejunal mucosa was found on histopathological examination. The other histopathological findings included the excised liver tissue with normal hepatic architecture; cartilage & fatty tissues from the ear, tooth anlage, salivary glands, squamous epithelium and cartilage from the structure resembling the face. There were also small cystic structures containing mucus which showed ciliated columnar epithelium and cartilage indicating a developing lung.

Fig. 5 The Post operative picture of the baby after excision of the parasitic twin

Fig. 6 The heteropagus twin is attached to the left side of chest & anterior abdominal wall showing well developed genitalia

No renal, cardiac and splenic tissues could be found in the parasite. No embryogenic tissues could also be found Fig. 5 shows the post operative picture of the baby after excision of the parasitic twin.

Case Report II

A 10 day old full term male baby with a heteropagus twin attached to its left chest & abdominal wall, weighing 2.8 kg presented to our OPD. The parasite had a pelvis with two

Fig. 7 C.T.Scan of the baby with catheter in the urethra of the heteropagus twin

Fig. 8 CECTscan showing a kidney like structure at the junction of the baby & the Heteropagus twin

well developed limbs, an empty scrotum with no testis & a penis (Fig. 6). There was no anal orifice in the parasite.

Haematology & biochemical work-ups were within normal limits. Straight x-ray revealed two well formed limbs, and a pelvis. C.T.Scan done along with an ascending urethrogram through a catheter inserted through the urethral orifice of the parasite (Fig. 7) revealed a rudimentary bladder and an accessory kidney in the parasitic part (Fig. 8). Besides this a gut like structure was also seen on C.T. Scan in the parasitic part. ECG & Echocardiography revealed no cardiac anomalies.

The parasite was separated successfully after 6 months. On exploration the excised part contained a rudimentary bladder and an accessory small kidney, ureter and duplication of the large gut with appendix along with

Fig. 9 Completely resected specimen showing duplicated large gut like structure & the accessory kidney

Fig. 10 The post operative picture of the baby after separation from its parasitic twin

the two limbs & external genitalia (Fig. 9). No testis like structure was seen in the excised part. Figure 10 shows the post operative picture of the baby after separation from its parasitic twin.

The excised part which was sent for histopathological examination confirmed the rudimentary structure to be a kidney and also the presence of bladder and ureter.

Discussion

There is considerable debate over the development of conjoined twins. One theory proposes that conjoined twins develop from a single fertilised ovum and result from failure of division of the embryonic disc until after day thirteen of conception [3].

Heteropagus twinning can also be explained by the concept of monozygotic duplication. One twin is superior & the disadvantaged twin is reduced in size and can survive only if it is able to draw its sustenance from the superior one by anastomosis with the chorionic circula-

tion. This asymmetrically broken up monozygotic twin can remain attached resulting in heteropagus twinning (ectoparasitic twinning); or it may be inside the body of the superior twin resulting in Fetus-in-Fetu(endoparasitic twinning).

According to some, this results from ischaemic atrophy of the body of the monozygotic conjoined twins at an early gestational age leading to selective atrophy of the cranial part of one of the monozygotic twins [4] as seen in our case two.

According to some others, the circulation in the deformed twin is reversed i.e. deoxygenated blood by the umbilical artery of the normal This causes great malformations producing acephalic, paracephalic and amorphous subtypes of these twins [5].

There can be associated congenital heart disease and the baby can be stillborn; but no congenital heart disease was present in our two cases.

As in our case one; definite formation of bone and cartilage of structures of the oral cavity were seen and possibly also a developing lung was present which is very rare amongst the cases reported in literature so far about heteropagus twinning. The possibility of teratoma in case one was ruled out because of absence of embryonic tissues [6].

References

1. Potter EL, Craig JM (1975) Pathology of foetus and newborn, 3rd edn. Year Book, Chicago, pp 220–237
2. Willis RA (1962) Borderland of embryology and pathology. Butterworth, Austin
3. Gul A, Aslan H, Ceylan Y (2004) Prenatal diagnosis of pygopagus tetrapus parasitic twin: case report. BMC Pregnancy Childbirth 4:13
4. Raynal P, Petit T, Ravasse P, Herlicoviez M (2001) Un cas rare de jumeaux heteropages epigastriques. J Gynecol Obstet Biol Reprod 30:65–69
5. Biswas SK, Gangopadhyay AN, Bhatia BD, Bandopadhyay D, Khanna S (1992) An unusual case of Heteropagus Twinning. Journal of Pediatric Surgery 27(1):96–97
6. Willis RA (1962) The borderland of embryology and pathology. London, England Butterworth, pp 135–147, 458–462