Case Report

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Parasitic twin with major cardiac defect: a case report

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ABSTRACT

Parasitic twins are an extremely rare form of asymmetrical conjoined monochorial monoamniotic twins where one of them has a mostly intact body that is able to survive and which is referred to as 'autosite', while the counterpart, referred to as 'parasite', is only rudimentarily developed being physically attached to and nourished by the other twin. Our case is a baby boy with Single Ventricle Heart defect with a thoraco-abdominal mass (epigastric heteropagus twin) attached to the anterior abdominal wall near the umbilicus with minimal visceral sharing. The twins had two external genitalia both in host and parasite micturating separately. After high risk surgery the parasite could be separated completely from the host and postoperative recovery was uneventful.

Keywords: Double genitalia, Epigastric heteropagus, Parasitic Twin, Single Ventricle defect

INTRODUCTION

Parasitic twinning is a rare malformation characterised by a twin (the 'Parasite') living in or on another host twin called the 'Autosite'. It refers to the type of conjoined twin in which the 'Parasite' is incompletely formed twin, usually nonviable but survives in the host twin (the 'Autosite') which is fully developed.¹⁻⁴ The parasite has grossly recognizable fetal parts and is usually attached to the autosite by a soft tissue pedicle containing large blood vessels.¹⁻³ The site of attachment could be one of eight (thoracopagus, omphalopagus, craniopagus, cephalopagus, parapagus, ischiopagus, pyopagus or rachipagus).

Conjoined twins have expected frequency of 1 in 50000 to 100000 live births. Potter and Craig used the term of heteropagus for asymmetrical conjoined twins. Parasitic twins account for 1-2% of all conjoined twins.⁵⁻⁷

The etiology of heteropagus twinning is yet to be determined but there are 2 dominant theories. The "fission" theory suggests incomplete separation of the embryo while the "fusion" theory proposes coalition of 2

originally distinct parts.^{1,2,8} The most advanced theory postulates that it occurs due to vascular compromise in utero, leading to death and partial resorption of one of the twins.^{1,2,4}

Since many aspects of this rare condition remain unknown, it has been recommended that all cases be reported⁸. Authors also discuss some peculiar findings from our report.

CASE REPORT

Our case is a baby boy with a thoraco-abdominal mass attached to the anterior abdominal wall near the umbilicus. The child was delivered at term to a 30 years old mother of 2 children in a nonconsanguineous marriage setting. There was no family history of similar malformation or twin pregnancy. Her antenatal period was uneventful but defect was not detected antenatally. The baby was delivered vaginally with a birth weight of 3.5kg. He was first brought to our hospital on day 7 of age.

On physical examination there was a 8*10cm mass attached with the autosite. The mass contains 2 rudimentary upper limb, pelvis with buttock and male external genitalia with hypoplastic scrotum and 2 well-formed lower limb with 5 digits with nails (Figure 1).

There was no spontaneous movement of the limbs and a pes equinovarus deformity of feet. Micturition was observed from both penis (parasite as well as autosite). Though the baby was acyanotic since birth, on auscultation a holosystolic murmur was found suggesting a cardiac defect in the autosite. Echocardiography confirmed the diagnosis of Single Ventricle.

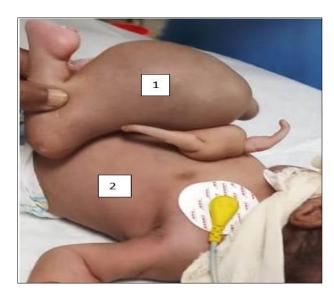


Figure 1: Parasitic twin (1) attached with host (2) near umbilicus.



Figure 2: CECT whole abdomen.

Ultrasonography shows a 3*4cm defect into anterior abdominal wall with gut loops entering into the 'parasitic twin'. CECT showed a soft tissue lobulated mass with fat and air components coming out of the anterior abdominal wall defect with calcification of lower limbs, absence of

vertebra, fused kidney like tissue and urinary bladder. Soft tissue shadow of external genitalia was also visible (Figure 2).

Cardiac defect was prime concern for both Surgeon and Anaesthesist. The mass could be separated from the autosite maintaining its vascularity and intestinal continuity was restored as there were no sharing of viscera between host and parasite. Rudimentary alimentary canal was present in the parasite. Abdomen was closed in layers. The mass with rudimentary limbs and genitalia was removed which on cut section shows the presence of fused kidney and bladder like structure (Figure 3).

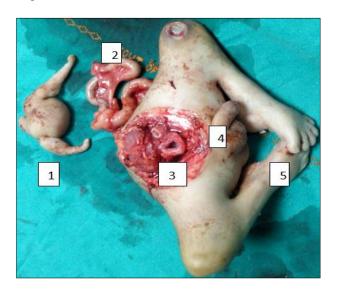


Figure 3: Specemen after excision (1) rudimentary upper limb, (2) rudimentary alimentary canal, (3) open bladder, (4) male external genitalia, (5) well developed lower limb with digits and nail.

Postoperatively patient was kept in NICU for cardiac defect and after stabilization he was discharged on POD 10. Cardiac abnormality need to be addressed later on.

DISCUSSION

Heteropagus twins are extremely rare and represent only 1 to 2 percent of all conjoined twins. 9,10 Most authors mention a clear predominance of the male sex with about 78% being males. 9,11 Until now, no particular risk factors for developing heteropagus twins have been reported. The most frequent form observed is the epigastric heteropagus, also called omphalopagus. Less common are ischiopagus, rachipagus and craniopagus. 10 This case is also a baby 'boy' and according to attachment 'epigastric heteropagus'.

This patient had acyanotic heart disease with a major defect (Single Ventricle). Single ventricle defects are rare. They occur in about 5 out of 100000 live births which include Hypoplastic Left Heart Syndrome (HLHS), Double outlet Right Ventricle, Tricaspid Atresia and

Double Inlet Left Ventricle¹². The autosites often have malformations themselves. Most common among these are congenital heart defects, especially Ventricular Septal Defects (VSD). Furthermore, the pumping twin can suffer from congestive heart failure due to the additional load on the cardiovascular system that has to supply the parasite twin as well. There is also a high incidence of associated omphalocele, especially in epigastric heteropagus twins. 9,11

In heteropagus twins, the parasite's limbs normally do not show any spontaneous movements. This is explained by the absence of neural innervation of the parasite myoblasts, which leads to incomplete differentiation and consecutively to skeletal muscular atrophy. Therefore the histology shows predominantly fat and bone but no muscle tissue. 9,11,13

Bony and visceral connections between the heteropagus twins can occur but are rarely observed. More frequently residues of parasite organs within the body of the autosite have been described. Vascular communications seem to be less marked than in symmetrical conjoined twins, making surgical separation less complicated.^{2,9}

Since organs aren't usually shared, preoperative imaging has usually been restricted to ultrasound, CT, or MRI. The aim is to reveal bony and soft tissue connections. However, since vascular connections are less complex, angiography is not required. Vascular communications can be determined intraoperatively without resulting difficulty, as was the case in our series.¹⁻³

The prognosis regarding survival of the autosite is good. In a review of cases by Sharma et al, the mortality rate was calculated to be 31 percent.² In order to reduce morbidity and for a good esthetic and functional results, a pregnant woman with the diagnosis of heteropagus twins should be referred to a perinatal center with experience in treating this disorder and be managed by a multidisciplinary team. Due to the rarity of this malformation, there is only limited literature available, mainly consisting of case reports.

CONCLUSION

Parasitic twins are rare, their sites and extent of attachment to the host twins varies. Result of such a major surgery varies due to abnormal sharing of viscera and associated anomalies. Our case improved remarkably inspite of major cardiac defect following successful excision of the "Parasitic Twin". Our case reporting might enlighten this rare spectrum of defects.

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