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Original Research Paper

Anatomy

PYOPAGUS CONJOINED TWINS WITH VENTRAL ABDOMINAL WALL DEFECT

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ABSTRACT Pyopagus conjoint twin represent a rare developmental defect with shared gluteal region and with fusion of sacrum facing away from each other. Pyopagus twins have an incidence of 6% among conjoined twins. We report a case of pyopagus conjoint twin with the site of fusion being the sacrococcygeal region. One of the twin was normal in appearance while the second twin had absent ventral abdominal wall below the umbilicus. Prenatal scanning makes an early diagnosis of the type of conjoint twin and determine the outcome of pregnancy. The surgical intervention can be determined according to the site of fusion and the organs shared. Pyopagus twin management involves separation and reconstruction of gastrointestinal, genitourinary and the reproductive tracts and the outcome of survival depends on the organs shared.

KEYWORDS : conjoint twins, pyopagus, ventral body wall, umbilicus, urogenital system.

INTRODUCTION

Conjoined twins present the most fascinating human congenital anomaly with incidence of about 1:50000 – 1: 200000 births[1]worldwide with higher incidence in Southwest Asia and Africa in a range of 1:14000 – 1: 25000. Conjoint twinning has also been reported in mammals, fishes, birds, reptiles and amphibians. Incidence was predominant in females over males around 2:1- 3:1[1]. The exact etiology and genetic cause of conjoint twin is unknown, most of them are monozygotic. They results due to incomplete division of embryonic disc more than 13-14 days after fertilization at about primitive streak stage.Two theories have been associated with conjoint twins one is fission and other fusion. In fission there is incomplete splitting of embryonic axis, whereas in fusion there is complete separation of zygote[2].

Eight different subtypes identified depending on the region of fused body parts: omphalopagus, thoracopagus, cephalopagus, ischiopagus, parapagus, craniopagus, rachipagus and pyopagus. Incidence of occurrence of the various conjoint twins are as follows: parapagus (28%), thoracopagus (19%), omphalopagus (18%), cephalopagus (11%), pygopagus (6%), ischiopagus (11%), and craniopagus (5%)[3].Most conjoined twins are born prematurely and almost 40% are stillbornwith 60% live births[4].

The first incidence of pyopagus was seen around 1100s[5]. Generally the pyopagus conjoint twins are fused in the gluteal region with the fusion of sacrum facing away from each other. These twins have a common sacral spine, rectum, anal canal, urinary and reproductive system[6].

Successful separation of conjoined twins requires skilled clinical assessment and detailed radiological study due to variable and complex anatomy with associated malformations. Prognosis of pyopagus twin is good with survival rate of 87%.

In the present paper for the first time we report a case of pyopagus conjoint twin with one twin (twin 2) presenting with ventral body wall defect and absent urogenital system.

CASE REPORT

A 11week old medically terminated pregnancy was brought to the Department of Anatomy following an antenatal diagnosis of pyopagus conjoined twin for fetal autopsy from Department of Obstetrics &Gynaecology, Government Medical College & Hospital, Chandigarh, India. Consent was taken from the parents to perform the autopsy. Parenteral history revealed maternal age of 34years with no significant medical illness. There was no family history of twinning.

Antenatal ultrasound findings showed twins in the same sac with fusion of lower back suggested of pyopagus conjoint twins.

Autopsy finding showed two fused fetuses with separate umbilical cords. Fusion was seen dorsolaterally in the sacrococcygeal region (Fig 1). Twin 1 was normal in appearance but twin 2 had absent ventral abdominal wall below the umbilicus. The abdominal contents (part of liver, small intestine) of twin 2 were exposed to the exterior (Fig 4). Umbilical cord of both the twins showed two vessels(single umbilical artery and umbilical vein).

The twins were separated by an incision in the fused region. The twin 1 was normal on internal examination with female internal genitalia. The twin 2 showed complete absence of urogenital system, partial formation of gastrointestinal system with absence of large intestine (Fig 4).

Radiological examination observed fused lower spine at the level of sacrum (Fig 5).

DISCUSSION

Conjoint twins are thought to be an incomplete separation of a single embryo. The other embryonic causes described in the literature are the result of (i) Duplication of inner cell mass (ii) incomplete duplication of embryonic disc; both resulting in monochorionic monoamniotic placenta[7]. The present case is an example of incomplete separation of two early blastomeres giving rise to two chorions, hence two placentae and two umbilical cords[4]. However, some authors are in the view that there is a secondary fusion of the two originally separate

monozygotic embryonic discs[1].

Pyopagusconjointwins occurs in 6% of the cases with two bodies joined back to back at the pelvis and lower spine, each with separate hearts, heads and limbs.Pyopagus twins have a lower mortality rate (23%) when compared to thoracopagus (51%)[8]. According to literaturepyopagus conjoint twins share a common gluteal region, terminal spine, gastrointestinal, urological and reproductive systems. However, in the present case there was absence of genital, urinary and parts of digestive system in twin 2, which was not reported earlier in the literature. The absence of urogenital system relates with the fact that the development of urinary and genital systems is complimentary to each other, that is one induces the development of the other.

Along with above mentioned defects we also observed ventral body wall defects which could be due to failure of migration of intraembryonic mesoderm below the level of umbilicus thereby resulting in ventral abdominal wall defects. Hedgehog signaling isconsidered as the key factor for the development of urogenital system and the ventral abdominal wall. The imbalance in the expressionof the Hedgehog (Hh) signaling pathway could be a hypothesis for the ventral abdominal wall defect and absent urogenital system.However, the relationship between Hh signaling and ventral body wall formation remains unclear.

Previous literature did not reportany defect in the umbilicus, but the present case for the first time reported two separate umbilicus containing single umbilical artery and umbilical vein.

Antenatal ultrasound helps in prenatal diagnosis of the type of conjoint twin and depending on the diagnosis early termination of pregnancy can be advised. The surgical management of conjoint twin can be determined according to the site of fusion and the organs shared. Pyopagus twin management involves separation and reconstruction of gastrointestinal, genitourinary and the reproductive tracts.

Conjoined twins are rare developmental defect withvariable and complex anatomy, among which pyopagus twins are one of the rarest types of conjoint twins. They are often frequently associated with malformations. Although the incidence of conjoint twins is low, their occurrence has allowed embryologists to further explore the dimensions of development in multi-fetal pregnancies. Further work at molecular level is required to rule out the exact etiology and associated risk factor for the occurrence of this interesting case.

FIGURES FIGURE LEGENDS

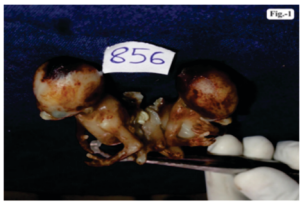

Fig 1: Pyopagus twins fused in the sacrococcygeal region.

Fig 2: Ventral body wall defect below umbilicus. Note: Exposed intestinal contents.

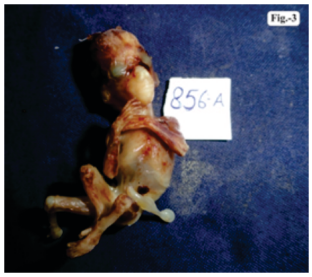

Fig 3: Twin 1 showing normal appearance.

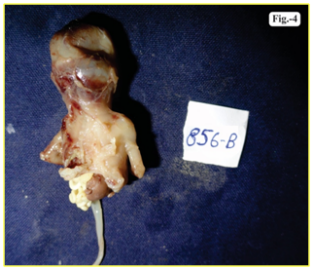

Fig 4: Twin 2 showing ventral body wall defect with absence of genitourinary system.

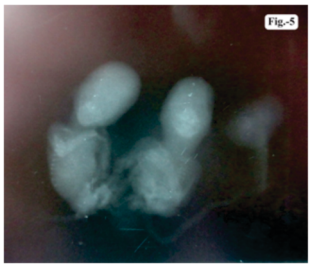

Fig 5: Radiograph of twins, showing fusion at the level of sacrum.

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