Ischiopagus Tetrapus, Parasitic Tripus Conjoined Twins: A Rare Anomaly

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ABSTRACT

Ischiopagus parasitic conjoined twins is a rare and intriguing anomaly with limited number of case reports in literature addressing clinical presentation and surgical intervention performed. A set of ischiopagus tetrapus heteropagus tripus conjoined male twins is reported who were not diagnosed on antenatal ultrasound. First stage surgical separation was done with uneventful recovery.

Key words *Ischiopagus, Parasitic twins, Surgical separation, Conjoined twins.*

INTRODUCTION:

A parasitic conjoined twins, also known as heteropagus twins, is a developmental fetal anomaly in which part of a defective twin is attached to an autosite, who may have other associated congenital anomalies. This is a rare occurrence with an incidence of less than I per million live births.[1] Many of such twins die early in postnatal period.[2] However, those who survive, provide an opportunity to understand more about their developmental anomalies and the challenge they pose in the management. In this report a rare form heteropagus conjoined twins is presented which adds to the existing knowledge about such malformations and highlights management issues.

CASE REPORT:

An 11-hours old male, full term set of twins, weighing 2.1 kg, presented to the emergency department, referred from another hospital with a parasite attached to normal looking baby. Antenatal workup did not suggest any fetal anomaly. Baby cried immediately after birth, passed urine from an anomalous site in the perineum. Meconium was also passed through the same site. There was no history of antenatal illness or drug intake in mother.

On examination a set of heteropagus conjoined twins, united at the perineal region found. The autosite was active and pink looking with a normal appearing head, torso and upper limbs. There were two lower limbs of which, the right lower limb was abnormally positioned (at hip and knee joints) with limited movement (only adduction and extension possible), knee joint cap faced medially with club foot. Left lower limb of autosite was normal except for its position, which was too lateral.

The umbilicus of the autosite was low lying situated over a bulge protruding through the abdominal cavity. A reddish pink mucosal surface was found adjacent to it, suspected as urinary bladder plate, along with an opening from which meconium was passed. A small, empty left scrotal fold was found lateral to the opening. Another empty scrotal fold was found between the non-functioning lower limbs of the parasite and the right abnormal lower limb of the autosite, along with a pit-like structure placed medial to it. The parasite had two non-functioning hypoplastic upper limbs of different sizes, a small bony structure, a third limb, which was attached to pelvic ring of autosite, probably head of the femur, a large cystic swelling in the middle, sparsely covered with tuft of hair with a small area of pinkish mucosa bulging and flickering occasionally (Fig. I).

The set of twins was admitted and work-up done to find out associated anomalies as well as assessment of anatomical structures within the parasite. A baby-gram along with pelvic x-rays, echocardiography, ultrasound abdomen, CT scan with oral and IV contrast were done. Biochemical profile was reported as within normal limits. There were no cardiac anomalies. Autosite had two normal appearing kidneys and ureters however urinary bladder was

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Fig. I: The Ischiopagus Parasitic Twin

Fig. II: Patient After Surgical Removal of Parasite

not identified. It was represented as exstrophied part in lower torso. A cystic area measuring 3.5cm x 1.5cm identified in the pelvis of autosite while a large cystic space found in parasite. Examination under anesthesia showed a small rectal opening with bladder plate. No urethral opening was found. Left testis was palpated slightly upwards in inguinal canal. Right testis was not found.

Surgery was performed on elective basis for the separation of parasitic twin. Incision was made keeping in mind the difficulty in closure of the wound. The parasite has broad based attachment with autosite but separated easily and no major vessels were encountered. Two suspected upper limbs of the parasite attached to the cystic structure were removed and a third rudimentary extra suspected lower limb with femoral head attached to the pelvis on the right side was taken out along with a part of pelvic bone of autosite (Fig. II). Postoperative recovery was uneventful. Physiotherapy was advised for right lower limb. The baby is on follow up. Additional surgical procedures are in plan as baby grows older for improving quality of life.

DISCUSSION:

Many challenges are faced in the management of heteropagus conjoined twins specially when they are found in the vicinity of vital structures or along with other organ systems.[3] Ischiopagus parasites are attached to the lower abdomen and pelvis of an autosite. This malformation accounts for about 13% of all heteropagus twins.[4] In this type of malformation slight male preponderance is reported. The patient in our study was also a male baby. The anomaly was not picked up in antenatal period as reported in another case report from a developing country however, their patient was a female.

The workup of patient in our case was exhaustive so as to know the anatomy before undertaking surgical removal of parasite. It is important to have detailed assessment before embarking upon surgical procedure.[6] The surgery was easy without much difficulty in removing the organs of the twin, however, number of issues remained. There was no external genitalia. Urinary bladder was exstrophied with urinary incontinence. Fecal control is expected as intact anus was found. Orthopedic issues are also a challenge for the future although physiotherapy has resulted in more normal position of right lower limb. In future a multidisciplinary team including pediatric surgeon, urologist, orthopedic and plastic surgeons can plan stage procedures for improving quality of life in this patient.

REFERENCES:

1. Sharma G, Mobin SS, Lypka M, Urata M. Heteropagus (parasitic) twins: a review. J Pediatr Surg. 2010;45:2454-63. doi: 10.1016/j.jpedsurg.2010.07.002.

2. Khan YA. Ischiopagus tripus conjoined twins. APSP J Case Rep. 2011;2:5.

3. Mahajan JK, Devendra K, Mainak D, Rao KL. Asymmetric conjoined twins: atypical ischiopagus parasite. J Pediatr Surg. 2002;37:E33. doi: 10.1053/jpsu.2002.35446.

4. Gul A, Aslan H, Ceylan Y. Prenatal diagnosis of pygopagus tetrapus parasitic twin: case report. BMC Pregnancy Childbirth. 2004;4:13. doi: 10.1186/1471-2393-4-13.

5. Muhelo AR, Montemezzo G, Da Dalt L, Wingi OM, Trevisanuto D, Gamba P, et al. Successful management of a parasitic ischiopagus conjoined twins in a low-income setting. Clin Case Rep. 2018;6:385-390. doi: 10.1002/ccr3.1374.

6. Wood BC, Sher SR, Mitchell BJ, Oh AK, Rogers GF, Boyajian MJ. Conjoined twin separation: Integration of three-dimensional modeling for optimization of surgical planning. J Craniofac Surg. 2017;28:4-10. doi: 10.1097/SCS.0000000000003412.

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Riaz Ahmed: Concept, patient management, literature search and report writing.
Jamshed Akhtar: Concept, patient management, literature search and report writing.
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Ethical statement: Permission was taken from parents to present the data in a poster format as well as case report for dissamination of scientific knowledge.

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