Case Report

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Cloacal exstrophy with vesicoureteric junction obstruction: Rare association

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ABSTRACT

Cloacal exstrophy is an extremely rare congenital abnormality resulting in an exstrophy of the urinary, intestinal and genital organs and associated with anomalies of other organ systems. We report a complicated case of cloacal exstrophy associated with right vesicoureteric junction obstruction.

Key words: Cloacal exstrophy, hydroureteronephrosis, percutaneous nephrostomy, vesicoureteric junction obstruction

INTRODUCTION

Cloacal anomalies exhibit a wide variety of morphological types and accompanying clinical severity thought to result from a mesodermal abnormality. Anatomically, its main components are omphalocele, bladder exstrophy and imperforate anus. Other associated malformations include renal malformations and spine defects. Reported incidence is one in 200 000 to 400 000 births.[1] Cloacal exstrophy with vesicoureteric junction obstruction is a rare association, which prompted us to report this case.

CASE REPORT

A ten-days-old male neonate presented to a paediatric emergency department by his parents and on external examination, the ventral abdominal wall was found to be defective with a small omphalocele with an intact sac with exstrophy elements in the lower part [Figure 1a]. He was born by caesarean section delivery to a Muslim family, the product of consanguineous marriage, 4th in order, perinatal period uneventful, no records of antenatal follows-up available. Male sex was confirmed after examination of the external genitalia. Just above the epispadiac glans penis, there was a duplicated bladder patch with 2 ureteric orifices separated by an extrophied cecal patch. There was an imperforate anus with only a dimple seen in the perineum. The penis was epispadiac. On x-ray, sacral agenesis and spina bifida was detected. Further examination of the abdomen with sonography showed hydronephrosis along with intra-abdominal testicles. The child was passing stool from the blinding cecal patch. Urine was seen egressing from the exposed bladder mucosa. The child was thoroughly worked up and nutritionally built up for definitive surgery. While awaiting surgery, child urine output decreased slowly over a period of 48 hours. Ultrasonography was done, which showed gross hydronephrosis on right side. Urgent percutaneous nephrostomy (PCN) [Figure 2a] was done under ultrasonographic guidance. Antegrade dye study done through PCN showed obstruction in the vesicoureteric junction region [Figure 2b and 2c]. The renal dynamic scan also showed an obstructive drainage pattern on right side with hydroureteronephrosis on the ipsilateral side. After stabilizing the child, surgery

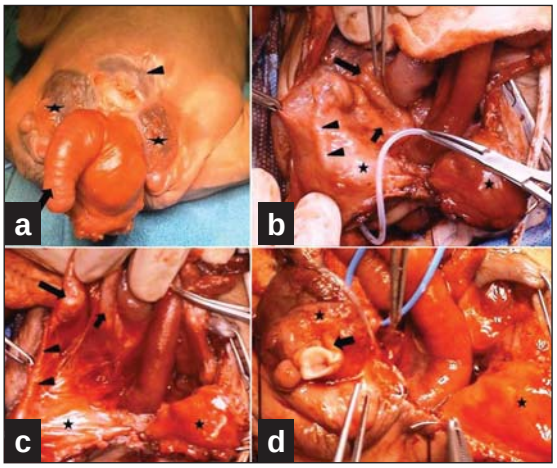


Figure 1: Cloacal exstrophy with two hemi-bladders (asterixes) and central prolapsed cecum (thick arrow) and omphalocele minor (arrow head) [1a]. Intra-operative photographs showing dilated right ureter (thick arrows) and narrow lower end including vesicoureteric junction (arrow heads) (asterixes: Two hemibladders) [1b and 1c]. Intra-operative photograph showing right ureteric re-attachment after excision of narrow segment (thick arrow) (asterixes: Two hemibladders) [1d]

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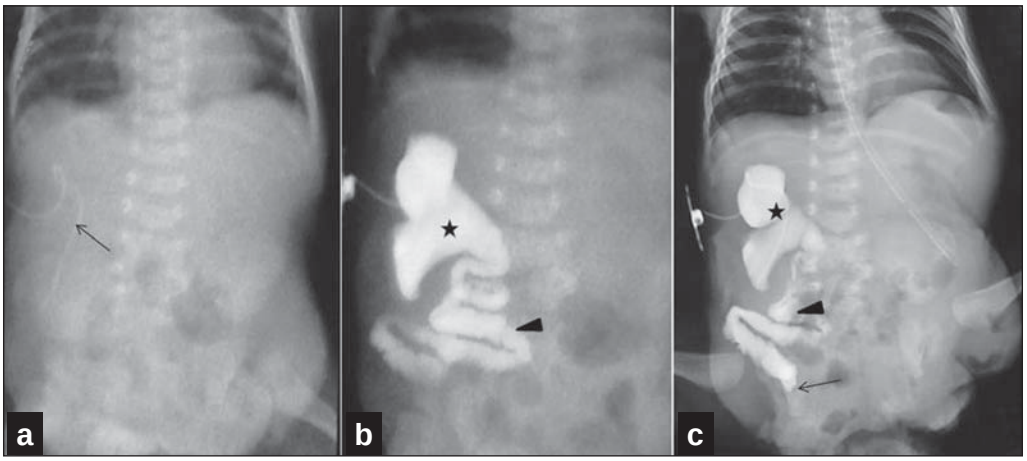


Figure 2: Post-PCN X-ray showing catheter *in situ* (thin arrow) [2a]. Antegrade dye study done through PCN (an early image: 10 min) showing dilated right pelvicalyceal system (Asterix) and dilated ureter (arrow head) [2b] and (delayed image: 2 hrs) showing retained contrast in dilated right pelvicalyceal system (Asterix) and dilated ureter (arrow head) and contrast cut off at vesicoureteric junction (thin arrow) [2c]

was performed at 4 weeks of life. Intraoperatively, right ureter was dilated and tortuous with narrow lower end including vesicoureteric junction [Figure 1b and 1c]. Primary bladder closure with re-attachment of the right ureter [Figure 1d] along with excision of cecal patch and abdominoperineal pull through was done. Postoperatively, child was electively ventilated for 72 hours. Presently, child is doing well and is in close follow-up awaiting epispadias repair.

DISCUSSION

Cloacal exstrophy represents a spectrum of rare congenital anomalies that are thought to arise from mal-development of the cloacal membrane, which prevents migration of mesenchymal tissue and impedes normal development of the lower abdominal wall. The defect is thought to be because of premature rupture of the cloacal membrane before descent of the urorectal septum at approximately 5 weeks gestation.[2] Cloacal exstrophy has complex anatomy, with omphalocele present at the superior extent of the ventral abdominal-wall defect and exposed bowel and bladder at the inferior extent with two hemibladders, which are separated in the midline by a zone of intestinal mucosa. Each hemibladder may have a ureteral orifice. The intestinal zone separating the hemibladders may have the orifices of the proximal gut superiorly and the distal gut inferiorly, with one or two appendiceal orifices in between. The proximal bowel orifice (cecal plate) often prolapses in the characteristic "elephant-trunk" deformity. The distal gut is a blind pouch as these infants all have imperforate anus. Occasionally, a colonic duplication may be found. Although cloacal exstrophy was recognized as early as 1909, it was not until 1960 that the first successful reconstruction was reported.[3] It is only in recent decades that repair of this complex anomaly has been routinely undertaken. For years, most infants were allowed to die because of the multiplicity and complex nature of the congenital anomalies.[4] Survival has increased significantly over last two decades from 0% to 90%.[5, 6] Cloacal exstrophy is associated with anomalies of organ systems other than the central defect in up to 85% of cases.[6] Various studies has reported associated incidence of urinary tract anomalies from 40-60 %.[6-9] Vertebral anomalies occur in 48% to 78% of patients[6,8,9] myelodysplasia, and tethered cord in 29% to 46% of patients[6] Over the last 30 years, the focus has drifted towards improving patient outcome in term of quality of life, which includes an appropriate gender assignment, independence from stoma appliances, higher level of physical and social independence, and mobility. Nowadays, the priority is given to urinary, gastrointestinal and genital reconstruction, designed to adapt the patient as a free person and of appropriate psychological gender. To our knowledge, association of cloacal exstrophy with vesicoureteric junction obstruction is not reported yet. In a series of 06 patients, which we have operated till date, vesicoureteric junction obstruction was seen in 01 case only. We performed corrective surgical procedure in neonatal period itself either as single-stage total reconstruction or as a staged procedure. In the present case, primary bladder closure with right ureteric re-attachment along with abdominoperineal pull through was done. Postoperatively, child is doing well and is in close follow-up. From this, we can conclude that cloacal exstrophy with or without associated malformation in itself is not a hopeless situation. A dedicated and comprehensive surgical planning is required for achieving a reasonable outcome in these cases.

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