Case Report

Cloacal Malformation Variant in a Male Neonate

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ABSTRACT

Cloacal malformation is a rare entity and is invariably referred only to females. We are reporting a very rare case of cloacal malformation variant in a 6-day-old male neonate who presented with absent anal opening along with passage of urine and meconium from an abnormal opening in the perineum.

KEYWORDS: Cloacal malformation, male, neonate, perineal opening, variant

INTRODUCTION

Cloacal malformation is a very rare entity with a frequency of 1:50,000–1,25,000 new born.[1] It is invariably referred only to females.[2] On the contrary, one of the earliest reported cases of persistence of the cloaca was a male fetus.[3] There are only a few cases of male cloaca reported in the recent literature. In addition, there is a lack of consensus regarding the entity in males and its management.[1,4–6] We share our experience with an extremely rare case of cloacal malformation in a male neonate.

CASE REPORT

A 6-day-old male neonate, weighing 2100 g, presented to us with passage of urine and meconium from a single perineal opening along with abdominal distension and bilious vomiting. On examination child was afebrile, mildly dehydrated, and tachypneic. The abdomen was soft and moderately distended and there were visible veins over the abdomen. Perineal examination revealed absent anal opening (without anal pit) and presence of a single midline anterior perineal opening at the proximal part of the scrotum, just distal to the junction of scrotum and perineum [Figure 1]. Urine was seen coming out of the perineal opening along with small amount of thick tenacious meconium. On careful inspection, there was a small cavity underneath the perineal opening; two smaller openings, one anteriorly and other posteriorly placed, were seen in the posterior wall of the cavity [Figure 1]. Testes were bilaterally descended and severe chordee with perineal hypospadias along with penoscrotal transposition was present [Figure 1]. An initial VACTERL workup was done. A left transverse colostomy was performed after preoperative optimization.

The patient was investigated at 8 months of age. Laboratory values revealed anemia (Hb-8.1 g%) and raised total leukocyte count (13,400/mm³). Chromosomal analysis revealed normal male (XY) karyotype. Internal organs were male; both testes were normal sized and well placed in the scrotum. Distal colostogram revealed rectum tapering before entering into the common channel [Figure 2].

Cystoscopy showed a common channel with a length of 1 cm with separately placed urethral opening (anteriorly) and rectal opening (posteriorly). The proximal urethra was normal. There were flakes in the urinary bladder with features of cystitis; ureteric openings were normal. Under general anesthesia, anterior urethral opening in common channel was inspected and bladder catheterization was done. Infant feeding tube was placed in posteriorly placed rectal opening to confirm the rectal opening in the common channel [Figure 1]. Posterior sagittal anorectoplasty was performed; common channel was repaired and kept as urethra. The patient did well postoperatively and correction of perineal hypospadias is being planned.

DISCUSSION

The cloaca is a single common channel which is formed at the 3rd week at the tail fold through the confluence of...
Male cloacal malformation

Photographs showing single midline perineal opening at the proximal part of the scrotum with small cavity underneath it (a); two smaller openings, one anterior (with Foley’s catheter in situ shown by yellow arrow) and other posteriorly placed (black arrow), are seen in the posterior wall of the cavity (b); infant feeding tube placed in posteriorly placed rectal opening and irrigation of the distal colonic loop done to confirm the rectal opening in the common channel (c); bilaterally descended testes, penoscrotal transposition, severe chordee, and perineal hypospadias along with neoanus are seen (d)

Figure 1: Photographs showing single midline perineal opening at the proximal part of the scrotum with small cavity underneath it (a); two smaller openings, one anterior (with Foley’s catheter in situ shown by yellow arrow) and other posteriorly placed (black arrow), are seen in the posterior wall of the cavity (b); infant feeding tube placed in posteriorly placed rectal opening and irrigation of the distal colonic loop done to confirm the rectal opening in the common channel (c); bilaterally descended testes, penoscrotal transposition, severe chordee, and perineal hypospadias along with neoanus are seen (d)

The allantois and the hindgut. Series of events lead to its partition and separation of urinary system, genital tract, and anorectum. Persistent cloaca is conventionally defined as “a single perineal orifice formed as a confluence of urinary, genital, and rectal systems ending in a common channel of varying lengths.” According to Livingston et al., all patients with persistent cloaca are female and have normal ovaries. Most of the surgical literature refers only to females as having cloacal defects. However, there is every chance of the development of cloacal anomalies in males as during embryogenesis; both sexes have to pass through the stage of a cloaca.

Male cloacal variants were defined as a defect in the male with a single perineal opening, in which rectum and urinary tract, with or without vagina, converge into one common channel. Male cloaca was referred as a single opening in perineum for passage of urine and meconium. We define male cloaca as common channel of varying lengths with a single perineal orifice, containing separate openings for the urinary tract (anteriorly) and the rectum (posteriorly).

As per Peña et al., the term posterior cloaca refers to a malformation in which the urethra and vagina are fused, forming a urogenital sinus that deviates posteriorly to open in the anterior rectal wall or immediately anterior to the anus. The rectum is essentially normal or may be minimally anteriorly mislocated. Hendren describes posterior cloaca as rare cloacal malformation in boys. As the common channel in cloaca can deviate anteriorly or posteriorly, thus posterior cloaca in boys as reported by Hendren could be a subset of a male cloacal malformation. The differences in cloacal anatomy are a result of the divergent differentiation of the genitourinary tracts in the two sexes. The spectrum of cloacal abnormalities may result from a partial to complete urorectal septum malformation.

Male cloacal variants may present in the neonatal period and up to 5 years. Late presentation (28 years) has also been reported. Single opening may be located anywhere from anterior perineum to the tip of glans. Length of the common channel ranged from 0.5 to 8 cm. Vaginal remnant (not seen in our case) has been reported in 50% cases of male cloacal variants. Genital abnormalities are severe in cloacal malformation, and proximal hypospadias is a common finding. Renal abnormalities have also been reported.

The differential diagnosis of male cloaca is anorectal malformation with rectourethral fistula (or rectovesical fistula) with proximal (perineal) hypospadias. In males, the spectrum of cloacal abnormalities is difficult to recognize. The presence of common channel should be confirmed by careful visual examination followed up by distal colostogram and cystoscopy. Cystoscopy helps in delineation of fistula tract, length of common channel, and evaluation of urethral and bladder abnormalities. In addition, a search for the presence of any vaginal remnant should be performed.

Colostomy as part of initial management was also performed by other authors. Definitive management consists of either (a) posterior/anterior sagittal anorectoplasty and urethroplasty or (b) posterior sagittal anorectourethroplasty or (c) perineal urethrostomy and anoplasty followed by urethroplasty. Common channel should be preserved.

Figure 2: Nonfluoroscopic well-tempered pressure-augmented distal colostogram: Anteroposterior view (a) showing rectum tapering before entering into the cavity along with complete bladder filling; lateral view (b) showing rectum descending below the “I” line; rectum (red arrow) and urethra (yellow arrow) communicating with a common channel (yellow bracket). TC: Transverse colon, C: Colostomy, S: Sigmoid colon, R: Rectum, B: Bladder

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at the time of anorectoplasty. This common channel functions as male urethra and is incorporated at the time of urethroplasty. Furthermore, colostomy closure should be undertaken after complete correction of the hypospadias to prevent soiling of the hypospadias repair and also to prevent ascending urinary tract infection due to its proximity to the neoanus. Mortality is not rare (because of associated multiple anomalies). [2]

**CONCLUSION**

A high index of suspicion for the presence of common channel should be present in a male patient of ARM with single perineal opening associated with genital abnormalities.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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