Importance and difficulty of correctly diagnosing covered cloacal exstrophy for adequate reconstruction: A case report

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Abstract

Covered cloacal exstrophy (CCE) is extremely rare condition. In patients with a single perineal orifice and no pubic bone separation, it is very difficult to suspect and/or diagnose CCE based on external signs alone. We present the case of a 2-month-old girl diagnosed with CCE based on cystography, ileostomy contrast study and cystoscopy.

Covered cloacal exstrophy (CCE) is extremely rare condition. Although the visceral features of CCE are similar to those of cloacal exstrophy (CE), affected individuals have an apparently intact lower abdominal wall, unlike those with CE [1]. In patients with a single perineal orifice and no separation of the pubic bones, it is very difficult to suspect and/or diagnose CCE on the basis of external signs alone [2]. Patients with CCE can be misdiagnosed as having cloacal malformation and thus undergo attempted colostomy. We present the case of a 2-month-old girl whom we highly suspected of having CCE based on cystography (CG), ileostomy contrast study and cystoscopy. The definitive diagnosis of CCE was made based on visceral features at exploratory surgery.

1. Case report

A female neonate weighing 2132 g was born by vaginal delivery at 35 weeks' gestation. She was found to have a single perineal orifice with an imperforate anus but no evidence of separated pubic bones. Bilateral hydronephrosis was detected with ultrasonography. The patient was diagnosed with cloacal malformation and colostomy was attempted on her first day of life. However, ileostomy was performed, because her colon could not be identified. After ileostomy the patient suffered from recurring urinary tract infections and metabolic acidosis. She was transferred to our hospital for further management at 41 days of age.

At the time of admission, we suspected cloacal malformation based on the patient's external physical characteristics: a single perineal orifice and no pubic bone separation, and recurrence of urinary tract infections. Based on the patient's clinical presentation, cystography, ileostomy contrast study and cystoscopy, we suspected that she had CCE. We performed exploratory surgery to confirm the CCE diagnosis and to ligate the enterovesical fistulae to prevent repeated urinary tract infection. We found bilateral ovaries and fallopian tubes and a bicornuate uterus. Anomalies of the gastrointestinal tract were consistent with the results of the contrast study. The patient's visceral features were similar to CE, and raised suspicion that she had CCE.

We performed exploratory surgery to confirm the CCE diagnosis and to ligate the enterovesical fistulae to prevent repeated urinary tract infection. We found bilateral ovaries and fallopian tubes and a bicornuate uterus. Anomalies of the gastrointestinal tract were consistent with the results of the contrast study. The patient was transferred to our hospital for further management at 41 days of age.
the colon was short, with narrow segment (Fig. 4a–d). We made a definitive diagnosis of CCE. Because there was narrow segment of the colon, we were not able to perform short-colon-sparing surgery. We performed ileostomy closure, ligation of the colovesical fistulae, and creation of a new permanent end ileostomy. Pathological examination revealed tissue similar to gastric pyloric glands with mild fibrosis at the site of colon narrowing.

After surgery, the patient’s urinary tract infections, bilateral hydronephrosis, and metabolic acidosis improved. She has been growing adequately with oral nutrient intake alone. In the future, we will evaluate the patient’s genital anomaly and urinary function.

2. Discussion

It is difficult to distinguish CCE from cloacal malformation based on external signs alone. Bischoff et al. evaluated 31 patients with CCE and found that low implantation of the umbilical cord in association with separated pubic bones and anorectal malformation are the most common signs of the condition [2]. Patients with CCE are misdiagnosed surprisingly often as having cloacal malformation and receive an ileostomy or proximal colostomy. After ileostomy, such patients suffer from recurrent urinary tract infections and hyperchloremic acidosis resulting from colonic absorption of urine.

Patients who undergo ileostomy or proximal colostomy need a “rescue operation” consisting of ileostomy closure, ligation of the colovesical fistula, and creation of a true end colostomy to allow growth of the colon [3,4]. Levitt et al. reported that pull-through is possible in patients with CCE. To maximize the bowel’s ability to form solid stool, it is crucial to use all available hindgut for the initial colostomy and to avoid using the colon for urologic or genital reconstruction [3]. It is ideal to perform ligation of the colovesical fistula and creation of a true end colostomy at initial surgery. However, distinguishing between CCE and cloacal malformation is very difficult.

If the surgeon cannot identify the colon during the initial transverse colostomy in the cloacal malformation patient, CCE should be suspected. In these cases, exploratory surgery or diagnostic laparoscopy should be performed to confirm the structure of the gastrointestinal tract and urogenital organs before ileostomy. This is the only way to correctly diagnose the condition and allow for preservation of a short colon.

In this case, the patient had already received an ileostomy; the CG, ileostomy contrast study and cystoscopy were useful in determining the correct diagnosis and we were able to perform the rescue operation immediately. Cooperation with pediatric urologist was essential to correctly diagnose the urogenital organs condition.

We couldn’t preserve the short colon because the oral side of colon was narrowing. If we could do the colon strictureplasty and performing a true end colostomy, it gives the patient the opportunity of a future pullthrough or, if not possible, leaving that gastrointestinal tissue for future bladder augmentation.
In our patient, the lack of a bladder neck and resulting small bladder may cause urinary incontinence; bladder augmentation will likely be required. Oshita et al. reported the creation of a continent urinary reservoir from cecum and blind-ended colon. The bladder and cloacal canal were used as a vagina and a single orifice was adequate for future sexual intercourse [5]. Reconstructive surgery must be specific to each case.

Fig. 3. Cystoscopy. (a) Enlarged bladder neck and two inlets of enterovesical fistulae. (b) Hindgut bound to the ileum. (c) We identified the balloon catheter inserted from the ileostomy into the distal side ileum. (d) Blind-ended hindgut.

Fig. 4. Surgical findings. (a) Single appendix aided identification of the colon. (b) Ileum bound to bladder neck. (c) After transect enterovesical fistulae. (d) Resected colon. The colon is short, approximately 10 cm in length, with narrow segment.
3. Conclusion

CCE is extremely rare. We think that true end colostomy or rescue operation is the best procedure to preserve the short colon and it gives a patient opportunity of a future pull-through operation. An early correct diagnosis is important for adequate reconstructive surgery for CCE.

Conflict of interest statement

There are no financial or personal conflicts to disclose.

References