Bladder extrophy and postoperative intussusception due to Meckel's diverticulum: A confluence of congenital anomalies

Wesley W. Ludwig a,*, Seth D. Goldstein b, John P. Gearhart a

a Department of Urology, Johns Hopkins Hospital, Baltimore, MD, United States
b Department of General Surgery, Johns Hopkins Hospital, Baltimore, MD, United States

1. Case report

1.1. Patient history

The patient is an otherwise healthy fourteen month-old female with CBE. She presented to the Johns Hopkins Hospital for consultation and was taken to the operating theater for an exam under anesthesia at two weeks of life. Her bladder template was demure, measuring 2×2 cm with five large polyps resected at that time. Pelvis x-ray revealed a pubic diastasis of 3.84 cm. At four months of age another exam under anesthesia was performed to determine if her bladder had undergone any significant growth. Her bladder increased to 2.7 by 2.5 cm, but at eight months her bladder remained the same size. Additionally, her bladder was extremely fibrotic and she had developed recurrent bladder polyps that required polypectomy. It was determined there was no possibility of significant bladder growth, and due to recurrent bladder polyp formation and urine-induced cutaneous irritation and fungal infections a definitive procedure was performed at one year of age.

1.2. Surgical approach to bladder extrophy

A midline incision was made and the bladder was freed from the rectus fascia down to the pubis. Due to the demure, fibrotic nature of the bladder, the ureters were transected and a cystectomy was performed. A Masson's trichrome stain showing severe fibrosis can be seen in Fig. 1A segment of alloderm was sewn to the medial aspect of the levator hiatus bilaterally to serve as a protectant under the pubic closure. A perforation was created in the peritoneum and the left ureter was brought posterior to the mesentery of the sigmoid colon to meet the right ureter. The medial aspect of both ureters were sharply incised and joined. The ureters were exteriorized through rectus fascia in the lower abdomen, below the level of the umbilicus and a cutaneous ureterostomy was created. The peritoneum was closed and the pubic bone was reapproximated. A genitoplasty was also performed. The mucosa of the vagina and distal urethra were brought to the medial aspect of the clitoral halves and the bifid clitoral hoods were joined in the midline. There were no intraoperative complications. Following the urologic...
procedure, bilateral anterior and posterior iliac osteotomies with placement of an external fixator was performed by the orthopedic surgery team. She was then extubated and transferred to the pediatric intensive care unit with a nasogastric tube (NGT) in place.

1.3. Hospital course

Initially, the patient did very well post-operatively and progressed in the expected manner. Due to the extensive surgery, osteotomies and external fixator, she was maintained on bedrest in traction. She remained NPO with NGT to suction until post-operative day (POD) 3 when she had a bowel movement. NGT clamp trials were started but she began to experience emesis. Following this, she continued to have low NGT output but no further BM’s. An abdominal x-ray (AXR) as seen in Fig. 2 was obtained and showed small bowel distension consistent with ileus. Throughout the rest of her postoperative course she had intermittent bowel movements, but her abdominal distension persisted and she had significant NGT output. This course was initially thought to be consistent with ileus typically seen following CBE repair, but on POD 8 she was started on parenteral nutrition and general surgery was consulted. After a period of expectant management with serial examinations and radiographs without significant improvement, the patient was taken POD 14 by the general surgery and urology teams for open surgical exploration.

1.4. Surgical approach to intussuscepted Meckel’s diverticulum

The patient was positioned supine on the operating table and sterilely prepared without disrupting the protuberant external fixators. Her lower midline incision was reopened and extended to the superior border of her umbilicus. The small bowel was completely eviscerated and run from the Ligament of Treitz to the ileocecal valve. Twenty cm proximal to the ileocecal valve there was an area of small bowel to small bowel intussusception noted. This segment of small bowel immediately proximal to the intussusception was dilated and appeared compromised as can be seen in Fig. 3. Despite great efforts the small bowel could not be manually reduced and the intussuscepiens had to be sharply incised. This revealed an ischemic intussusceptum with a lead point that was an MD that had itself turned into the main intestinal lumen. The affected segment was resected and deemed sufficiently far from the cecum that ileocecectomy was not required. The bowel was re-anastomosed in side-to-side, functional end-to-end fashion and the patient was then closed. The patient did well post-operatively, and after a short period of bowel rest her diet was advanced. She was discharged home on POD 44, following removal of her external fixator.

2. Discussion

The above patient with CBE underwent cystectomy and cutaneous ureterostomies and developed symptoms consistent with persistent post-operative ileus or small bowel obstruction. After surgical exploration, this was found to be due to intussusception with MD as the lead point.

Embryologically, MD occurs due to incomplete obliteration of the vitelline duct in the eight week of development.[7] Although
not fully understood, CBE is thought to be due to a failure of the cloacal membrane to fully develop in the fourth week of development [8]. Therefore, the embryologic underpinnings of CBE and MD are likely distinct and it does not appear that MD is related to or co-occurs with other congenital anomalies. However, it is possible that these anomalies may cosegregate for reasons not yet understood, which would put CBE patients at higher risk for MD related complications.

Intussusception in the pediatric population most frequently occurs before age two and has a male to female ratio of 2:1 [9,10]. A lead point is found in less than 10% of intussusception cases, adding to the unique nature of this case [11]. Postoperative intussusception is rare, but has been reported following a variety of procedures in pediatric patients [12]. It occurs in approximately 0.01–0.25% of pediatric patients undergoing laparotomy, and is the source of 5–10% of postoperative intestinal obstructions [13–15]. The etiology of postoperative intussusception is not well understood, but may involve altered gastrointestinal peristalsis, bowel manipulation, electrolyte imbalances, anesthetics and opioid analgesics [16]. It is unclear if any alterations in intra or postoperative care could have improved the course in the current case. While most postoperative patients that experience a slow return of bowel function will ultimately be due to a common etiology, rare causes including intussusception should always be considered.

3. Conclusion

CBE is a rare disorder with associated congenital anomalies in other organ systems. MD is a less rare anomaly and intussusception is a common related complication. The above patient with CBE underwent cystectomy with cutaneous ureterostomies, and developed a postoperative intussusception with MD identified as the lead point. This was successfully treated with reduction and small bowel resection. To our knowledge, this is the first report of intussusception due to MD in a patient with CBE.

References