Inflammation and infection

Cecal volvulus with gangrene following Mitrofanoff procedure

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Introduction

The appendico-vesicostomy (Mitrofanoff procedure) is a procedure for continent urinary tract reconstruction usually in children and young adults.\textsuperscript{1} This conduit originally fashioned from the appendix is easily catheterized through an abdominal wall stoma. It is most commonly indicated for neurogenic bladder but has also been performed for ex-strophy and epispadias. The most frequent complications are stomal stenosis and leakage which sometimes require revisional procedures. Bowel obstruction is rare following this procedure.\textsuperscript{2} To our knowledge there are no reports of cecal volvulus following the Mitrofanoff procedure. We report on a case of cecal volvulus which progressed to frank gangrene with perforation, peritonitis and sepsis.

Case presentation

The patient is a 20-year old female paraplegic following a car accident and a neurogenic bladder. She had undergone an uneventful laparoscopic Mitrofanoff procedure at another institution at age16 years. She presented to a community hospital with a one-day history of severe right sided abdominal pain, nausea and vomiting with a presumptive diagnosis of bowel obstruction. She was transferred to our institution for tertiary management. On arrival she appeared gravely ill, with blood pressure 69/47 mmHg, a pulse of 168 and a serum base deficit of 10 mmol/L. Her abdomen was very distended, tympanitic and exquisitely tender on the right side. A plain abdominal X-ray showed a significantly distended cecum in the right upper quadrant (Fig. 1). Computed tomography (CT) revealed swirling adjacent to the cecum and dilated small bowel (Fig. 2). She was initially treated with nasogastric decompression, fluid resuscitation and broad-spectrum antibiotics. Despite adequate resuscitation, she had progressive abdominal pain and developed diffuse peritonitis with worsening sepsis. A decision was therefore made to explore her.

At exploration, the patient had fecal peritonitis from a gangrenous and perforated cecum (Fig. 3). The appendiceal conduit was also frankly ischemic. The cecum and ascending colon were very mobile from prior mobilization from the Mitrofanoff procedure. A right hemicolectomy was performed, and a second look operation scheduled to evaluate for residual ischemia prior to bowel reconstruction. She returned 48 hours later and with no evidence of ischemia, an ileo-transverse colon anastomosis was performed. She had an uneventful postoperative course and discharged. She returned to clean intermittent catheterization with plans for another continent reconstruction via an alternative conduit.

Discussion

Clean intermittent catheterization via the urethra (CIC) popularized by Lapides in 1972 is the preferred initial method of drainage for the neurogenic bladder. Limitations such as obesity, cognitive impairment, altered dexterity, wheel chair dependence, restrictions from orthopedic appliances and braces may make CIC via the native urethra difficult. In 1980, the Mitrofanoff continent cystostomy was first reported as a novel procedure providing a reliable, convenient continent catheterizable conduit. Originally described using the appendix, other suitable conduits have been developed using distal ureter, fallopian tube and tapered ileum.

The initial part of the Mitrofanoff procedure involves mobilization of the cecum and proximal ascending colon from their retroperitoneal attachments to allow the appendix to be sufficiently rotated medially. This reduces stretching of the appendicular blood supply and allows for a tension-free anastomosis to the bladder and the umbilicus.

The most frequent complication resulting from continent conduits is stomal stenosis which limits catheterization in up to 24% of cases.\textsuperscript{3} Others include stomal leakage, prolapse and urolithiasis. In a 20-year follow up review of 23 cases of patients who underwent the Mitrofanoff procedure, five patients had developed a bowel obstruction. There were 2 cases of adhesive obstruction and 3 cases of small intestinal volvulus around the vascular pedicle of ileum used for the bladder augmentation.\textsuperscript{3}

Cecal volvulus was first described in 1837 by Karl von Rokitansky. It is a twist of the cecum and proximal ascending colon. This is a surgical
emergency because untreated it could lead to a closed loop obstruction, ischemia, bowel perforation, sepsis and death. A predisposing factor for its occurrence is a cecal mesentery that is not appropriately adherent to the retroperitoneum, which then allows it to twist. This may occur inherently from congenital abnormality or arise secondary to abdominal operations. Other risk factors for cecal volvulus include constipation, pregnancy, weakness of the abdominal wall and urinary tract infections predisposing to ileus. Bowel obstruction from adhesive bands, although rare, have been described after Mitrofanoff procedure but no case of cecal volvulus has ever been reported.

The clinical presentation of a cecal volvulus is usually that of lower abdominal pain, distension and obstipation with abdominal distension and tenderness. Diagnosis may be difficult clinically, and imaging is often required. On plain abdominal X-rays, cecal dilatation, a single large air-fluid level in the right lower quadrant and paucity of colonic gas characterize the diagnosis. Previously, barium enema was the standard imaging modality with the diagnosis based on the “beak sign” Currently, computed tomography (CT) of the abdomen and pelvis is preferred modality. A 'whirl sign’ is typical, representing spiraling of the cecum around the accompanying vasculature.

If the bowel is viable, simple detorsion with or without cecopexy is acceptable although associated with higher recurrence rates. Cecostomy to create adherence of the cecum to the abdominal wall appears to carry more morbidity and is no longer popular. Right hemicolecctomy may be the preferred option even when the bowel is viable because there is no risk of recurrence and current morbidity rates are low, but this remains controversial. Gangrenous cecum is effectively treated with right hemicolecctomy with or without primary anastomosis.

In our patient’s case, the volvulus was believed to have resulted from significant mobilization of the cecum and ascending colon at the time of her original Mitrofanoff procedure. A strategy to reduce the risk of this complication is to ensure precise mobilization of the cecum and ascending colon allowing just enough redundancy to preserve the appendicular blood supply.

Conclusion

A high index of suspicion is required in patients with the Mitrofanoff procedure presenting with signs of bowel obstruction. Early detection may allow timely detorsion and cecopexy of the redundant cecum with preservation of the conduit. Early surgical intervention is the key to a favorable outcome. Alternative conduits are available if continent catheterizable reconstruction is still desired.

Declarations of interest

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Fig. 3. Gangrenous cecum and ascending colon with mildly ischemic terminal ileum.