Intussusception of bypassed ileal segment following ileal exclusion for progressive familial intrahepatic cholestasis

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Abstract

Progressive familial intrahepatic cholestasis (PFIC) is a rare cause of hepatocellular cholestasis. The disease is typically refractory to medical management and often requires surgical intervention for long-term management. Accepted surgical therapies include biliary diversion, ileal bypass and ultimately orthotopic liver transplantation. Ileal exclusion is felt to be an attractive alternative as it avoids the cutaneous ostomy required with external biliary diversion. We describe a case of ileocolic intussusception following ileal exclusion for PFIC presenting as abdominal pain and hyperbilirubinemia. After reduction of the intussusceptum, the terminal ileum was resected and the ileocolostomy was taken down with re-establishment of bowel continuity. The patient had immediate correction of her hyperbilirubinemia and pruritus post-operatively. This case report describes a unique complication of ileal diversion that has not been previously reported.

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Progressive focal intrahepatic cholestasis (PFIC) is a rare cause of intrahepatic cholestasis refractory to medical management. Surgical therapy includes partial external biliary diversion; however, this approach is associated with usual ostomy complications as well as concerns with cosmesis in the adolescent population. Alternatives include internal ileal exclusion which obviates the need for an external ostomy. While an attractive alternative, complications from this approach remain which surgeons must be aware of. We describe a case of intussusception following ileal exclusion, a previously unreported complication of surgical therapy for PFIC.

1. Case report

A 19-year-old female of Inuit descent was followed at our facility for progressive focal intrahepatic cholestasis (PFIC). She was diagnosed with PFIC as an infant and underwent cholecystojejunostomy for external diversion at 10 months of age. Except for occasional episodes of jaundice, her bilirubin levels were within the normal range with only mild elevations in her serum transaminases. She had no pruritus, pain or clinical jaundice since her early external drainage although she did require a revision of her jejunostomy due to prolapse as a teenager. Liver biopsies showed stable mild periportal fibrosis and bridging fibrosis. She requested closure of her external ostomy and creation of a new internal biliary diversion due to cosmetic and social considerations.

An excision of the jejunostomy with cholecystectomy and ileal exclusion was performed. A length of 100 cm (15%) of small bowel was measured from the ileocecal valve and removed from continuity. An ileocecostomy was performed to bypass this segment. The patient initially did well postoperatively, but within several days she developed pruritus, jaundice and abdominal pain with a progressive conjugated hyperbilirubinemia to 28.0 mg/dL (Fig. 1). Serial ultrasounds revealed no evidence of intrahepatic ductal dilatation or abnormal fluid collection. A percutaneous transhepatic biliary drain was placed with no improvement of her pruritus or jaundice. Minimal biliary drainage was obtained from the drain despite several attempts at replacement. An attempt an external biliary drain placement via laparotomy was aborted due to the small diameter of the common bile duct on intra-operative imaging. A percutaneous transhepatic biliary drain was placed with no improvement of her pruritus or jaundice. Minimal biliary drainage was obtained from the drain despite several attempts at replacement. An attempt an external biliary drain placement via laparotomy was aborted due to the small diameter of the common bile duct on intra-operative imaging.

After resolution of the intussusception, the terminal ileum was resected and the ileocolostomy was taken down with re-establishment of bowel continuity. The patient had immediate correction of her hyperbilirubinemia and pruritus post-operatively. This case report describes a unique complication of ileal diversion that has not been previously reported.
and she was discharged eight weeks following the initial ileal exclusion.

The patient represented with worsening abdominal pain on the day following discharge and she was evaluated with an abdominal CT scan. This revealed an intussusception of the excluded portion of the terminal ileum into her cecum and extending to the mid-transverse colon (Fig. 2). On exploration of the abdomen, the intussusception appeared chronic and was difficult to reduce. Upon reduction it was evident that a large portion of the intussusceptum was ischemic, and there were multiple perforations along its length. No pathologic lead point was identified. The ischemic portion was resected leaving 10 cm of intact native terminal ileum. The ileocolostomy was taken down and an ileal–ileal anastomosis was performed thereby leaving her without a bypassed segment. Following this procedure, her jaundice, pruritus and abdominal pain quickly resolved. Her conjugated bilirubin decreased from 12.0 mg/dL at time of emergent re-operation to 1.5 mg/dL by postoperative day 12. She has done well with no recurrence of symptoms now seven months after discharge.

2. Discussion

Progressive familial intrahepatic cholestasis was initially described by Clayton et al., in 1965 after recognizing this in the Amish family of Joseph Byler [1]. To date, medical treatment has shown limited benefit in preventing the ultimate progression of PFIC. Liver transplantation remains the only curative option in the management of PFIC [1]. The associated morbidity from transplantation and ongoing shortage of organ donors led to the development of alternative surgical options that would at least delay the onset of ESLD. In 1988, Whittington and Whittington first described a non-transplant option for the management of PFIC using partial external biliary diversion (PEBD) through creation of a permanent stoma by cholecystojejunostomy [2]. Retrospective studies have shown excellent results in patients treated with PEBD; clinical and biochemical improvements were seen in 75% of patients and partial resolution in an additional 12.5% [3]. The remaining patients had progression of their disease, although these patients all had evidence of cirrhosis on liver biopsy performed at the time of diversion. Despite the excellent results seen with PEBD, it is associated with typical stoma complications including prolapse, biliary reflux, dehydration, parastomal hernia and concerns for social stigmatism [4].

Ileal exclusion has become a viable surgical option in the treatment of PFIC when PEBD cannot be performed [5]. In this operation, 15% of the small bowel is measured proximal to the ileocecal valve and excluded with an ileocolostomy. The excluded terminal ileum is classically left in situ and can be placed back in continuity if liver transplantation is needed. This approach has demonstrated early resolution of symptoms and laboratory abnormalities while avoiding the complications of a stoma [5]. A recently published review of the literature analyzed 11 studies with a total of 77 patients and found that patients had successful outcomes from both PEBD and ileal exclusion. Treatment failures were usually due to advanced liver disease prior to surgical intervention [6]. Only one study retrospectively compares PEBD to ileal bypass. The data demonstrate that ileal bypass, although initially effective, did not confer a long-term benefit [7].

Other surgical options including partial internal biliary diversion have been proposed but long term studies are lacking [8,9]. PIBD involves the internal diversion of bile flow into the colon, thereby avoiding the terminal ileum in an attempt to prevent reabsorption of bile salts in the enterohepatic circulation. This has been described using an excluded segment of jejunum or colon to create a cholecystoenterocolostomy [8,9]. These methods accomplish the goals of PEBD without an external ostomy. Despite excellent short-term results, the direct effect of bile salts on the colonic mucosa and the potential for biliary reflux into the terminal ileum have yet to be studied. Additionally, many patients who suffer from PFIC have had previous bowel resections, and further intestinal resection to create an internal bypass may lead to malabsorption and nutritional deficiency.

We report a case of ileocolic intussusception of the excluded ileal segment following ileal bypass for PFIC. The intussusception was the likely source of the patient’s persistent hyperbilirubinemia and abdominal pain after ileal exclusion. The mucosa of the intussusceptum was positioned in the lumen of the ascending colon and was exposed to its luminal contents. Thus, bile salt absorption through the terminal ileal mucosa continued to occur. Effectively, the terminal ileum was no longer bypassed. After manual reduction of the intussusception, the patient’s abdominal pain was ameliorated, and she had a rapid normalization of her hyperbilirubinemia. To our knowledge, this is the first reported case of this unique complication from ileal exclusion for PFIC. The diagnosis of intussusception should be considered in patients who present with recurrent hyperbilirubinemia and abdominal pain after ileal exclusion. Radiographic
imaging for patients with refractory pain, CT scan in particular, should be obtained. In our case, it is possible that the diagnosis of intussusception may have been made earlier in the post-operative course had a CT scan been performed.

While this complication is most likely rare, resection of the bypassed ileum may be an option to prevent post-operative intussusception after ileal exclusion as seen in this case. However, ileal exclusion patients who ultimately receive a liver transplant may subsequently have the excluded ileal segment restored into continuity thereby minimizing long-term risk of vitamin and nutrient deficiency. In addition, resection of the terminal ileum may have untoward effects related to gastrointestinal hormone production which may remain active despite this segment being excluded from the fecal stream.

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References