CASE REPORT

Intestinal obstruction secondary to adhesions in an infant with cystic fibrosis

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SUMMARY

We report the case of a 9-week-old boy with cystic fibrosis (CF) who presented with intestinal obstruction. He was initially diagnosed with distal intestinal obstruction syndrome (DIOS) and started on laxatives and gastrograffin. This conservative management was unsuccessful and he therefore underwent an exploratory laparotomy. Despite not having undergone any previous surgery, extensive intra-abdominal adhesions were identified as the cause of the intestinal obstruction. The only previous abdominal pathology occurred in the neonatal period prior to his diagnosis of CF, when he was managed conservatively for possible necrotising enterocolitis. We hypothesise that this episode was associated with a subclinical bowel perforation that led to the formation of intra-abdominal adhesions. Although DIOS is the commonest cause of intestinal obstruction in children with CF, this case highlights that surgical causes should always be considered. It also demonstrates that the intra-abdominal adhesions can occur in the absence of previous surgery.

BACKGROUND

Intestinal obstructions are relatively common in patients with cystic fibrosis (CF), the cause varying according to the patient's age. Meconium ileus (MI) is unique to CF and is characterised by complete intestinal obstruction in the neonatal period caused by the accumulation of inspissated meconium. In total, 13–17% of all patients with CF present with MI, the majority requiring surgery.¹ Constipation and distal intestinal obstruction syndrome (DIOS) are the main causes of intestinal obstruction in older children and adults with CF. DIOS is an acute, complete or incomplete, faecal obstruction in the ileocaecum, whereas constipation is defined as gradual faecal impaction of the total colon.² The prevalence of DIOS in children with CF is 7–8%, the lifetime prevalence rising to 14–16%. The prevalence of constipation in childhood and the lifetime prevalence are both around 40%.¹ Both DIOS and constipation can usually be successfully managed with an intensive regimen of laxatives. Colonic strictures causing intestinal obstruction have been reported in patients with CF, possibly related to high doses of pancreatic enzyme replacement.³

We present the case of an infant with CF and intestinal obstruction caused by intra-abdominal adhesions. Adhesions are bands of fibrous tissues that form between structures in the intra-abdominal cavity as a result of inflammation, injury or surgery. The patient had not undergone any previous

surgery and we hypothesise that the adhesions were caused by a subclinical bowel perforation in the neonatal period.

CASE PRESENTATION

A 9-week-old boy with CF presented to the emergency department with a 24 h history of non-bilious vomiting and fever. His oral intake was poor but he continued to pass urine and open his bowels normally. Full systemic examination was unremarkable apart from mild dehydration. A diagnosis of viral gastritis was made and he was admitted to the paediatric ward for observation. Over the next 48 h he continued to vomit and developed increasing abdominal distension and constipation. He became tachycardic, tachypnoeic and his skin appeared mottled. Examination of his chest revealed subcostal recession and tracheal tug but his chest was clear. His abdominal distension was resonant to percussion. Bowel sounds were present. An abdominal radiograph (figure 1A) showed massively distended gas-filled loops of small bowel, consistent with obstruction.

A presumed diagnosis of DIOS was made and he was started on gastrograffin via nasogastric tube and enema. The respiratory distress was thought to be caused by the distended abdomen compressing the lower lobes but intravenous cefuroxime was started to cover possible lower respiratory tract infection. The conservative management of his intestinal obstruction was unsuccessful as he did not open his bowels and the abdominal distension and vomiting continued. He was reviewed by the surgical team and underwent an exploratory laparotomy. During this procedure extensive intra-abdominal adhesions were identified as the cause of the intestinal obstruction. Four segments of the ileum (total 17.2 cm) were resected and end-to-end anastomoses made. A broviac line was inserted and total parental nutrition (TPN) was started in the postoperative period. He made a good postoperative recovery and by day 14 he was fully enterally fed and the TPN was therefore discontinued. He has had no further episodes of abdominal distension although his subsequent weight gain has been slow.

The cause of the patient's adhesions is unclear. He had not undergone previous surgery but did have a history of possible abdominal pathology in the neonatal period. He had been born in good condition at 35 weeks gestation but developed respiratory distress, tachycardia and abdominal distension at 18 h of age. An abdominal radiograph (figure 1B) showed some non-specific gas filled,

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Figure 1 (A) An abdominal radiograph at 9 weeks of age showing massively distended gas filled loops of the small bowel consistent with obstruction. (B) An abdominal radiograph taken at 18 h of age showing non-specific gas-filled, mildly distended loops of bowel. Arrow shows a loop of bowel with a soap bubble appearance most commonly seen in meconium ileus.

mildly distended loops of the bowel. He had not passed meconium at this point. A diagnosis of possible necrotising enterocolitis (NEC) was made which was managed conservatively with intravenous antibiotics, intravenous fluid and subsequently total parental nutrition. Over the next 10 days his abdominal signs and symptoms resolved. Enteral feeding was established and he began to open his bowels normally. At 15 days of age he was diagnosed with CF by newborn screening.

DISCUSSION

We hypothesise that the adhesions causing this patient's intestinal obstruction were secondary to a subclinical bowel perforation in the neonatal period. This perforation may have been due to NEC in keeping with his initial diagnosis. However, it is also possible that it was caused by MI that resolved spontaneously, helped by the intravenous fluids. The original X-ray showed gas filled, distended loops of bowel and a loop in the left upper quadrant demonstrating a soap bubble appearance (see arrow in figure 1B). This is seen when gas is mixed with meconium. It is likely that if the diagnosis of CF had been known at this early stage MI would have been considered in the differential diagnosis. Unfortunately this presentation occurred before the dried blood spot was collected for newborn CF screening, which occurs in the UK on day 5.

Approximately half the patients with MI are defined as 'uncomplicated' where the intestinal obstruction is caused by the accumulation of inspissated meconium in the terminal ileum and proximal colon. The remainder are defined as 'complicated' where the intestinal obstruction is caused by meconium-laden loops of bowel twisting and becoming ischaemic causing volvulus, perforation or atresia.⁴ MI may present with various symptoms. Treatment options for MI remain challenging. A recent review⁴ has shown that conservative management of neonatal MI with a gastrografin enema has a success rate of 36–39% but can cause bowel perforation. Surgical management with primary anastomosis results in a higher complication rate than delayed anastomosis. Despite such invasive management, patients with MI at birth appear to have similar clinical outcomes later in life in comparison to patients with CF without MI.¹

This case highlights the multiple causes of intestinal obstruction in patients with CF and the need for clinicians to consider

a surgical cause. It is also important to remember that the cause of intra-abdominal adhesions may not be clear and can occur in the absence of previous surgery.

Learning points

- ▶ Intestinal obstruction is relatively common in patients with cystic fibrosis (CF) and the cause varies according to the patient's age.
- ▶ Meconium ileus presents before the CF newborn screening results are available.
- ▶ Although distal intestinal obstruction syndrome and constipation are the commonest causes of intestinal obstruction in children and adults with CF, surgical causes should always be considered.
- ▶ The cause of intra-abdominal adhesions may not always be clear and can occur in the absence of previous surgery

Contributors MJ was working with the CF team at the time of the presentation. GK reported the X-rays and wrote the first draft of the case report. FJG and WL were involved in editing the article. All authors have seen and approved the final version.

Competing interests None.

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