Case Report

Distal intestinal obstruction syndrome and mechanical small bowel obstruction : a rare but life-threatening combination in cystic fibrosis patients

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Keywords

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Abstract

This case report illustrates a distal intestinal obstruction syndrome in a child with cystic fibrosis, complicated by an adhesive small bowel obstruction. Both conditions can occur simultaneously in cystic fibrosis patients.

Early recognition and appropriate multidiciplinary medical management are essential to avoid complications.

Introduction

Distal intestinal obstruction syndrome (DIOS) is a common complication associated with cystic fibrosis (CF). The prevalence is estimated around 5 to 12 episodes per 1000 patients per year in children (1). DIOS results from an accumulation of viscous faecal material with sticky mucous secretions in the bowel lumen that adheres to the intestinal wall of the terminal ileum and caecum, and was first described by Jensen in 1962 (1-3).

In 2010, a consensus gave rise to a definition of the syndrome with a distinction between the incomplete and complete form (4). The incomplete form is characterised by history of abdominal pain and the presence of a subostructive faecal mass in the right colic quadrant. The complete form is more severe, with signs of complete intestinal obstruction such as bilious vomiting and secondary mechanical ileus with hydro-aeric distension of the intestines visible on plain radiographs.

Most cases resolve with medical treatment. However, in case of a lack of improvement despite conservative management, an alternative diagnosis should be considered, such as an adhesive small bowel obstruction. We describe the occurrence of both DIOS and mechanical small bowel obstruction due to adhesions in a child with CF and a history of abdominal surgery in the neonatal period.

Case report

A 9-year-old boy with CF, genotype F508del/R1162X, with a pancreatic insufficiency and a history of meconium ileus leading to bowel resection at birth, was admitted at the emergency department for abdominal pain and gastric fluid vomiting. Abdominal radiographs revealed hydro-aeric levels with stercoral stasis. The abdominal ultrasound did not show any sign of bowel ischaemia. Blood tests was normal apart from hyponatremia at 127 mmol/L (normal value: 135-145mmol/L). Initial management consisted of saline enemas and oral macrogol administration. The hyponatremia was corrected through intravenous fluid therapy. The patient was transferred to a CF reference centre 72 hours after the initial management.

On admission, the clinical examination revealed a tense and bloated abdomen. The bowel sounds were weak and the abdominal palpation was painful. Blood tests were normal. Management consisted in placing a nasogastric tube and the administration of gastrografin enemas, without success. Barium enema showed faecal impaction in the right lower quadrant with small intestinal distension. 48 hours after, due to persistent abdominal pain, bile stained gastric residuals, increase of inflammatory markers and ascites with evidence of small bowel obstruction on ultrasound (Figure 1), surgical intervention by laparotomy was indicated. A significant intestinal dilatation proximal to an ileal stenosis with numerous adhesions and bowel impaction was found. A complete adhesiolysis and a 10 cm necrotic ileal resection were performed. There were no complications in the immediate post-operative period and a residue-free diet was progressively introduced. 72 hours after refeeding, signs of obstruction reappeared. A re-intervention was performed and consisted of a 15 cm resection of intestinal stenosis (due to bowel impaction) and ileostomy. After re-intervention, refeeding was well tolerated. Due to high stoma output and salt loss, electrolyte replacement was necessary, first with parenteral administration and then with enteral feeding. The patient left the hospital after 6 weeks. The ileostomy was closed 3 months after the surgery, following imaging assessment (radiography and barium enema) confirming the recovery of a normal intestinal calibre upstream and good permeability downstream of the ileostomy.

Discussion

DIOS can occur at any age, but is most common after the age of 15, with a peak in young adults of 20-25 years old (3). The principal differential diagnosis of DIOS is constipation, also common in patients with cystic fibrosis.

The diagnosis is often clinical. Sometimes, an abdominal radiograph may help to show the faecal mass in the bowel lumen with multiple air-fluid levels in the dilated small bowel (1-3). Risk factors include cystic fibrosis genotype with non-functional CFTR-protein (cystic fibrosis transmembrane conductance regulator protein), pancreatic insufficiency, a history of meconium ileus at birth as this could share the same pathophysiology, irregular pancreatic enzyme intake and dehydration or dietary changes (3). After lung transplant, the incidence is also higher and this may be due to a post-operative ileus and use of analgesics (like opioids) (1).

The management is often symptomatic and conservative (91% of episodes of non-

complicated of DIOS are successfully treated) (3). In a case series of 80 patients a surgical treatment was required in less of 3.9% (3). In mild or moderate DIOS, a combination of orally administered laxatives and osmotic laxative enemas are recommended (3). A nasogastric tube may be required to give large volumes of oral laxatives or to decompress the abdomen in cases of complete intestinal obstruction. Prokinetics have been suggested, but there is a lack of evidence to use them in cases of DIOS (3). In rare cases, when oral treatment fails, a gastrografin enema can be used to clear the impacted faecal mass in the terminal ileum (3). This procedure must be performed by an experienced radiologist to minimize the related complications such as intestinal perforation, shock or necrotising enterocolitis (1).

In our case, surgical treatment was required because conservative treatment failed.

DIOS probably worsened a chronic small bowel subocclusion in an abdomen full of adhesions. In this case, the adhesions around the neonatal surgery probably contributed to the concomitant involvement of digestive stenosis. We can also imagine that there were previous incidents of repetitive intussusceptions which may have contributed to acute incident.

If conservative management fails, a diagnosis of mechanical small bowel obstruction due to adhesions must be considered. According to Subhi et al., the risk of surgical intervention in patients with a history of meconium ileus was statistically significant, maybe due to previous laparotomy (5). Surgery for DIOS is rare in the paediatric population and may be necessary if oral treatment fails or if there are complications such as intussusception or volvulus (3-6). In severe cases, a surgical procedure is required with laparotomy and intestinal lavage often without ileostomy (3). In some cases, bowel resection with stoma is required to allow intestinal recovery (3). In all cases, the patient must be referred to a CF centre with a multidisciplinary team. Early recognition and appropriate medical management are necessary to avoid complications (such as intussusception) or surgical treatment (3).

Frequent in acute situation among CF patients and especially in case of digestive problems, hyponatremia may contribute to create a third space which helps to underline the gravity of the situation. Further investigations are required to quickly reach the diagnosis.

There is a lack of research analysing options to prevent DIOS in CF patients (3). It is important to follow these patients in CF centres, to monitor their feeding habits to achieve good hydration status, and to supplement pancreatic insufficient patients with fat-soluble vitamins and pancreatic enzymes. Early detection of signs of constipation and prevention of possible faecal impaction are essential in CF patients.

Conclusion

This case report highlights that DIOS, a classical gastrointestinal complication in cystic fibrosis, and adhesive small bowel obstruction can occur concurrently, making the diagnosis more difficult. Early recognition and appropriate multidiciplinary medical management are essential to avoid complications. The purpose of this case report was to demonstrate that clinical deterioration despite medical treatment must raise the suspicion of another diagnosis such as an obstruction due to adhesions.

Conflict of interest statement: The authors have declared that no conflict of interest exists.

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Figure 1: *Ultrasound of DIOS described by an impaction of faecal mass with bowel distension complicated by a small bowel obstruction described by a bridle imprint*