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Case Report

Intestinal obstruction and cystic fibrosis: a case report

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ABSTRACT

Distal intestinal obstruction syndrome is one of the frequent digestive complications of cystic fibrosis; due to a more or less complete small bowel obstruction. Proximal intestinal obstruction syndrome is a rarer variant, but has been more often described since the improvement of the management of CF that has led to enhance CF patient survival. We described a 19-year-old male followed in the pediatrics department of the HMIMV for cystic fibrosis complicated by exocrine pancreatic insufficiency and type 1 diabetes, who presented an intestinal obstruction syndrome for two days, consisting on abdominal pain, bloating, bilious vomiting and cessation of stool and gas. Imaging has suspected a mechanical obstruction of the small bowel however no transitional zone could be identified. A conservative treatment was start and because of the inefficiency of this first approach, the decision was made to proceed with a laparotomy. Intra-operatively, a mid-jejunum point with inssipationed stool was noticed to be the transitional aera of the obstruction and conducted us to proceed for an entrotomy and digitaly empty the proximal bowell. PIOS is a more rare intestinal complication but should be considered when the ileo cecal clinical location is absent in an obstruction syndrome. Its treatment is based on laxatives however surgery should wisely be considered in front of failure of the conservative approach or critical condition.

Keywords: Distal intestinal obstruction syndrome, Case report, PIOS

INTRODUCTION

Cystic fibrosis (CF) is an inherited disease caused by mutation of the CFTR (a) gene. It's a life-limiting disease that affects all the exocrine glands (sweat glands, pancreas, etc.) of the body.[1]
The improvement in the management of CF, once the prerogative of pediatric services, has meant that other medical services are facing fragile CF adults with complications from other systems, in particular the gastro intestinal tract.[2] One of the frequent digestive complications of cystic fibrosis is distal intestinal obstruction syndrome which produces a more or less complete intestinal obstruction.[3,4]

Only few cases of proximal intestinal obstruction were described.

We report the case of a proximal intestinal obstruction in a young patient followed for cystic fibrosis since his young age. The aim is to share our unique experience in the management of this rare complication.

CASE REPORT

A 19-year-old male followed since the age of 5 in the pediatrics department of the HMIMV for cystic fibrosis complicated by exocrine pancreatic insufficiency and type 1 diabetes.

During its hospitalization for pulmonary exacerbation of cystic fibrosis, the patient presented an intestinal obstruction syndrome, consisting on abdominal pain, bloating, bilious vomiting and cessation of stool and gas.

Biological findings (leukocytes, lipase) were normal.

Abdominal CT showed a distended aspect of the jejunal loops with air fluid levels, and doubted about a transitional zone and collapsed distal ileum. The CT scan also revealed hypo-dense material containing air bubbles in the transitional bowel area, a dilated appendix containing intraluminal dense material and a large peritoneal effusion (Figure 1).

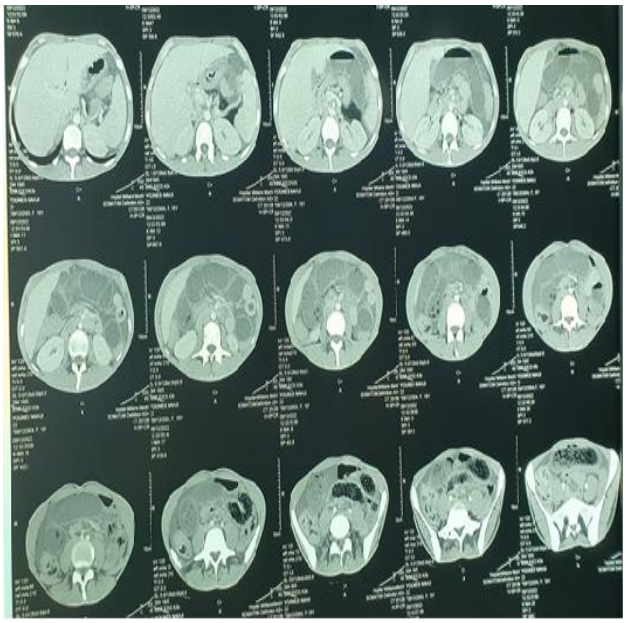

Figure 1: CT scan picture shows dilated small bowel and air fluid levels, fesses sign.

Conservative treatment involving nasogastric tube, electrolytes replacement, rehydration and oral osmotic aperient, was started but with no clinical improvement after 48 hours. Therefore; consent was obtained for surgery.

On a midline laparotomy, the exploration found an inflamed appendix filled with mucoid materiel (Figure 2a), a very abundant peritoneal effusion and a dilated suffering first jejunal loops among an intraluminal material followed an empty collapsed remaining small bowel. The colon was flat and free.

The procedure consisted on a jejunal enterotomy in a non-suffering zone that permitted to evacuate by digital expression, 200 cc of dry and solid mucus and stool, simple nots of 3/0 (Figure 2b) PDS were used for enterotomy closure, and then an appendectomy was performed. The post-operative courses were simples and hospitalization was only maintained for respiratory criteria. 15 days after the operation, the patient reinstalled another occlusive episode, which responds to gastrographin ingestion and osmotic laxatives.

The resumption of the intestinal transit was effective 6 months after the procedure. The histological study showed a congestive and edematous appendix with no sign of malignancy and the extracted material consisted on a mucofecal composite.

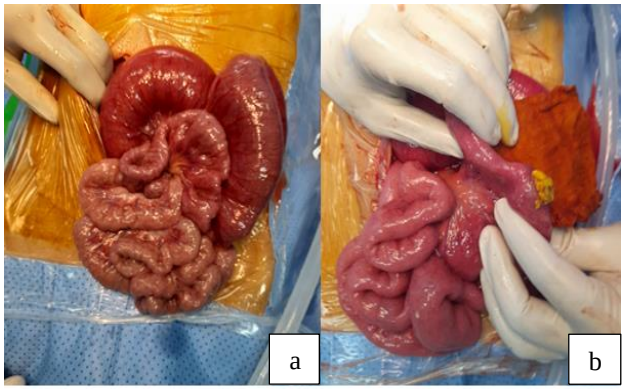

Figure 2: Operative view showing the proximal occlusion (a) dilated proximal loops and hyperemia, normal empty distal small bowel; and (b) digital expression of the insisipated material through the enterotomy.

DISCUSSION

Cystic fibrosis is an inherited disease caused by mutation of the CFTR (a) gene. This mutation is responsible for the dehydration and hyperviscosity of the surface fluid lining the bronchial epithelium, resulting in poor mucociliary clearance.

All of the exocrine glands of the body are affected.[1] Almost all the morbidity and mortality in CF patient is secondary to respiratory disease. The improvement in the management of these complications, once the prerogative of pediatric services, has meant that we face nowadays fragile CF adults with complications from other systems, in particular digestive.[2,4]

Proximal intestinal obstruction syndrome is a rare entity and only few cases were described. Intestinal obstruction in CF is secondary to a fluid and chloride (secretion/absorption) dysregulation, combined to multi-factorial motility abnormalities.[5,6] The crossing of the anatomical barrier, the ileocecal junction; by thick stool explains the higher frequency of distal intestinal obstruction compared to a more proximal obstruction.

The proximal intestinal obstruction syndrome PIOS presents symptoms of complete or impending small bowel obstruction; acute abdominal pain and bloating, vomiting of bilious material. The abdominal pain has no specific location and no mass can be found compared to the DIOS where the pain is often located in the right lower quadrant and where a mass is more usually to be found.[7] Abdominal examination should seek for peritoneal signs that can reveal an intestinal necrosis and constitute a formal surgical indication. The prevalence of PIOS is poorly known, only few cases were published, in the other hand,

DIOS is estimated to be at 10-20% of all CF patients regardless of age and sex. Inappropriate fluid intake or supplementations of pancreatic enzymes are the most frequent factors leading to occlusion in CF patients.[8] Abdominal X-rays shows fecal loading and small bowel air-fluid levels in the setting of a complete obstruction. CT scan of the abdomen demonstrates proximal small bowel dilatation with inspissated fecal material in the bowel.[9] Appendicitis, appendicular mucocele or complete occlusion must be ruled out. The treatment of PIOS is above all empirical in the absence of controlled and randomized studies.

As the CF Patients constitute a high risk surgical candidates , treatment start with rehydration if necessary, combined with oral osmotic laxatives containing polyethylene glycol (PEGS) at a dosage of 20-40 ml/kg/h with a maximum of 1 l/h over 8 hours; emptying is then obtained in 2 to 6 hours.[10-12]

Complete resolution of symptoms should be achieved and abdominal X-ray can be used to show resolution of PIOS. The non-response to the medical treatment mandates laparotomy, as well as the development of peritoneal signs. Laparoscopy can be run, with a high risk of conversion to laparotomy for technical difficulties; and it's a useless if a manual disimpaction of the stool into the colon is to attempted.[13] In our case, the long distance to the colon constituted a great risk of inefficiency of disimpaction and small bowel laceration, and lead us to consider enterotomy. Extraction of the impacted materiel through enterotomy is a contaminating procedure that should be well controlled. Prevention is based on maintaining a good state of hydration and optimizing the treatment of exocrine pancreatic insufficiency.

CONCLUSION

PIOS is a more rare intestinal complication and should be evoqued when the ileo cecal clinical location is absent. Treatment is based on laxatives and surgery should wisely be considered in front of critical conditions.

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