CELIAC DISEASE PRESENTING AS ACUTE COLONIC PSEUDO-OBSTRUCTION


Reva MATTA, Elie ARAMOUNI, Pierre MOUAWAD, Nabil DIAB

ABSTRACT: Acute colonic pseudo-obstruction rarely presents in children. Upon diagnosis, initiating conservative management and identification of an underlying etiology is mandatory. We describe the case of an 8-year-old girl who presented with signs of acute abdominal distention with no evident etiology. The diagnosis of celiac disease was suspected and confirmed by intestinal biopsy. Based on this case and other reported cases in the literature, we discuss this unusual mode of presentation of celiac disease, the underlying etiology and suggest management options to avoid unnecessary aggressive interventions.

INTRODUCTION

Celiac disease is a complex autoimmune enteropathy that affects the small bowel in genetically predisposed individuals [1]. Typical symptoms of general malabsorption are failure to thrive, large pungent stools, and a distended abdomen [2]. Celiac may also present in variety of other atypical symptoms but very rarely as acute colonic pseudo-obstruction. Acute colonic pseudo-obstruction (ACPO) also known as Ogilvie’s syndrome is defined as acute colonic dilatation in the absence of obvious colonic disease or mechanical obstruction [3]. ACPO is rarely defined in pediatric population. The purpose of this article is to review the literature describing ACPO in pediatric population and its possible causes, particularly the mode of presentation for celiac disease to avoid unnecessary aggressive interventions.

CASE REPORT

An 8-year-old girl, previously healthy (weight 24 kg, height 125 cm), presented to the emergency room with severe abdominal pain and distention. Six hours prior to presentation the patient suddenly experienced severe, diffuse abdominal pain associated with abdominal distention and obstipation. Physical examination revealed hypoactive bowel sounds, and distended abdomen with diffuse tenderness. Complete blood count, electrolytes, liver function tests, C-reactive protein and erythrocyte sedimentation rate were within normal range. Plain abdominal radiography (Fig. 1a) showed markedly distended sigmoid and rectum, with marked fecal material proximal to the colo-sigmoid junction.

The diagnosis of functional ileus of the rectum and sigmoid was raised. Abdomen and pelvis CT confirmed the findings and showed severe dilatation in the rectum and sigmoid. The patient was initially started on IV hydration, and kept NPO with nasogastric decompression. A rectal tube was inserted to release the colonic distention and the symptoms subsided. Post-decompression, repeat abdominal radiography (Fig. 1b) showed normal gas pattern distribution. Colon contrast enema showed free passage from rectum to cecum without extrinsic compression or stenotic lesions. Anorectal manometric study showed the presence of a recto-anal inhibitory reflex, therefore excluding Hirschsprung’s disease. After resolution of colonic dilatation, a colonoscopy was performed and was of normal macroscopic aspect. The patient was diagnosed to have Ogilvie’s syndrome with no evident underlying etiology and was discharged home with no medical treatment while waiting for the biopsy results.

Four days later, the patient presented again to the emergency room with the same complaints. A rectal tube was placed and again the patient’s symptoms were resolved. Colonic biopsy results showed lymphocytic colitis that...
raised the suspicion of celiac disease. Therefore a gastroscopy was performed revealing marked scalloping of duodenal folds. Anti-endomysium antibodies were positive and anti-human tissue transglutaminase antibodies were remarkably elevated (25.7 > 10 µ/ml). Duodenal biopsies revealed severe villous atrophy with significant intraepithelial lymphocytosis and crypt hyperplasia, confirming the diagnosis of celiac disease (Fig. 2). Patient was started on a gluten free diet and has been symptom free for four years.

DISCUSSION

Celiac disease is a complex autoimmune enteropathy that affects the small bowel in genetically predisposed individuals. It results from an inappropriate T cell-mediated immune response against ingested gluten protein [1]. The characteristic lesion of the small intestinal mucosa includes loss of absorptive villi and infiltration of the lamina propria with inflammatory cells [1]. Typical symptoms are those of general malabsorption: failure to thrive, large pungent stools, and a distended abdomen [2]. Celiac may present in a variety of other atypical symptoms but very exceptionally as acute colonic pseudo-obstruction.

Acute colonic pseudo-obstruction (ACPO), or Ogilvie’s syndrome, is a gastrointestinal motility disorder characterized by marked colonic dilation in the absence of evident underlying causes. It is defined as acute colonic dilatation in the absence of obvious colonic disease or mechanical obstruction [3]. It is an uncommon but potentially fatal condition with significant morbidity and mortality [1-2]. ACPO is seen in the adult population, but exceptionally in children and usually develops in association with serious underlying medical or surgical illness [3-4]. The mortality rate in adult ACPO is 15% with early appropriate care but increases to 36% if the patient progresses to colonic ischemia and perforation [3]. It is of paramount importance to make an early diagnosis to reverse the outcome of this potentially fatal condition, than to search for the underlying etiology. The diagnosis is based on history, physical and radiological examination, and exclusion of other diagnoses. The diagnostic evaluation should exclude mechanical obstruction, toxic megacolon, perforation, and ischemia [3-4]. Conservative management is the initial mode of therapy consisting of bowel rest, nasogastric decompression, fluid and electrolyte correction, and aggressive treatment of any underlying reversible medical conditions [3]. Spontaneous resolution is seen in 85% of cases with a 15% chance of recurrence [4].

Only five cases of ACPO in the pediatric population are reported in the literature [5-7]. All the reported cases had an underlying etiology. Our patient’s presentation is typical to that of Ogilvie. The workup done on our patient excluded any metabolic or septic or underlying surgical cause for her pseudo-obstruction. Further evaluations by

![Abdominal radiography](image1)

**Figure 1.** Abdominal radiography showing (a) markedly distended sigmoid and rectum with marked fecal impaction proximal to colosigmoid junction; (b) normal gas pattern distribution following post decompression.

![Duodenal biopsy (H&E)](image2)

**Figure 2.** Duodenal biopsy (H&E) showing severe villous atrophy with significant intraepithelial lymphocytosis.
gastroscopy and duodenal biopsies were necessary to identify the underlying illness. As soon as conservative management was initiated, the symptoms completely subsided. None of the pharmacological treatments for Ogilvie that are described in the literature [5-7] such as erythromycin, cisapride and neostigmine were necessary in our case.

A similar reported case by Pittschierler et al. [8], describes the case of a 13-month-old boy who presented with an intestinal pseudo-obstruction. A jejunal biopsy showed total villous atrophy. The patient did not relapse with the same symptoms on a gluten challenge. In our case, the patient had a recurrence of her disease after discharge when the underlying etiology was not eliminated. In addition to that, once the diagnosis of celiac disease was confirmed, the patient was started on gluten free diet with no recurrence of her symptoms.

The pathophysiology of Ogilvie is not completely understood, but it is believed that it is due to inhibition of colonic motility by autonomic imbalance either by suppression of sacral parasympathetic nerves or by an increase in sympathetic tone [4]. Koklu et al. [9] reported two cases of patients aged 51 and 39 who were known to have celiac disease presenting for intestinal obstruction. Gluten driven mucosal inflammation caused motor abnormalities by affecting smooth muscle contraction or impaired release of neurotransmitters [9-10]. Furthermore, manometric investigations showed that motility disorders improved or even became normal following a gluten free diet [9-10].

This case offers the strongest argument compatible with clinical and biochemical findings that celiac disease may cause intestinal pseudo-obstruction. It should be considered especially in children and young adults with such presentation, in order to avoid unnecessary medical and surgical interventions. ACPO occurs in patients with an underlying disease. The take home message is to immediately start conservative treatment to avoid unnecessary complications and then to search for and treat the underlying etiology.

ACKNOWLEDGMENTS

Authors declare no conflict of interest and no sources of support requiring acknowledgment or funding declaration.

REFERENCES