Intestinal pseudo-obstruction: adult Hirschsprung’s disease and Ogilvie’s syndrome. Clinical case report

Authors: Ernesta Utakienė1, Vestina Strakšytė1, Irina Gineikienė1, Saulius Švagždys2, Dainius Jančiauskas3, Gediminas Kiudelis4, Algidas Basevičius1, Mantas Vilčinskas5

1 Department of Radiology, Hospital of Lithuanian University of Health Sciences Kauno klinikos, Eiveniu street 2, Kaunas, Lithuania.
2 Department of Surgery, Hospital of Lithuanian University of Health Sciences Kauno klinikos, Eiveniu street 2, Kaunas, Lithuania.
3 Department of Pathology, Hospital of Lithuanian University of Health Sciences Kauno klinikos, Eiveniu street 2, Kaunas, Lithuania.
4 Department of Gastroenterology, Hospital of Lithuanian University of Health Sciences Kauno klinikos, Eiveniu street 2, Kaunas, Lithuania.
5 Hospital of Lithuanian University of Health Sciences Kauno klinikos, Eiveniu street 2, Kaunas, Lithuania.

ABSTRACT
Colonic pseudo-obstruction is a clinical and radiological syndrome of the large bowel dilatation without mechanical obstruction. We report two similar clinical cases of this syndrome with different origins – adult Hirschsprung’s disease and Ogilvie’s syndrome. Both conditions are rare and actual prevalence is unknown. Early recognition and management are extremely important because it can prevent occurrence of such complications as ischemia, volvulus or perforation and reduce morbidity with mortality.

Keywords: hypoganglionosis, computed tomography imaging, acute colonic pseudo-obstruction, megacolon, constipation.

INTRODUCTION
The term intestinal pseudo-obstruction denotes a syndrome characterized by a clinical picture suggestive of mechanical obstruction in the absence of any demonstrable intestinal lumen obstruction [1]. According to presentation, pseudo-obstruction syndrome can be subdivided into acute and chronic forms.
Acute colonic pseudo-obstruction (ACPO) also known as Ogilvie’s syndrome is characterized by massive colonic dilatation in the absence of mechanical obstruction [2]. ACPO is an important cause of morbidity and mortality. Ischemia and perforation are the threatening complications of ACPO. Spontaneous perforation has been reported in 3-15% of cases with a mortality rate estimated at 50 % or higher when this occurs [3].
Chronic intestinal pseudo-obstruction (CIPO) is a syndrome defined by the presence of chronic intestinal dilatation and dysmotility due to various causes [4]. One of them is Hirschsprung’s disease (HD), which is considered a pediatric illness, as the vast majority of cases are diagnosed during the neonatal period [5]. Rare cases of Hirschsprung disease can be present in adulthood. The incidence of adult Hirschsprung’s disease is unknown, mainly because it is frequently overlooked in the adult population. The diagnosis of HD is usually much more difficult in adult than in infants, partly because of the rarity of the disease and the higher incidence of short or ultrashort segment aganglionosis in adults [6].
We should distinguish patients with ACPO from CIPO mainly because of treatment possibilities. However, despite the increasing awareness of these conditions its diagnosis remains difficult and is often delayed [2, 7]. We present two clinical cases of colonic pseudo-obstruction with different origins – Ogilvie’s syndrome and Hirschsprung’s disease.
CASE PRESENTATIONS

CASE 1

A 49-year-old man was admitted to the hospital with suspicious bowel obstruction. The anamnesis was poor because of patient’s speech and hearing disorders. Other serious underlying medical or surgical conditions were unknown. Physical examination findings were massive, rigid abdominal distension, diffuse lower abdominal pain, absence of peristalsis and no clinical signs of peritonitis. Digital rectal examination was negative (rectum empty). Routine laboratory blood test results were within the normal values.

Plain film of the abdomen showed marked diffuse dilatation of the colon (up to 11,2 cm diameter) without air-fluid levels and free air (Fig.1). Abdominal ultrasound examination was uninformative because of gas filled bowel loops. A colonoscopy procedure failed due to retained stools in the rectum. A computed tomography (CT) scan confirmed persistent massive dilatation of the all large bowel (cecum diameter reaching 11,5 cm, rectum - 9,8 cm) filled with faeces and gas without evidence of mechanical obstruction (Fig.2).

The patient’s condition was improving and he was managed conservatively with intravenous fluids and enemas.

The diagnosis of Ogilvie's syndrome was made based on the clinical and CT imaging findings.

CASE 2

A 27-years-old man was admitted to emergency department with severe abdominal pain followed by vomiting. He was unable to pass stool for two months and had a years history of obstipation. The patient had developmental disorders and epilepsy. Measurements of basic physiological parameters were within normal levels - respiratory rate 16 breaths per min, heart rate 60 beats per min, temperature 36,5°C, blood pressure 120/85 mmHg, oxygen saturations 98% on room air.

During a physical examination massive abdominal distention, diffuse abdominal pain, hyperactive bowel sounds, with no evidence of peritonitis were identified. Digital rectal examination demonstrated a hard mass of stool just above the anorectal ring. Routine and biochemical blood tests values were normal.

Whole abdomen did not fit in plain film of the abdomen (43 x 35 cm), because of the huge size (Fig. 3). Ultrasound examination was hard to interpretate because of distended bowel loops. A computed tomography (CT) scan was also performed and showed a massive dilatation of the rectosigmoid bowel, reaching 23 cm in diameter, with heterogeneous matter, other large bowel parts dislocated and dilatated 5 cm in diameter (Fig. 4).

Patient was treated conservatively as there was no evidence of perforation and no definite cause of mechanical obstruction. The Hirschsprung’s disease was not suspected. Despite management with intravenous fluids and enemas the patient’s condition became worse and the decision to perform a surgery was made. Sigmoid colon was resected and an intraoperative frozen sectional histological analysis has been performed (Fig.5).

Histopathological examination of the resected colon showed the absence of ganglion cells in the myenteric and submucosal plexus. Diagnosis of Hirschsprung’s disease was confirmed (Fig.6). The patient’s postoperative course was uneventful, and the colostomy was closed 3 months after this operation. Thereafter, his defecation status became almost normal.
Fig. 2. Case 1. Computed tomography scan of the abdomen demonstrating persistent massive dilatation of the entire large bowel.

Fig. 3. Case 2. Plain film showing massive air fluid level and dilatation at the hepatic flexure (arrow). Small bowel loops are invisible, x-ray film can't contain diaphragm cupola and pelvis because of huge abdomen size.
Fig. 4. Case 2. Computed tomography scan of the abdomen showing a dilated colon packed with feces.

Fig. 5. Case 2. Operative findings showing a massively dilated (mega) sigma.

Fig. 6. Case 2. Histological appearance of distal resection line. No ganglion cells were observed (arrows). H and E staining, x24.
DISCUSSION

We presented two cases that were very similar radiologically but different in patients age, pathology, clinical manifestations and treatment. Both conditions are rare and actual prevalence is unknown. The Hirschsprung's disease occurs in approximately one in 5000 live births. Ninety-four percent of these cases are diagnosed before the patient is aged 5 years. In rare milder cases of these disorders, the patient may not receive a diagnosis until he or she reaches adulthood [8] probably because the proximal colon is often hypertrophied to compensate for the distal obstructed aganglionic rectum [6]. Do MY et al. hypothesized that adult-onset Hirschsprung disease manifests as a mild form of HD because this disease is associated with a lower degree of genetic abnormality (RET transmembrane receptor system and the endothelin receptor system) than childhood HD [9].

The exact incidence of adult HD is unknown because those cases are frequently misdiagnosed as chronic constipation or undiagnosed at all [6]. Grove and Ahlawat suggested the incidence of adult HD high as 2 % of the population [7]. HD is characterized by aganglionosis (absence of ganglion cells) in the distal colon and rectum and is thought to either occur from a failure of neuroblasts in neural crest cells to migrate into bowel segments or degeneration of already migrated neuroblasts. It affects cells both in the myenteric and submucosal plexuses [10].

Ogilvie's syndrome is a rare but well recognized complication in the severely debilitated patient, after trauma, major surgery, or sepsis [11]. The pathophysiology of Ogilvie syndrome is not completely understood although it likely results from an alteration in the autonomic regulation of colonic motor function [12]. The vast majority of patients (>95 %) with ACPO have the syndrome in association with one or multiple predisposing factors or clinical conditions. In a large retrospective series of 400 patients, the most common predisposing conditions were non-operative trauma (11%), infections (10 %) and cardiac disease (10 %) [13]. The mechanisms through which these different conditions temporarily suppress colonic motility and induce dilatation are unknown [14]. The exact prevalence of ACPO is unknown. In our case we don't know exact predisposing condition because of patient's speech and hearing disorders.

Clinical presentations of Ogilvie's syndrome and adult HD are different. Most often ACPO affects those in late middle age (mean age of 60 years), occurs in hospitalized or institutionalized patients with serious underlying medical and surgical conditions and associated with a wide spectrum of illnesses [15] or adult HD mean age 27.8 year [8]. Ogilvie's syndrome is characterized by abdominal distention, abdominal pain, nausea and/or vomiting, with a failure to pass flatus and stools documented in up to 60% of patients. Abdominal distention usually develops over 3-7 days but can occur as rapidly as 24 h [13]. In surgical patient, symptoms and signs develop at a mean of 5 days post-operatively. Massive colonic dilatation may cause ischemia and perforation, with the subsequent clinical finding of peritonism [15]. The approximate risk of spontaneous perforation is 3 percent, with an attendant mortality rate of 50 percent [8]. Physical examination in the uncomplicated situation typically reveals a tympanic, non-tender abdomen, with high-pitched “tinkling”, reduced or absent bowel sounds. Patients with complications present with marked abdominal tenderness and systemic features such as fever, tachycardia and leukocytosis [15].

A life-long history of refractory constipation is typical clinical manifestation of adult Hirschsprung's disease, like in our second case. Patients manage this condition by using cathartic agents, bulk formers, and enemas [8]. When functional decompensation of the colon occur, fecal impaction and megacolon may warrant urgent surgical intervention [16]. Rarely these patients may experience rapidly worsening subacute intestinal obstruction or even acute presentation with complications such as enterocolitis or volvulus [17].

Diagnosis relies on accurate clinical observation and plain abdominal radiography showing various degrees of colonic dilatation, mainly involving the proximal colon. Plain abdominal radiog-
raphy can also give some indication of colonic diameter as well as detecting the presence of free air, suggesting perforation. CT should be performed to differentiate mechanical obstruction from pseudo-obstruction; CT with intravenous contrast has a sensitivity and specificity both of 91% [15].

The correct diagnosis of adult Hirschsprung’s disease is based on barium enema, anorectal manometry and, most important, the full-thickness rectal biopsy findings. Plain abdominal radiographs identify only nonspecific findings of massive distention of the colon, often with air-fluid levels in both conditions. A small narrowed distal segment in adult Hirschsprung’s disease may not be detected in approximately 20% of patients [8]. This finding may be due to a short, or more commonly, an ultrashort diseased segment [6]. Although CT is considered a common imaging modality for excluding other diseases such as colorectal cancer, which also causes chronic constipation in adults [8].

The HD diagnosis was confirmed after histological examination. Histologic examination of colorectal specimens for the presence of ganglion cells remains the standard method of evaluating patients with Hirschsprung’s disease and forms the basis for surgical treatment [18].

In both cases CT imaging was performed, and findings were very similar. In case 1, the diagnosis of Ogilvie’s syndrome was properly based on the grounds of massive bowel dilatation, absence of mechanical reason and patient clinical course. In case 2, HD was not suspected radiologically mainly because characteristic rectal narrowing was not identified on CT imaging. Both conditions should be distinguished mainly because of treatment possibilities. It is extremely important to diagnose HD because surgical management is effective with satisfactory long-term functional results and significantly improves quality of life [19, 20]. The principles of pull-through surgery are firstly to remove all the hypoganglionic segments and secondly to achieve bowel continuity between the normally innervated bowel and the anal canal in order to provide bowel continence in the long term [20, 21]. The most common procedure performed is the Duhamel procedure [17]. This operation is associated with lower rate of major postoperative complications than the other procedures, and the higher rate of good long-term results [21]. In our case patients condition with HD was not properly assessed. Only after surgery and histological evaluation true diagnosis was made.

Treatment of Ogilvie’s syndrome depends on the severity of the clinical picture and the perceived risk of imminent ischemia and perforation [15]. Conservative treatment with nasogastric suction, enemas, and neostigmine is highly effective [19]. Colonoscopy is a safe and effective method for treatment of this syndrome when conservative treatment has failed. Surgical operation is indicated only for realized or imminent perforation, or in patients who have not responded to maximum non-surgical measures [15].

In both cases learning point is that massively dilated bowel loops and patients with anamnesis of obstruction may be representation of life threatening conditions such as Hirschsprung’s disease and Ogilvie’s syndrome.

CONCLUSIONS

Proper diagnostic and management techniques allow to avoid life-threatening conditions such as adult Hirschsprung’s disease and Ogilvie syndrome.

STATEMENT OF CONFLICTS OF INTEREST

The authors state no conflict of interest.
REFERENCES