



## A CASE OF IDIOPATHIC MEGACOLON IN ADULT FEMALE: CHALLENGE IN DIAGNOSIS

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### ABSTRACT

Idiopathic megacolon is one of the culprit for chronic constipation and is a rare condition. Megacolon is divided into 3: Acute megacolon in which there is no obvious colonic disease (colonic pseudo obstruction); chronic megacolon which could be congenital, acquired or idiopathic; toxic megacolon, which occurs in association with inflammation of the colon. Until date, the exact prevalence of idiopathic megacolon is unknown. The onset of symptoms either in childhood or adulthood and more common among females. The objective is to report idiopathic megacolon as a rare cause of chronic constipation. We report a 55-year old woman, who presented with 10 days history of no bowel open and abdominal distention. She been having chronic constipation for the past 5 years and dependent on laxatives. Contrast enhanced computer tomography abdomen revealed grossly dilated descending and sigmoid colon with no obvious pathology. She was took to Operation Theater for exploratory laparotomy in view of grossly distended painful abdomen. Intra-operatively, grossly dilated descending and sigmoid colon with no obvious pathology detected. The others part of colon were normal in calibre. Decision for Hartmann's procedure was made and the final histopathological examination (HPE) reported as idiopathic megacolon.

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### INTRODUCTION

We report a 55-years old woman, who presented with 10 days history of constipation and abdominal distention. However, she was still able to pass flatus and no vomiting. Further history, she been having chronic constipation for the past 5 years which she opened her bowel only once in every three days or sometimes once in a week. She was dependent on oral laxatives for the past but no history of digital evacuation. Prior to this, her bowel habit was once per day during childhood and early adulthood. There was no significant drug history. Her abdomen was distended with mild tenderness over left hypochondriac region. There was no mass palpable on digital rectal examination and the anal sphincter was normotonic. There was no sign of sepsis on presentation. White cell count and electrolytes on admission were within normal range. Thyroid function test was performed later and it was normal. Initial abdominal radiography revealed grossly dilated colon filled with fecal material (**Figure-1**). Contrast Enhanced Computed Tomography (CECT) abdomen revealed grossly dilated sigmoid and descending colon with no obvious pathology detected (**Figure-2**). Other parts of the colon were within normal caliber. She was booked for emergency exploratory laparotomy as she still not able to pass motion and abdomen was marked distended with localized tenderness after 24 hours of admission. Intra-operatively, grossly dilated

descending and sigmoid colon with no obvious pathology detected (**Figure-3**). The others part of colon were normal in caliber. Decision for Hartmann's procedure was made and she recovered well post-operative.



Figure 1 Abdominal radiography with dilated large bowel and faecal loaded

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Figure 2 CECT-abdomen with grossly dilated sigmoid colon

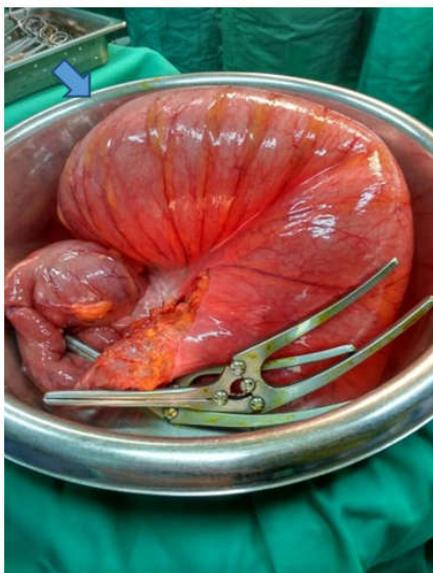


Figure 3 Grossly dilated descending and sigmoid colon intra-operative

The final histopathological examination (HPE) reported as idiopathic megacolon with present of ganglion cells. Subsequently, a full-thickness rectal biopsy was performed under spinal anesthesia and the HPE was reported to be normal with ganglion cell seen. Colonic transit time was performed and it was normal. She underwent reversal of Hartmann 4 months later and recovery was uneventful. Her bowel frequency improved to one in 1-2 days after the surgery.

## DISCUSSION

Idiopathic megacolon or idiopathic megarectum are rare and poorly understood condition. Until date, the exact prevalence of idiopathic megacolon is unknown. This disease could affect either the colon or the rectum alone, a situation respectively called megacolon and megarectum. Megacolon remains relatively poorly defined from the morphological point of view. Normal values for proximal and sigmoid colon diameters on single contrast enemas and double contrast enemas have been reported, however, there is a wide normal range and the diagnosis of megacolon remains subjective [1,2]. Megacolon can be classified into three groups: abnormality of ganglion cells, symptomatic megacolon (known etiology) and idiopathic megacolon. Clinical characteristics of Hirschsprung's disease may have one of several abnormalities of the myenteric plexus, including not only absence of ganglion cells, but also patchy or zonal loss, abnormal neurones or neuronal dysplasia [3]. Chagas disease

is one of the acquired megacolon, which is due to trypanosome infection that may lead to extensive destruction of ganglion cells in the peripheral autonomic system and may result in gross enlargement of the oesophagus, colon, and heart [4]. Idiopathic megacolon is a diagnosis of exclusion. The onset of the symptoms can occur in early or late childhood life or adult life and the etiology will be different at each onset. [1,2,3,4]

Initial assessment of constipation includes detail history and physical examination. Biochemical investigations such as electrolytes level and thyroid function test are vital as abnormality in both can result in constipation as well. Any patient who have red-flag symptoms or strong family history of colorectal carcinoma should undergo endoscopy examination. Abdominal radiography to look for the diameter of colon and retain of faeces in megacolon and megarectum cases. If the above tests are normal, colonic transit time and ano-rectal manometry should be order to further identify the cause of constipation [5].

Idiopathic megacolon describes an abnormality of colon characterized by permanent distension of bowel diameter in absence of identifiable cause. The pathological basis of underlying idiopathic megacolon is unknown. Postulated mechanisms include abnormalities in the extrinsic nerves, enteric nervous system, intestinal smooth muscle or neurotransmitter. Abnormalities of any of these components can cause colon dilatation and impaired colon motility. Histologically, idiopathic megacolon was characterized by a total atrophy of the collagenous tendinous connective tissue membrane of the myenteric plexus and the tendinous collagen fibre net of the muscularis propria. Thus, atrophy of the tendinous fibre net abolishes peristalsis and allows for unlimited distension of the colon [6,7].

Interestingly, the incidence of idiopathic megacolon in those of the female sex was seven times more frequent than in the male sex with mean age of 52 years but the true prevalence of this disorder is unknown [6,8]. Both Lane and Todd (1977) and Stabile et al (1991) reported adults tend to present with constipation, abdominal pain and distension while children presented with faecal impaction and soiling [9].

The diagnosis of the megacolon is made by clinical and radiological findings. Due to presence of bowel gas filling the dilated colonic segment, the rule of ultrasonography is limited. Contrast Enhanced Computed Tomography (CECT) with oral and rectal contrast is a better modality to visualize the dilated colonic segment, any mechanical obstruction and complications such as perforations. Colonoscopy can be used to exclude mechanical obstructive causes or even for colonic decompression, however the risk of perforation during procedure is high. Anorectal manometry may help to differentiate acquired and congenital forms. Rectal biopsy is necessary to establish the diagnosis [10]. Idiopathic megacolon and megarectum remained a diagnostic challenge, as there is no well establish diagnostic criteria available currently.

Many patients with idiopathic megacolon can be managed with long-term laxatives although some may require surgery later. Conservative management of patients with idiopathic megacolon is frequently ineffective. Surgery is indicated for those who fail conservative management or having disabling symptoms or who develop a complication which require urgent surgical intervention, such as perforation or ischemia [10,11]. As in this case, surgical measure was initiated during

this admission due to uncertainty in the diagnosis and her abdomen still distended and localized tenderness. Hartmann's procedure was performed instead of colectomy and anastomosis as the aganglionic segment of distal colon was one of our differential diagnosis. Thus, full thickness rectal biopsy and colonic transit study were performed after the surgery.

In this case, the clinical and radiological differential diagnosis at that point of time can be obstructed colorectal tumour, adult Hirschsprung's disease, closed loop intestinal obstruction, sigmoid volvulus, pseudo-obstruction and etc. Idiopathic megacolon is rare and thus is lower down in the list. The diagnosis of idiopathic megacolon was obtained from the HPE result post-surgery. Thus, it posed a challenge during the initial diagnosis and management. In contrast to Battal etc., they reported an elderly woman who presented with chronic painful distended abdomen with constipation. Examination revealed dilated abdomen with huge mass in the left lower quadrant. CECT-abdomen showed dilated sigmoid and rectum almost 18 cm with no obstructive pathology. The colonic wall was intact but thinner and no intramural gas. Idiopathic megacolon was diagnosed by them and she was treated with laxatives [11].

## CONCLUSION

Idiopathic megacolon should be considered in patients who have chronic constipation and by making this diagnosis, a series of investigations have to be performed to exclude organic causes.

**Declarations of interest:** All the authors state that there are no commercial financial incentives related with publishing current work.

**Consent:** Written consent obtained from patient.

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