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IDIOPATHIC MOTILITY DISORDER OF THE BOWEL

I Bombil, MD, FCS (SA); M Govender, MBChB, FCS (SA); M Tun, MBBS, FCS (SA), FRCS (I); S M Mulira, MBChB; A Kiss, MD, FCS (SA)

Department of Surgery , Chris Hani Baragwanath Academic Hospital and Faculty of Health Sciences, University of the Witwatersrand, Johannesburg, South Africa
Email: bombil@telkomsa.net

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Abstract

Introduction. Motility disorder of the bowel is rare and complex. Progress has been made towards establishing the diagnosis but unfortunately it does not translate into improved patient care. Often patient spend years on laxatives before surgical options are considered. For the sake of simplicity; this condition is divided in three categories: Motility disorder of the bowel (colonic inertia), functional pelvic outlet obstruction and combination thereof which further subdivides into congenital and acquired.

Objective. To present the unusual cases of idiopathic motility disorder of the bowel in order to reflect on its challenging management

Method. Retrospective review of 6 cases treated at Chris Hani Baragwanath Academic Hospital from 2007 to 2010. Parameters studied: patient demographics, clinical presentation, time of onset, management, investigations and outcome.

Result. 6 cases of chronic constipation. All of black African origin. Mean age of 18 years (11-32), predominantly male with a 5:1 ratio. 5 patients presented with severe constipation (1 stool per month) since birth and one patient had acquired constipation. Their mainstay of treatment has been chronic laxatives with insignificant benefit. 5 patients had total colectomy (of which one as an emergency) and one had segmental resection. All patients improved except the only one in the acquired limb who relapsed and the condition progressed proximally in the small bowel. In the congenital limb, 4 patients are off laxatives and one had partial response and remains laxative dependent. There was no evidence of any pathology on histology.

Conclusion. Rare condition with unknown aetiology and difficult to manage. Surgery offers satisfactory result in selected cases.

Introduction

Idiopathic motility disorder of the bowel is a rare entity not well understood, very complex . The pathophysiology is not well defined ; it represents an amalgam of disorder affecting the nervous and muscular system that interfere either with the emptying mechanism (functional pelvic outlet obstruction) where there is rectosphincteric incoordination or the motility of the bowel (slow

transit) resulting in chronic constipation [1-5]. Idiopathic constipation is the most common cause of motility disorder with unknown true incidence. Male appears to be affected more than female. There is a range of differential diagnosis divided into congenital (neuropathies, myopathies and idiopathic) and acquired (neurological, systemic, metabolic, psychological and idiopathic)

The hallmark of the clinical presentation is a chronic constipation which results on the long run in abdominal distension. The constipation is very frustrating, unremitting and force the patient in a regimen of chronic laxatives use with little satisfaction. Significant progress has been made towards establishing the diagnosis with little impact on the management.
The investigations include [6-12]:
Laboratory test: to rule out electrolytes abnormalities, hypothyroidism and metabolic causes.
Imaging

1. Abdominal X-Ray: Initial screening: often shows significant fecal loading and dilated large bowel.
2. Contrast enema. Confirm the X-Ray finding and rule out mechanical obstruction
3. Colonic marker transit studies. Give an idea whether we are dealing with colonic inertia or functional pelvic outlet obstruction based on the distribution of the ingested radiopaque tablets
4. Colonoscopy (Often unsuccessful due to inability to clear the bowel because of excessive fecal loading) and biopsy
5. Cine-defecography. It assesses the anorectal angle and the motion of all pelvic musculature during defecation of barium paste. It is seldom performed in children and unfortunately its findings do not guide the management toward a specific therapy.

Others (emptying studies):

1. Anorectal manometry to assess the presence of rectoanal inhibitory reflex: It implies relaxation of the internal anal sphincter when the rectum is distended by inflating a balloon. It may differentiate congenital from acquired megacolon
2. Total colonic manometry. To measure the intraluminal pressure and the contraction of the whole colon in an attempt to predict the extent of colectomy.
3. Pudendal nerve latency testing. Deals with matter (peristalsis, anatomy or neurology) related to evacuation.

Histology
Mucosal rectal biopsy: Suction Biopsy is adequate in most cases for Hirschsprung. In hollow visceral myopathy, degenerative smooth muscle can be confused with ischaemic change due to bowel distension, whereas in idiopathic megacolon both the ganglia and smooth muscle are normal.
Medical management[13]
Mainly to address the underlying causes. Various medications are being utilized targeting different pathophysiologies but without major break through. When constipation is due to psychological condition, retraining the bowel habit can be beneficial. In idiopathic motility disorder, the patient has to cope with the stress of lifelong laxatives uses.

Surgical management [14,15]

The Surgical approach can sometime be very uncertain due to the complex pathophysiology of this condition. Rarely curative but can make the use of laxative more manageable.
Besides, understanding the underlying condition does not necessarily mean improvement of patient care. The benefit of surgery is essentially when the removed organ (bowel) is the culprit. In idiopathic motility disorder, total colectomy removes the source of the problem if it is related to amotile bowel rather than to the rectosphincteric dyssynergy. There are
Various options but the principle is to balance the risk of incontinence (proctocolectomy) with the risk of residual constipation when a procedure lesser than a total colectomy is applied.
Several options are available[16]:
- Total Colectomy/Total proctocolectomy with ileostomy.
- Total colectomy/ total proctocolectomy with ileorectal or ileoanal anastomosis
- Segmental resection./diverting colostomy.
- ACE (antegrade colonic enema) procedure. Whether or not it improves motility remains questionable

Objective
To present the unusual cases of idiopathic motility disorder of the bowel in order to reflect on its challenging management

Method
Retrospective review of 6 cases treated at our colorectal unit at Chris Hani Baragwanath Academic Hospital (CHBAH) from 2007 to 2010. Parameters studied include patient demographics, clinical presentation, time of onset, management, investigations and outcome. Ethic approval for the study was obtained from the Human Ethics Committee of the University of the Witwatersrand.

Result
The demography is shown in table 1. The histology and outcome are depicted in table 2

Table 1. Patient demographics (N=6)

Mean , n (years), range	18 (11-32)
Male, n (%)	5
Female, n (%)	1
Male : female ratio	5:1
Blacks, n (%)	6 (100%)

Table 2. Histology findings and outcome

Case	Biopsy pre-operative	Histology post-operative	Outcome	Laxative dependent
1	no abnormality	No neuropathy or myopathy	4-6 stool /day vs 1/month preoperative.	No more
2	no abnormality	No neuropathy or myopathy	4-6 stool /day vs 1/month preoperative.	No more
3	no	No neuropathy or	4-6 stool /day vs 1/month	No more

	abnormality	myopathy	preoperative.	
4	no abnormality	No neuropathy or myopathy	ileostomy satisfactory	No more
5	no abnormality	No neuropathy or myopathy	Partial improvement	Yes but less frequently
6	no abnormality	No neuropathy or myopathy	1 stool/ month preoperative to 4 stool/day post operative then back to 1 stool/month within a year	Yes

Case 1

A 16 years old male had suffered severe constipation from birth passing one stool per month. He had had multiple admissions with gross abdominal distension and fecal loading managed medically with laxatives. Repeated rectal biopsies had excluded Hirschsprung's disease (HD) and hollow visceral myopathy (HVM). The patient required an emergency total colectomy with an ileostomy for toxic megacolon followed by ileorectal anastomosis a year later. Histopathological examination of the resected colon revealed normal myenteric plexus with ganglian cells with some hyperplasia of the nerve and thickening of the muscularis propria. There was also evidence of peritonitis with autolysis.

The patient improved markedly from 1 bowel movement per month to 4-6 per day with normal defecation. The skin condition (ichtyosis) showed also a significant improvement. Three years later, he remained stable, no more on laxative. We concluded that the abnormality affecting this patient is related to the colon itself and the emptying mechanism (pelvic floor) appears to be intact.

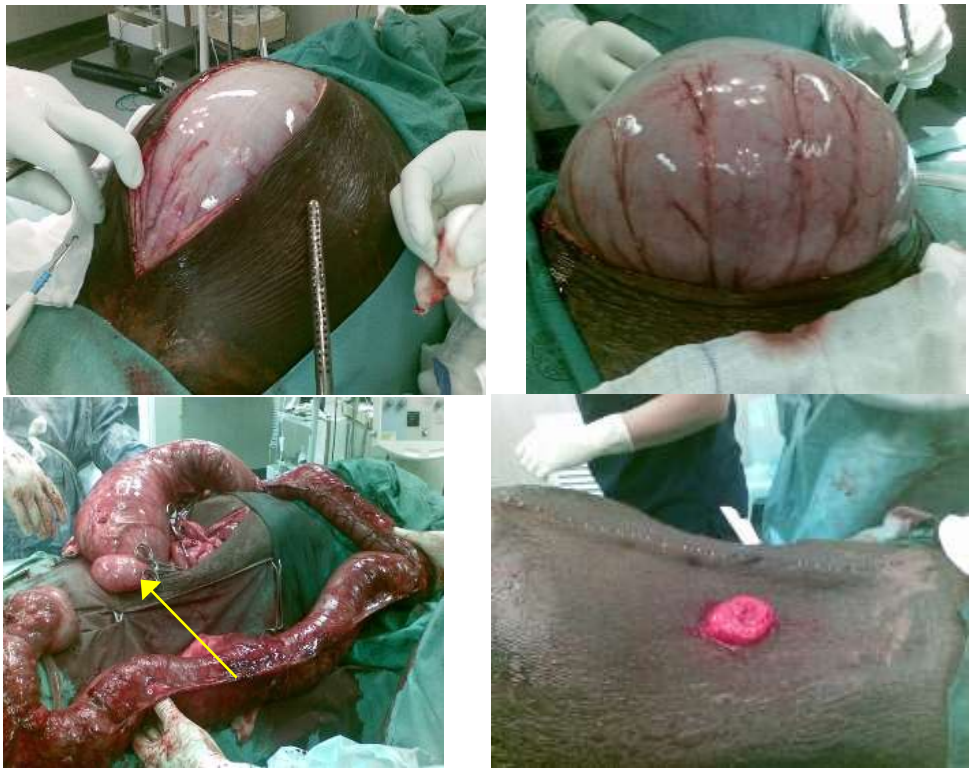

Fig.1. 1a,b: significantly dilated colon, 1c: notice a floppy bladder due to chronic compression, 1d: end ileostomy.

Case 2

A 17 years old male presented with intractable severe congenital constipation. At the age of one year he had had surgery for posterior urethral valve. At that time a full thickness rectal biopsy was normal. The patient underwent an elective total colectomy with an ileorectal anastomosis (fig.2) at the age of 17 years. Post operatively, the patient was noted to pass stool normally 4-5 times per day. The colectomy specimen reports no abnormality. At two years follow up, the patient remained well and no longer required laxatives



Fig.2. 2a. dilated colon , 2b. dilated colon loaded with faeces, 2c: ileorectal anastomosis

Case 3

A 15 years old male with severe chronic congenital constipation had an elective total colectomy with ileostomy. Histopathology confirmed intact ganglia with normal distribution and no evidence of HVM. Following restoration of intestinal continuity with an ileorectal anastomosis three month later, the patient had a normal bowel habit 4-5 times a day without laxative intake. At one year follow up the patient remained well.

Case 4

A 11 years old male was admitted with a diagnosis of large bowel obstruction having previously undergone two laparotomies (fig.3a). Notice the floppy bladder caused by chronic compression from the significantly loaded colon (fig.3d) and the right ureter adherent to the sigmoid colon making it vulnerable to injury during surgery (fig.3c).

A total colectomy was performed with an end ileostomy. The post operative course was complicated by a leakage from the rectal stump and the patient is awaiting reconstitution of the large bowel. Histology of the resected specimen revealed no evidence of HD or HVM. Post operatively the patient is off laxative.

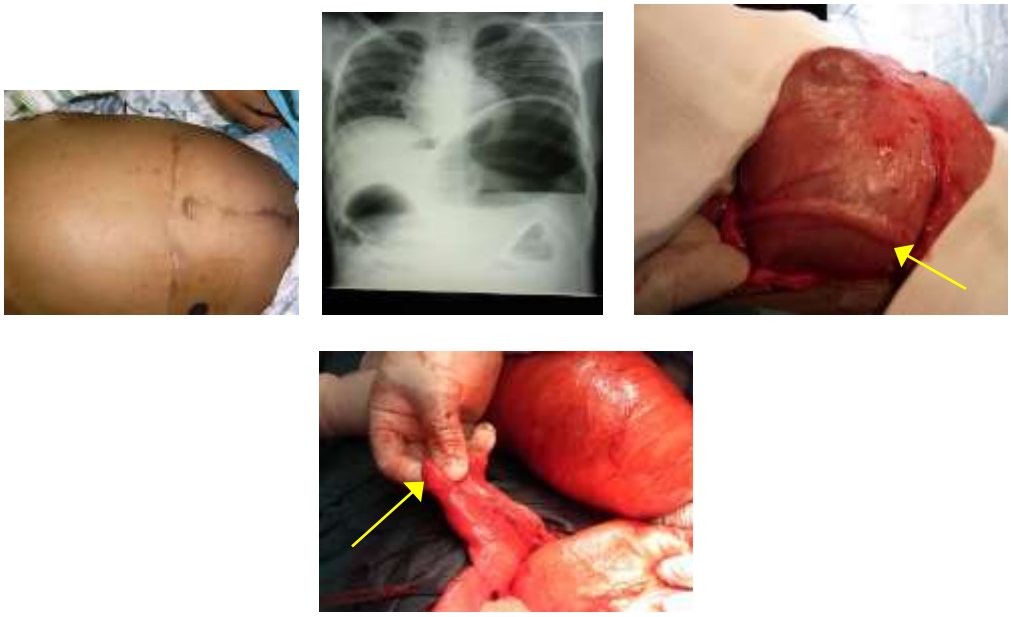

Fig.3. 3a: transverse and midline laparotomy scar, 3b,: dilated large bowel with fecal loading, 3c: note the right ureter adherent to the sigmoid colon, 3.d: floppy bladder due to chronic compression

Case 5

A 19 years old female, constipated since birth. At 4 years of age, she had a colostomy done followed by closure the same year. At the age of 17 years, she underwent a laparotomy for ovarian mass but the intra-operative finding was rather a very dilated sigmoid colon heavily loaded with feces, the rest of the colon was less dilated. The following year, she underwent a sigmoidectomy with a Hartmann colostomy after a full thickness rectal biopsy revealed no HD and no HVM (fig.4). She reported an improvement of bowel movement but a year later her constipation worsened after colorectal anastomosiswhich prompted a diverting transverse colostomy. This picture is suggestive of a combined motility disorder with dysfunctional pelvic outlet. She is still laxative dependent. The histopathological report of the resected specimen was normal.

Fig. 4. 4a,b,c: Extremely dilated sigmoid colon, 4d: Significant fecal loading of the rectum

Case 6

A 32 years old male with a very long history of adult onset chronic severe constipation. He is a known epileptic on medication. His constipation was refractory to laxative and rectal biopsy revealed no abnormality. The abdominal X-ray showed a markedly dilated large bowel. At the age of 32 years, after 5 years of conservative management, a total colectomy was performed with an ileorectal anastomosis. The initial good response of 4-6 bowel movement per day shifted gradually back to the pre-operative picture of one stool per month within a year of follow up. The histopathological report showed no HD and no HVM. On follow up, the disease progressed proximally to the small bowel (fig.5). He is totally laxative dependent. This picture is more suggestive of a hollow visceral myopathy although not confirmed on histology.

Fig.5. Dilated small bowel mimicking colon (a year post ileorectal anastomosis)

Discussion

Challenging condition both for the doctors and patients. Repeated biopsy yield nothing more and most of the patients spent all their childhood with no clear management plan due to the uncertainty of the diagnosis. Some of them had previous laparotomies with no evidence of any procedure (colectomy) done. The frustration is when we realized that all medical options we suggested have been offered already with no satisfactory result. Of the long list of investigations, only few were practical and did not contribute much toward the diagnosis. The real benefit was obtained by the removal of the affected organ (colon), but where there was a presumed functional pelvic outlet obstruction, we had no better option than to divert the fecal stream with a

stoma. The patient in case 5 behaved more like an anorectal dysfunction with significant dilatation of the sigmoid colon. Liu S et al reported on a 30 patients with idiopathic colonic constipation who were investigated with water perfused manometric system to evaluate the minimum relaxation volume, defecatory threshold, maximal tolerable volume and the compliance of the rectum; all these parametres revealed an abnormally high value compare to the control group of normal individuals[12]. There are promising researches toward the role of sacral nerve neuromodulation to treat incontinence and constipation associated with the motility disorder of the bowel[14].

There is a potential risk to develop complication at anytime. One patient developed a toxic megacolon prompting an emergency total colectomy. The psychosocial implications on these adolescents were significant; having to deal with a lifelong use of laxative, with a chronically distended abdomen and multiple unfruitful consultations. In contrast to the exceeding joy they experienced when surgery (total colectomy) provided a satisfactory result in favorable cases. It was much easier for the patients to appreciate a 4-6 bowel movement per day in contrast to one stool per month. Even with the post operative histopathological report, we still can not tell the underlying pathology. In all cases there was representation of ganglia cells and no evidence of HVM although the ischaemic change due to excessive bowel distension can be mixed up with myopathy. The interstitial cell of cajal are responsible for the motility of the bowel[4,10]. Dysfunctional cell of cajal is an option that is reported in the literature but we do not have any information to entertain this possibility in our cases. In a case report of idiopathic motility disorder of the bowel it was noted an ineffective response of the smooth muscle to the stimulation by acetylcholine[8]. Only the patient with acquired disease behaved much like hollow visceral myopathy but with no histological confirmation. The total colectomy in this case yielded a temporary relief with subsequent relapse back to square one within a year and there was a radiological evidence (markedly dilated small bowel) of proximal progression of the disease. This patient is bound to laxative with little benefit and had multiple admission for functional bowel obstruction that the unwary doctor can easily attribute to adhesions.

Conclusion

Rare condition with unknown aetiology and challenging management. Only when the affected organ (bowel) is removed does surgery offer a gratifying result. Although the understanding of the underlying pathophysiology has improved, we still do not have a better way of correcting the functional pelvic outlet obstruction except a diverting colostomy living the patient still frustrated with the idea of permanent stoma and lifelong laxative.

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