INTRODUCTION

Hollow Visceral Myopathy (HVM) is encompassed by intestinal motility dysfunction without the presence of mechanical obstruction [1]. It falls under a general classification of Chronic Intestinal Pseudo-obstruction which is a term describing signs and symptoms of gastrointestinal obstruction in the absence of a mechanical cause [2]. Broadly, enteric visceral myopathies encompass a cluster of genetic disorders that are characterized by smooth muscle dysfunction and exhibit clinical features that may include abdominal pain, features of malabsorption, and abdominal distension [3]. The more severe forms of enteric visceral myopathies typically present in the neonatal period, however, the mild forms may only present in adulthood [4]. Because HVM is one of the milder and rarer forms of enteric visceral myopathies, it is often misdiagnosed and underdiagnosed and, thus, literature on it is scanty [5]. A number of case reports show patients of HVM presenting with chronic symptoms of nausea, vomiting, and abdominal distension with pain [6]. A diagnosis is reached by excluding mechanical obstruction and establishing the presence of impaired intestinal motility through radiological and histological investigations [7].

Morphologically, the earliest lesion in HVM is an isolated smooth muscle degeneration of the muscularis propria of bowel – particularly the colon. Pathological changes noted are: lysis and disappearance of the bowel wall muscularis propria that then leads to dilatation and intra-luminal stasis, and progressive involvement results in large areas of defective muscularis propria and classical honeycomb fibrosis [8].

This case report presents as a clinical conundrum with intra-op findings of a para-duodenal volvulus along with a megacolon in a patient with a previous reversal of Hartmann's colostomy secondary to an initial sigmoid volvulus resection. Once again, highlighting the importance of history, examination and investigations in the work up of HVM, with the crucial multi-disciplinary approach and management that these patients require.

CASE PRESENTATION

Presentation

A 40-year-old black South African male presented to a tertiary Surgical Emergency Unit (SEU) hospital complaining of an eight-day history of constipation and increasing abdominal distension and pain associated with a one-day history of vomiting. Past surgical history encompassed a sigmoidectomy for a necrotic sigmoid volvulus (Histology: inflamed and ischemic mucosa, no evidence of malignancy of infection) with a Hartmann's stoma 18 months prior with an uncomplicated reversal of stoma 6 months after the initial surgery. Patient reported no other medical
history and was a non-smoker with no ethanol intake. The patient’s father had demised many years prior due to ‘a distended abdomen’.

**Clinical examination**

On general examination: severely emaciated patient, no generalized palpable lymph nodes, heart rate (HR)=122 beats per minute, Blood pressure (BP) = 118/68mmHg, Temperature = 36.2°C, respiratory rate (RR) = 24 breaths per minute, patient was severely wasted. Abdominal examination: massively distended, tense, peritonitic and tympanic on percussion; previous midline laparotomy scar with a left lower scar of the previous stoma site. Per rectum examination: empty rectum, no lesions, normal prostate. Cardiovascular and respiratory examinations were normal.

**Pre-operative management**

Two large bore intravenous (IV) lines were inserted and one litre crystalloid fluid bolus given with subsequent crystalloid infusion rate of 125ml/hr IVI. Urine catheter inserted drained 400ml clear urine. Nasogastric tube (NGT) inserted and two litres of feculent material drained. Chest Xray: bilaterally elevated diaphragms with small but clear lung fields and no free air under the diaphragm, Abdominal Xray: massively dilated loops of bowel, no air in the rectum, multiple air-fluid levels. COVID swab: negative. Patient was booked for emergency laparotomy for peritonitis with bowel obstruction.

**Intra-operative Management**

Theatre:

1) Massively dilated colon from caecum to cuff of rectum (more than 10cm diameter) but viable (Figure 1).

2) Paraduodenal volvulus with ischaemic but viable bowel (Figure 2) (of note: two stitches noted in the mesenteric region of ligament of Treitz – query of previous para-duodenal volvulus on initial laparotomy of sigmoidectomy – however, theatre notes did not indicate this and no mention of small bowel pathology was noted.)

3) Multiple, large mesenteric lymph nodes

4) Distended small bowel

5) Ten litres of liquid stool drained from small and large bowel via a large bowel enterotomy

6) Multiple chronic liver adhesions, normal spleen, normal gallbladder

**Procedures undertaken:**

- Detorsion/untwisting of paraduodenal volvulus – all loops of bowel viable, thickened mesentery, no other surgical intervention taken
- Subtotal colectomy: resection of terminal ileum to cuff/proximal rectum: rectal stump formed and Brooke’s ileostomy brought out
- Intra-abdominal washout, all remaining bowel viable with good peristaltic movements
- Abdomen closed and patient remained intubated and transferred to ICU on 0.2 micrograms of adrenaline infusion.

![Fig 1: Dilated Colon with ischaemic small bowel](image)

**Table: Arterial Blood Gas**

<table>
<thead>
<tr>
<th>Arterial Blood Gas</th>
<th>Value</th>
</tr>
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<tbody>
<tr>
<td>pH</td>
<td>7.549</td>
</tr>
<tr>
<td>PCO2</td>
<td>28</td>
</tr>
<tr>
<td>PO2</td>
<td>79</td>
</tr>
<tr>
<td>Saturation</td>
<td>97</td>
</tr>
<tr>
<td>Lactate</td>
<td>3.2</td>
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<tr>
<td>Base Excess</td>
<td>+5.6</td>
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<tr>
<td>Bicarbonate</td>
<td>29</td>
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<tr>
<td>Chloride</td>
<td>81</td>
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<tr>
<td>Sodium</td>
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</tr>
<tr>
<td>Potassium</td>
<td>2.1</td>
</tr>
<tr>
<td>Haemoglobin</td>
<td>13.3</td>
</tr>
</tbody>
</table>

**Table: White Cell Count**

<table>
<thead>
<tr>
<th>White Cell Count (x10⁹/L)</th>
<th>Haemoglobin (g/dL)</th>
<th>Mean Corpuscular Volume (fL)</th>
<th>Platelet count (x10⁹/L)</th>
<th>Urea (mmol/L)</th>
<th>Creatinine (micromol/L)</th>
<th>C-Reactive Protein (mg/L)</th>
<th>Amylase (U/L)</th>
</tr>
</thead>
<tbody>
<tr>
<td>10.84</td>
<td>12.4</td>
<td>80.7</td>
<td>373</td>
<td>7.9</td>
<td>72</td>
<td>20</td>
<td>50</td>
</tr>
</tbody>
</table>
Hollow Visceral Myopathy: Diagnosis and Management

HISTOLOGY REPORT

MACROSCOPY
Specimen comprises of terminal ileum, caecum, ascending colon, transverse colon, and descending colon.

Proximal small intestinal resection margin measures 40x15mm and appears viable. The distal colonic resection margin appears viable and measures 70x20mm.

Terminal ileum measures 90x45x15mm
The colon is markedly dilated as:
- The cæcum measures 120x40x45mm
- The ascending colon measures 180x90x45mm
- The transverse colon measures 250x60x35mm
- The descending colon measures 330x90x30mm
- The appendix measures 85x12x8mm

No perforations or fibro purulent exudate noted on the serosal aspect.
On opening of the bowel, the mucosa appears haemorrhagic throughout.
No perforations or ulceration noted. There are no polyps, diverticulae or tumours identified.

MICROSCOPY
The sections of terminal and large intestine display mucosal erosion
The muscularis propria of the colon is hypertrophic and there is myocyte vacuolation and hyper eosinophilia. Within the external longitudinal layer of the muscularis propria, there are distinct areas of fibrosis which are highlighted by Masson Trichome stain
Ganglion cells are present throughout. There is no neural hypertrophy
The resection margins, inclusive of terminal ileum are viable
No neoplasia is evident
No microscopic colitis is identified
No features of inflammatory bowel disease is present
No vasculitis, thrombosis or emboli are identified
No viral cytopathy, granuloma, protozoa, parasites or spirochaetosis is evident

DIAGNOSIS: features suggestive of hollow visceral myopathy

Post-operative management
Patient weaned off inotropic support overnight and patient was subsequently extubated. Six hours post extubation the patient was initiated on oral sips per os. The ileostomy was pink and viable with 50ml of intestinal output in 24 hours. Prophylactic thrombolytic therapy and Enhanced Recovery After Surgery (ERAS) protocol was followed. Constipating agents initiated (codeine phosphate, loperamide) with...
electrolyte replacement (calcium, magnesium, phosphate, potassium) started in order to decrease the ileostomy output of 1.1litres per day, alongside increased caloric and high-fibre dietary intake. Patient educated on stoma care and management. On Day 12 post admission patient maintained a stoma output ranging from 250–400ml daily for 3 days. All formal bloods were normal with electrolytes normal. Day 15: Patient discharged home.

**DISCUSSION**

Intestinal pseudo-obstruction comprises various disorders and because of its rarity and vague symptoms, it is easily misdiagnosed. Its origins may be primary (idiopathic) or secondary (due to other diseases), namely: encephalitis, cerebral vascular accident, Ehlers-Danlos Syndrome, Systemic Lupus Erythematosus (SLE), Amyloidosis, and scleroderma. With regards to primary intestinal pseudo-obstruction, the myenteric plexus may be defective involving the nerves and ganglia (visceral neuropathy), or be a result of smooth muscle degeneration and fibrosis (visceral myopathy).

Once again, because of the varying symptoms (from minimal to severe abdominal pain/distension/vomiting/constipation) the diagnosis may be missed; some patients may present with associated urinary retention or recurrent urinary tract symptoms. Therefore, differential diagnoses are of utmost importance: Hirschsprung’s Disease, leiomyomatosis/angio-lymphangioleiomyomatosis, hamartomatous lesions. Invariably, visceral myopathy is rarely linked to malignancies.

In this case study, in the acute presentation HVM was not considered, and it was neither considered when the patient had presented with a sigmoid volvulus 18 months prior, nor during the time of his reversal. Considering the patients age, previous surgical history and his current presentation, the differential diagnosis above should have been considered. The appropriate management of pseudo-obstruction is dependent on its consideration and thorough work-up with investigations including, colonoscopy with biopsy, contrast studies with plain X-rays, anorectal manometry, and histology. Full-thickness or mucosal suction biopsies are adequate to denote the presence or absence of ganglion cells.

The management requires a multidisciplinary approach involving gastroenterology, dietician, and surgery. Acute relief of symptoms may involve decompression with nasogastric tube and rectal tube insertion, and if this fails decompression with colonoscopy may be performed. Typically, if no perforation is noted then the primary management would involve non-operative conservative management that may involve high-fibre diet/bulk ing agents, as-needed facal disimpaction (using suppositories and enemas), correction electrolyte abnormalities, and removing aggravators (i.e.: narcotics, calcium channel blockers, or anticholinergics). However, if dilatation continues to persist the operative options are total colectomy with ileorectal anastomosis or total colectomy with end-ileostomy, with the former being the operation of choice.

With all the above considered, this patient may have benefited with more extensive investigations such as colonoscopy with biopsy and contrast studies during his re-presentation. This would have allowed for a trial of non-operative management with a multidisciplinary approach, and only once failed, progression to surgery with the consideration of and ileorectal anastomosis under controlled elective conditions.

**CONCLUSION**

Hollow visceral myopathy is a very uncommon host of diseases. It should be suspected in patients with atypical signs and symptoms such as: chronic intermittent attacks of abdominal distension/pain with associated constipation or diarrhoea, no radiological evidence of mechanical intestinal obstruction, and possible family history of similar symptoms; and, thus, making its consideration crucial in the differential diagnosis stage. This allows for appropriate and targeted investigations of colonoscopy and biopsy with consideration of contrast studies to be included in the work-up. The confirmation of a functional obstruction permits the inclusion of a multidisciplinary team of gastroenterology, surgery, and dietician in order to pursue a trial of non-operative conservative management. Failure of this then allows for the possibility of surgery involving adequate pre-operative fluid resuscitation, correction of metabolic abnormalities, and a planned surgical approach, followed by target post-operative management that may involve and stomo output management and ensuring patients return to an independent life.

This case demonstrates the vague symptomatology, recurrent abdominal complaints, and the importance of a differential diagnosis of HVM in such presentations to guide with further management of these patients.

**Declaration of Conflicting Interests**

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**Ethics Approval**

Our institution does not require ethical approval for reporting individual cases or case series.

**Informed Consent**

Signed informed consent was obtained from the patient for their anonymized information to be published.
REFERENCES