

## Visceral Myopathy Causing Intestinal Obstruction: A Case Report

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### ABSTRACT

Visceral myopathy (VM) is a rare, severe, and often misdiagnosed pathological condition. It is characterized by impaired intestinal function and motility in absence of mechanical obstruction. It affects the smooth muscles of gastrointestinal tract without involving myentric neurons. A seventy year old lady presented with mucoid discharge and bleeding per rectum on and off usually during and after passing stools since one year and altered bowel habits for eight months. Radiological investigations revealed thickening of rectal wall. An endoscopic biopsy was performed which on microscopy revealed degeneration, thinning and fibrous replacement of smooth muscle and dysplastic changes in glands. An abdomino-perineal-resection (APR) was done which on histopathology was reported as visceral myopathy. This report emphasizes on the fact that visceral myopathy, though rare, should be considered whenever a patient presents with uncharacteristic abdominal symptoms, recurrent attacks of abdominal distension and pain.

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**Key words:** Visceral myopathy; Smooth muscles; Intestinal obstruction

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### INTRODUCTION

Visceral myopathy (VM) is a rare pathological condition characterized by impaired intestinal function and motility in absence of mechanical obstruction. It affects the smooth muscle of the gastrointestinal tract (GIT) and sometimes the urinary tract. It is a form of chronic intestinal pseudo-obstruction<sup>[1]</sup>. The diagnosis can be delayed due to the rarity of VM, great variation in symptoms, and the many similarities it has with other more common gastrointestinal diseases. Furthermore, there is almost always absence of specific radiological features that indicates the diagnosis VM<sup>[2]</sup>. Of the VM, the cases are either familial or sporadic. Regardless of the inheritance pattern, VM is overwhelmingly attributed to an atrophic muscularis propria. The atrophy is variable but described as a marked vacuolar degeneration of myocytes, loss of muscle fibers, and with or without a highly characteristic honeycomb fibrosis. A hypertrophic pattern in VM, dominating the atrophic pattern, is exceedingly rare<sup>[3]</sup>. We report a case of visceral myopathy in a seventy years old lady presenting with signs and symptoms of intestinal obstruction.

### CASE REPORT

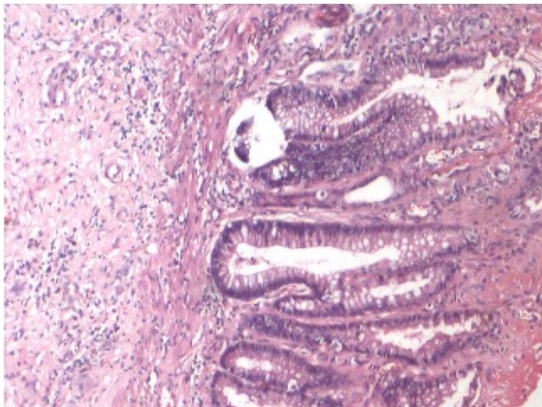
A seventy years old lady presented to surgery OPD with chief complaints of mucoid discharge and bleeding per rectum on and off usually during and after passing stools for one year and altered bowel habits for eight months duration. She also noticed streaks of blood with stool on defecation. There was no history of malena, significant weight loss, anorexia, vomiting, fever or jaundice. There was no history of chronic illness including diabetes or hypertension. There was no pallor, icterus, cyanosis, edema or lymphadenopathy on physical examination. Abdomen was soft with no organomegaly on palpation. Per rectal examination revealed decreased anal tone, circumferential thickening of rectum 5cm from anal verge partially obstructing the lumen. There was no evidence of any haemorrhoids, fistula or fissure. All routine haematological and biochemical investigations were within normal limit. Abdominal ultrasound revealed thickening of rectum wall. Rest abdominal organs were normal. CECT abdomen revealed thickening of wall of rectum with stranding seen in parametrial fat planes. Enlarged obturator group of lymph nodes on left side and pre sacral lymph nodes were also

seen. An endoscopic biopsy was performed and sent to the pathology department. On light microscopy the biopsy revealed degeneration, thinning and fibrous replacement of smooth muscle and dysplastic changes in glands (Figure 2).

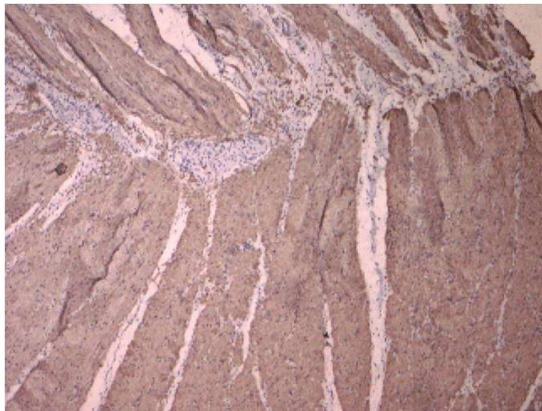
Laprotomy was planned and primary resection of recto-sigmoid with Hartman's procedure was done. In pathology department, we received a portion of sigmoid colon and rectum which was 18cm long. Mucosa revealed multiple ulcerative lesions and was atrophic and sloughed out at places. Ill defined nodular thickening were present in the wall (Figure 1). On light microscopy the H&E stained sections revealed that the nodule comprised of markedly hypertrophied and organized smooth muscle bundles (Figure 3). Hypertrophied nerve bundles and ganglion cells were also seen. The mucosa and submucosa over the nodule revealed extensive ulceration. Areas showing fibrinoid necrosis, marked oedema and disruption of muscularis mucosae were evident. Special staining with Von Geison



**Figure 1** Ill defined nodule at the distal end between mucosa and serosa.

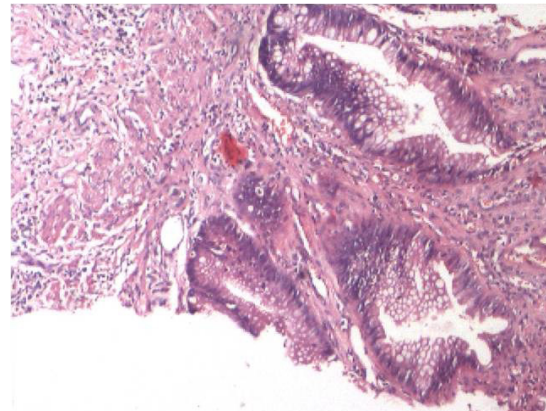


**Figure 2** Endoscopic biopsy showing high grade dysplasia with desmoplastic reaction around the glands indicating intramucosal malignancy (H&E, 40 $\times$ ).

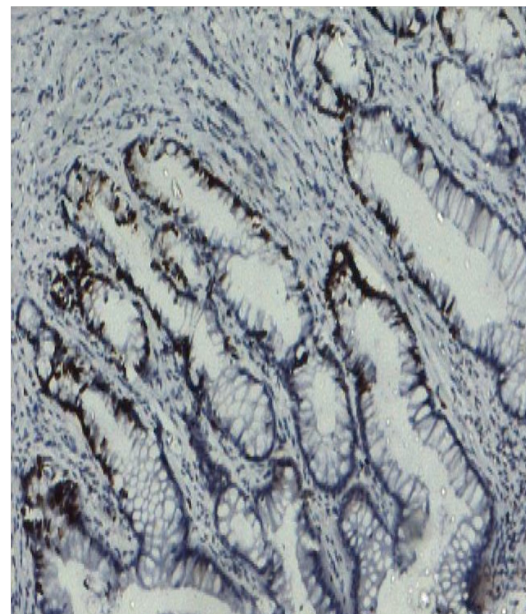


**Figure 3** Smooth muscle antigen showing positivity in hypertrophied muscles (40 $\times$ ).

was done which showed fibrosis of muscularis mucosae, submucosa and muscle. A small focus of regenerating mucosa in the distal cut end revealed mild to moderate cytoarchitectural dysplasia, fibrillary degeneration of muscularis mucosae and fibrosis of submucosa. It was a possibility that this dysplasia could actually worsen in continuity in the mucosa of the rectum left behind. Patient was taken for resurgery within a week. The distal rectal stump was removed and sent for histopathological examination. The H&E stained sections prepared revealed high grade dysplasia with desmoplastic reaction around the glands indicating intramucosal malignancy. It was not possible to rule out that muscularis mucosae was entirely free of microinvasion. Eventually abdominoperineal resection was performed. The APR specimen received in the department was 8cm long and a jumbled up mass showing multiple nodules in the wall more than the number seen in sigmoid colon (Figure 4). On light microscopy the H&E stained slides revealed ulceration and variable dysplastic changes including grade 3 along with foci of intramucosal malignancy in the mucosa. The regenerative hyperplasia at places gave impression of pseudoinvasion. Sections from the distal resected ends were negative for dysplastic changes. To assess the proliferative capacity of the epithelial cells Ki-67 staining was done and it showed >50% positivity in dysplastic glands (Figure 5). Hence finally diagnosis of visceral myopathy was suggested.



**Figure 4** The nodules from APR specimen showing marked cytoarchitectural dysplasia with desmoplastic reaction around the solid buds (H&E, 40 $\times$ ).



**Figure 5** Ki-67 positivity in dysplastic glands (40 $\times$ ).



## DISCUSSION

Intestinal pseudo obstruction encompasses a broad range of disorders and is classified as primary (idiopathic) or secondary to other diseases (scleroderma, systemic lupus, amyloidosis, Ehlers- Danlos syndrome, stroke, or encephalitis)<sup>[4]</sup>. The primary form of the disease may be due to inherent defect in myenteric plexus involving the nerves and ganglia (visceral neuropathy), or due to degeneration or fibrosis of smooth muscle (visceral myopathy). Disorders affecting smooth muscles without involving myenteric neurons are grouped under hollow visceral myopathy. This is considered to be the commonest cause of chronic intestinal pseudo obstruction<sup>[5]</sup>. Hollow visceral myopathy can be familial or sporadic. The familial form can be transmitted by a dominant or recessive gene<sup>[6]</sup>.

Patients suffering from VM vary greatly in their clinical manifestations. Some patients are completely asymptomatic, some patients will experience light abdominal symptoms such as nausea, vomiting, postprandial pain, abdominal distension, and diarrhea, and few patients experience repetitive attacks of bowel obstruction resembling small bowel or colonic obstruction<sup>[2]</sup>. No biliary involvement or uterine dysfunction have been reported. Few patients may present with recurrent urinary tract infections or urinary retention when visceral myopathy involves the bladder muscles<sup>[1]</sup>.

Light microscopy of such cases show typical features of degeneration, thinning and fibrous replacement of smooth muscle of GIT<sup>[5]</sup>. Myopathy preferentially involves the external longitudinal layer of muscularis propria characterized by vacuolar degeneration of smooth muscle cells distinguishing it from progressive systemic sclerosis<sup>[7]</sup>.

Differential diagnosis in cases presenting with intestinal obstruction includes Hirschsprung's disease, pseudo- Hirschsprung's disease, progressive systemic sclerosis involving gut, leiomyomatosis/ angio-lymphangiomyomatosis, hamartomatous lesion, age related neuropathies/ musculopathies. Preservation of ganglion cells in normal number, nodules of hypertrophied smooth muscles scattered throughout colon and rectum and absence of distal stenosed segment and proximal dilated segment rules out Hirschsprung's disease<sup>[8]</sup>. Smooth muscle proliferation is invariably associated with angiomatous proliferation; both the components present in variable proportion forming masses of disorganized mature tissue supports leiomyomatosis/angio-lymphangiomyomatosis/hamartomas<sup>[9]</sup>. Pseudo-obstruction due to myopathy alternating with hypertrophy of smooth muscle may cause pathological changes similar to those seen in rectal prolapse syndrome where regenerating mucosa often become dysplastic and if left behind may progress to invasive malignancy. This was seen in our case. The mucosal atrophy, ulceration, necrosis and fibrosis could be explained due to ischemia and pressure caused by passage of hard stool. Repeated regenerative epithelial proliferation with diminished checks and balances due to ageing in cell cycle would predispose for dysplastic changes.

Secondary chronic intestinal pseudo-obstruction is managed by treating the underlying condition. Intestinal decompression by colostomy or tube placement in a small stoma can also be used to reduce distension and pressure within the gut. The stoma may be a gastrostomy, enterostomy or cecostomy, and may also be used to feed or flush the intestines. Colostomy or ileostomy can bypass affected parts if they are distal to (come after) the stoma. For instance, if only the large colon that is affected, an ileostomy may be helpful. Resection of affected parts may be needed if part of the gut dies (for instance toxic megacolon), or if there is a localised area of

dysmotility. Gastric and colonic pacemakers have been tried. These are strips placed along the colon which create an electric discharge intended to cause the muscle to contract in a controlled manner<sup>[10]</sup>.

Not much cases of malignancy related to visceral myopathy have been reported. To our knowledge only one case of 35 year old male with visceral myopathy associated with bladder adenocarcinoma was reported in New York in 2002<sup>[11]</sup>. It was observed on autopsy of the patient that the small intestine was infiltrated by poorly differentiated adenocarcinoma with signet ring cell features. The primary tumor site was not identified.

## CONCLUSION

Visceral myopathy is a very rare group of diseases and they should be considered whenever a patient presents with uncharacteristic abdominal symptoms, recurrent attacks of abdominal distension and pain with no radiological evidence of intestinal obstruction. This case, however, shows that visceral myopathy in extremely rare cases can present with rectal bleeding and changes of dysplasia seen in microscopic sections.

## CONFLICT OF INTERESTS

There are no conflicts of interest with regard to the present study.

## REFERENCES

- Zaidi SHT, Arif M, Zaidi Z. Hollow visceral myopathy in a 5 year old boy: a case report. *J Pak Med Assoc* 2003; **53**(2): 82-83
- Burcharth J, Olsen C, Rosenberg J. Acute abdomen and perforated bowel with a rare pathology: Nonfamilial visceral myopathy. *Case Rep Surg* 2011; **2011**: 645349.
- Koh S, Bradley RF, French SW, Farmer DG, Cortina G. Congenital visceral myopathy with a predominantly hypertrophic pattern treated by multivisceral transplantation. *Hum Pathol* 2008; **39**(6): 970-974
- Pal K, Moammar H, Mitra DK. Visceral myopathy causing chronic intestinal pseudoobstruction and intestinal failure in a child with Sanjad-Sakati syndrome. *J Ped Surg* 2010; **45**(2): 430-434
- Martin JE, Benson M, Swash M, Salih V, Gray A. Myofibroblasts in hollow visceral myopathy: the origin of gastrointestinal fibrosis? *Gut* 1993; **34**(7): 999-1001
- Jones SC, Dixon MF, Lintott D, Axon AT. Familial visceral myopathy: a family with involvement of four generations. *Dig Dis Sci* 1992; **37**(3): 464-469
- Smith JA, Hause SC, Madara JL. Hollow visceral myopathy: a light and electron microscopic study. *Am J Surg Pathol* 1982; **6**(3): 269-275
- Fiorino K, Liacouras CA. Congenital aganglionic megacolon (Hirschsprung disease). In: Kliegman RM, Behrman RE, Jenson HB, Stanton BF, eds. *Nelson Textbook of Pediatrics*. 19th ed. Philadelphia, Pa: Saunders Elsevier; 2011:chap 324.3
- Chan JK, Frizzera G, Fletcher CD, Rosai J. Primary vascular tumors of lymph nodes other than Kaposi's sarcoma. Analysis of 39 cases and delineation of two new entities. *Am J Surg Pathol* 1992; **16**(4): 335-350
- Rex DK. Colonoscopy and acute colonic pseudoobstruction. *Gastrointest Endosc Clin North Am* 1997; **7**(3): 499-508
- Darvishian F, Basham K. Familial visceral myopathy with carcinoma of unknown primary. *Ann Clin Lab Sci* 2002; **32**(1): 93-97

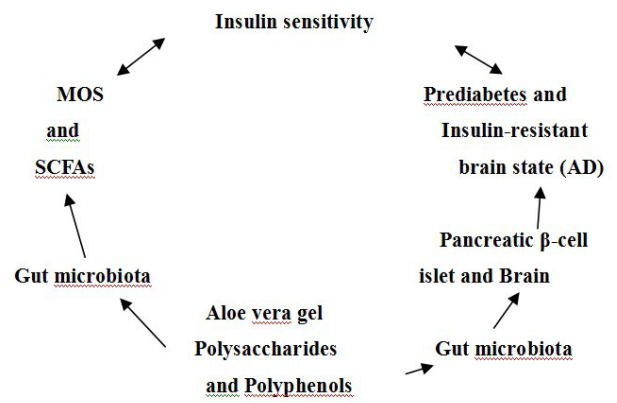
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Yagi A. Aloe vera gel metabolites



**Figure 1** Possible efficacy of aloe vera gel metabolites in long-term ingestion to insulin sensitivity.







