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REVIEW ARTICLE

Spontaneous Perforation of the Colon

Shariful Islam MBBS, DM, FACS, FASBrS, FCCS

¹Consultant, General, Laparoscopic and Specialized Breast Surgeon and Department of General Surgery; San Fernando General Hospital, Trinidad & Tobago and Associate Lecturer, Department of Clinical Surgical Sciences, University of West Indies, St Augustine, UWI, Trinidad & Tobago, West Indies

Abstract

Spontaneous colonic perforation is extremely rare. Several case reports and case series have been published on this topic in English literature. However, confusion still exists amongst General surgeons regarding its actual nomenclature. Different terminologies are used by many authors for these perforations i.e. spontaneous or idiopathic or stercoral. The aim of this topic is to make it clear to the surgeons about the difference between these types of perforations. This in turn will help them to have a better understanding of its exact terminology, diseases process and may guide them in the management of these patients.

Keywords: Colonic perforation, spontaneous perforation, stercoral perforation, idiopathic perforation

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INTRODUCTION

S pontaneous perforation of the colon is also referred to as sudden onset of colonic perforation. True spontaneous perforation of the colon in adults is a rare and unusual clinical entity. As the name suggests, there is no identifiable etiology; making this a diagnosis of exclusion. Similarly, it is very difficult to establish causality between the disease and associated patient's factor. The diagnosis is often delayed and usually made at laparotomy after excluding all possible causes ¹.

Sudden severe abdominal pain with frank peritonitis is a common surgical emergency and often can be life threatening once diagnosis and treatment are made late ². However, because of its rarity and the apparent lack of large case series on spontaneous perforation of colon in adults, its exact clinical features and outcomes are largely unknown. Mortality of this disease is secondary to the resulting fecal peritonitis and can be reduced by early recognition and timely

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Shariful Islam MBBS, DM, FACS, FASBrS, FCCS, Consultant, General, Laparoscopic and Specialized Breast Surgeon and Department of General Surgery; San Fernando General Hospital, Trinidad & Tobago and Associate Lecturer, Department of Clinical Sur-gical Sciences, University of West Indies, St Augus-tine, UWI, Trinidad & Tobago, West Indies



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surgical intervention.

Definition and Classification:

Spontaneous perforation of the colon is defined as an abrupt perforation of the colon without any identifiable cause. The first case was reported in a woman with spontaneous rupture of the rectum in 1872 by Brodie et al 1 .

Berry et al. divided it into- a) Idiopathic b) Stercoral perforation ³.

This classification is based on the etiological and pathological factors 4 . Since then, various terminology or descriptions were made in the English literature 5,6 .

There is a misconception amongst different authors about the exact terminology. There are differences between them but the descriptions are not consistent in literature because of the sporadic nature of case reports.

Currently, it has been described in literature as spontaneous colonic perforation (SCP), idiopathic perforation of the colon (IPC) and stercoral perforation of the colon (SPC). However, majority of the descriptions are reported as SPC ². Few authors have noted that the main difference between them were macroscopic as well as microscopic features^{1,5}.

Idiopathic perforation of the colon is defined as a spontaneous perforation of the colon in the absence of any colonic pathology, and without any history of chronic constipation.

Stercoral perforation is also defined as spontaneous perforation of the colon in the absence of any colonic pathology in patients with history of chronic constipation.

Incidence and Prevalence:

The true incidence of this disease is unknown. Similarly, available literatures have failed to document whether the disease patterns are the same between adults and children ^{7,8}. It accounts for 1% of all emergency colonic surgeries and 3% of all colonic perforations. However, the true incidence is much higher than previously thought ^{7,8}. The incidence is reported as 2% in an autopsy series. Recurrent spontaneous perforation is rarer than spontaneous colonic perforation ⁹. The increased incidence of the disease is commonly seen in debilitated, bed-ridden, mentally ill, or narcotic-dependent patients and is often related to a combination of factors; reduced activity, medication side effects and dehydration¹⁰.

It can occur in all age groups; the youngest being a six (6) year old and the oldest being a ninety six (96) year old ¹¹. Nevertheless, the prevalence of idiopathic colonic perforation favors the elderly with the average age of occurrence being 62.5 years with a male predilection 5,12,13 , but a recent study revealed that there was no gender difference ¹⁴.

The most common sites of perforation are sigmoid colon (54.1%), recto-sigmoid junction (21.5%), followed by the descending colon (13.6%), transverse colon [Figure 2] (9.6%), cecum (3.9%) and ascending colon $(1.3\%)^{5,8,13,15,16}$. The perforation usually occurs on the anti-mesenteric border of the bowel.

Etiology and pathophysiology:

Risk factors:

To date, the exact etiology of SCPA remains unknown. Few theories have been proposed to explain its pathogenesis including high intra-abdominal or intraluminal pressure, colonic implosion, ischemia, attenuation of the bowel wall, or laceration of the latter from hard feces. However, these factors were not supported by the existing evidence ^{17,18}.

The risk factors for SCPA are largely based on retrospective studies. Due to its rarity, prospective study or RCT is virtually impossible. Recent retrospective studies revealed that elderly patients with chronic constipation and associated unexplained abdominal pain were shown to be a high-risk group for SCPA.

Colonic perforations are usually associated with increased intraluminal pressure in patients with underlying pathology i.e. colitis, diverticulosis, malignancy, inflammatory bowel disease, foreign bodies, adhesions, irradiation, rectal and uterine prolapse, blunt trauma or iatrogenic injuries secondary to instrumentation ¹⁹. Nonetheless, it is also being reported in patients without any history of strain ²⁰.

The exact cause of spontaneous perforation of colon is unclear. Multiple medical conditions and medications have been related to SP, all of which share a common pathogenic factor of altered colonic motility.

a) Medical conditions:

Patients with comorbid conditions i.e. hypercalcemia ²¹, hypothyroidism, diabetic enteropathy ²², type IV Ehlers danlos ^{23,24}, prolonged immobility, peritoneal dialysis, chronic kidney disease, and renal transplant ^{25,26} often have intestinal hypo-motility, chronic constipation and /or fecal impaction ²⁷. These conditions put them into a high risk category for this type of perforation.

b) Drugs:

Patients on antacids, drugs containing codeine, narcotics ²⁸, non-steroidal anti-inflammatory drugs ²⁹, steroids, major tranquilizers, methadone, neuroleptics ³⁰ and tricyclic anti-depressants ³¹ have been reported as being at a higher risk of perforation.

Recently, it has been reported that IL-6Ra therapy for COVID-19 may be a potential mediator of colonic injury ¹⁶.

c) Chronic constipation:

Studies have shown that chronic constipation is an independent risk factor for SPCA. It present in 68.8% of patients in earlier studies ⁴ compared to 76.8% in recent studies ^{5,14}. However, spontaneous perforations are also reported in younger patients without any history of constipation, straining, trauma or underlying pathology ²⁰.

d) Anatomical weak points:

There are several weak points in the colonic blood supply resulting from incomplete anastomoses of the marginal arteries known as watershed areas. These are splenic flexure (Griffith's point), sigmoid colon, rectum (Sudeck's point) and ileocaecal region ¹². These areas are poorly perfused by both the superior and inferior mesenteric arteries (splenic flexure) and also by inferior mesenteric arteries, pudendal arteries and iliac circulations (Sigmoid colon, recto-sigmoid junction). Hypo-perfusion in these areas results in mucosal and mural infarction leading to ischemic necrosis and perforation (Figure 1).

Pathophysiology:

The pathophysiology of colonic perforation in patients with underlying diseases is well known.

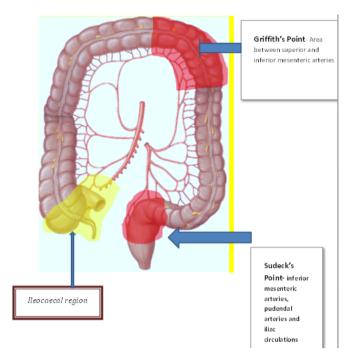


FIGURE 1: Anatomical weak points or watershed areas of the colon

Chronic straining causes raised intraluminal pressure resulting in the formation of recto-vesicle or uterine pouches, thereby thinning out the rectal wall. Sudden contraction of abdominal muscles during strenuous activities causes increased intraluminal pressure resulting in rupture of the colon or rectum through the thinned out area. This is usually at the antimesenteric border of the colon or rectum where the blood supply is poor. The reasons for perforation at the distal part of the colon are because of its physiological characteristics i. e higher intraluminal pressure due to its narrow luminal diameter, lower content of water in the stool, and relatively poor blood supply. It occurs commonly at the antimesenteric border of the colon or rectum.

Yang et al stated that the reason for the perforation at the recto-sigmoid junction was not only due its narrow luminal diameter but also due to the lack of anastomosis between the superior rectal artery and the lower branch of sigmoid arteries ³².

Patients with stercoral perforations usually suffer from chronic constipation. The impacted hard stools in the recto-sigmoid junction cause ischemic necrosis which extends from the mucus membranes to the muscular propria with resultant perforation of the colon $^{32-34}$. This result in a defect with necrotic edges, and a characteristic round to ovoid ulcer 35 (Figure 2).

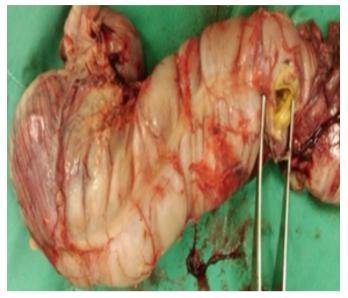


FIGURE 2: Intra op photograph of right hemi-colon showing a 2 cmround or oval defect on the an**ti**-mesenteric border of the proximal transverse colon

However, true idiopathic perforation of the colon has no identifiable cause. Two theories have been proposed for idiopathic perforation. The first one is the vascular theory and the second one is the weakness of the bowel wall associated with increased intra-luminal pressure ³⁶. These types of perforations are usually seen in a normal appearing colon and it tends to be a linear ulcer with clear edges with no microscopic evidence of any ischemia ³³.

Histo-pathological features of spontaneous perforations:

Chongxi R et al stated that the histo-pathological features of idiopathic (HIP) and stercoral perforations are unique¹⁴.

Both stercoral and idiopathic perforations are very difficult to identify at surgery as they differ both macroscopically and microscopically. Hence, histopathological examination is essential to reach a final diagnosis ¹². Additionally, there are no comparative studies focusing on the different types of histopathology for the SCPA.

The macroscopic and microscopic features of Idiopathic and Stercoral perforation are as follows 1,15,25,36,37

1a) Macroscopic features of idiopathic perforations are:

- Linear ulcer with clear edges,
- Normal underlying colon 5

1b) Microscopic features of idiopathic perforation are: (Figure 3a, b, c)-

- Normal mucous membrane of the colon
- Absence of abundant neutrophil infiltration around the perforation
- Absence of inflammatory bowel disease
- Absence of signs of ischemia or necrosis.
- Underlying muscle ends are broken but regular with non-specific changes in the mucosa.

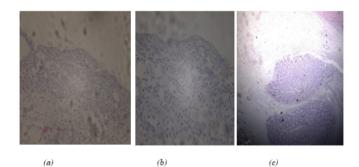


FIGURE 3: Histo-micrographs of idiopathic perforation-Microscopic view of mucosa (a, b), & serosa (c), showing the broken ends of the muscles are regular and there are no changes in the mucosa or signs of any ischemiaor necrosis

2a) Macroscopic features of stercoral perforations are:

- Typically round or oval
- Necrotic and inflammatory edges12

2b) Microscopic features of the stercoral perorations are: (Figure 4)

- Has a round or ovoid hole
- Necrotic and inflammatory edges
- Lies in the anti-mesenteric boarder
- Colonic mucosa becomes ischemic and necrotic
- Feculent ulcer

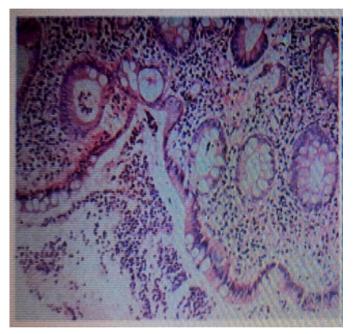


FIGURE 4: Histomicrograph of stercoral perforation showing superficial ulcer with numerous neutrophil infiltration at the edge of the ulcer

Key distinguishing features of Idiopathic and Stercoral Colonic Perforation: Table 1-

Clinical Features of SCPA:

The clinico-pathological features of SCPA are variable due to its extremely low incidence. It is very difficult to diagnose SCPA before surgery. Correct preoperative diagnosis rate varies from 11% to 20.6% ^{36,14}. The reasons for this low preoperative diagnosis of SCPA are as follows-

• The incidence of SCPA is very low

- The disease is not yet well known to the General Surgeons
- The imaging manifestations of SCPA are also not specific

Most of these patients usually present with unexplained abdominal pain, having no differences with other acute abdomen symptoms in the elderly. Abdominal findings can be either localized or generalized peritonitis. Most of these patients have a history of chronic constipation (76.8%), and the imaging findings are positive in 91.7% of them¹⁴.

Investigations of SCPA:

a) Plain abdominal Radiographs:

Is not specific and it only shows pneumo-peritoneum in < 50% of these patients 13,38 (Figure 5) -



FIGURE 5: Plain abdominal X-ray showing pneumo-peritoneum under both domes of diaphragm

b) Computed Tomography (CT) scan of the Abdomen and Pelvis:

Early CT scan of the abdomen and pelvis is very important in reducing mortality in these patients. It helps to diagnose the condition easily and shorten the interval to surgery. The important findings of SCPA

Key Fea- tures	Idiopathic colonic perforation (ICP):	Stercoral colonic perforation (STCP):
A. His- tory	No history of constipation	History of chronic constipation
B. As- soci- ated fac- tors	Usually no associated factors are present	 The use of aspirin, steroids, opiates, anti-cholinergic, and cytotoxic agents The patient on hemodialysis, history of renal or lung transplant and/or chronic renal failure
C. Oper- ative find- ings	No fecaloma seen protruding through the colon or lying outside the colon	Fecaloma within the colon, protruding through the perforation site or lying within the abdominal cavity, are the essential features of STCP
D. His-	 Linear The mucosal edge is clear 	 Has a round or ovoid hole Necrotic and inflammatory edges
	-	
tol-	3. Does not extend to the serosa, the broken	3. Lies in the anti-mesenteric boarder
ogy	ends of the muscular layer are regular	4. Involves ischemia and necrosis of colonic
	4. No ischemia	mucosa
	5. No feculent ulcer	5. Feculent ulcer

TABLE 1: Characteristic features of idiopathic colonic perforation (ICP) and stercoral colonic perforation (STCP)

on CT scan are as follows - free intra-peritoneal air, fat streaking, free fluids, extra-luminal fluid collection, and thickening of the bowel wall around the perforation site ⁴¹. The sensitivity of CT scan for SCPA is documented as 91.7%¹⁴. However, it cannot identify the exact cause of the perforation.

The presence of free intra-peritoneal air on radiograph is often believed to be diagnostic. It can be seen on plain abdominal radiographs especially on erect films, and on the CT scan, however it is less likely to determine the exact cause ⁴⁰. Pneumo-peritoneum accounts for approximately 85-95% of the visceral perforations, including SCPA ^{40,41}.

Pneumo-peritoneum in absence of fever or leukocytosis with minimal abdominal pain, distension, or peritoneal signs should be considered as a nonsurgical cause and should be treated conservatively ⁴¹.

Management of SCPA:

Currently, there is no guideline regarding the management of SCPA because of its rarity. They are usually managed following the protocol of traumatic perforation. Early clinical suspicion, timely diagnosis, and treatment are of paramount importance to reduce the complications and mortality rate in SCPA patients.

The type of surgery performed may vary depending on the hemodynamic stability of the patient, underlying medical conditions, the site and size of the lesion as well as the degree of peritoneal contamination. It can vary from laparoscopic lavage, open repair of the perforation, Hartmann's operation or colectomy with or without covering ostomy 13,42 .

Open Hartmann's procedure is the most frequently performed procedure and remains as the procedure of choice for SCPA patients ¹⁴. Patients with underlying comorbid condition, unstable hemodynamic status, poor nutritional status and moderate to severe degree of peritoneal contamination are best managed by this procedure. Although this is a morbid procedure and requires a second procedure to reverse it but, it has proven over the years as to be the safest procedure to

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reduce further perioperative complication as well as mortality.

Colectomy with primary anastomosis is now increasingly being performed by many surgeons as it is a one stage procedure and no further surgery is required. It is associated with the risk of anastomotic leak with its associated morbidity. It is feasible in SCPA patients with minimal peritoneal contamination, stable hemodynamic condition, and mostly in patients with right colonic perforation and in selective cases of left sided colonic perforations ^{13,14,42}. However, the ultimate decision of the type of surgery lies with the managing surgical team.

With the advancement of laparoscopic surgery, it is now increasingly being used in perforated peritonitis including colonic resection and anastomosis with or without de-functioning stoma or laparoscopic lavage and drainage ⁴³. Several recent studies showed that laparoscopic lavage is feasible and safe for perforated diverticulitis with purulent peritonitis ^{43–45}. Although laparoscopic lavage has been suggested as an alternative method for perforated peritonitis but its use is still limited especially for the SCPA.

Postoperative Complication and Mortality:

Prognosis of HIP patient are appeared to be better than that of HSP patients. The mortality of spontaneous perforation of colon depends on how early the perforation is recognized, underlying comorbid conditions, degree of peritoneal contamination, and adequacy of resuscitation and timing of the surgery. This mortality is a result of fecal peritonitis, which ranges from 35-45% with some authors estimating a higher mortality 5,12,46 .

Idiopathic perforation has a favorable outcome compared to stercoral perforation. With regards to the site of the perforation, the mortality from caecal perforations is noted to be highest ranging between $30\%-72\%^{47}$.

Similarly, amongst the different types of surgery; Hartmann's procedure was associated with lower mortality rate compared to other procedures ³⁶.

The postoperative complication rate is reported as 67.7%. Age of the patient is an important risk factor. Elderly patients with chronic constipation were noted to be a high-risk group ¹⁴.

Prevention of SCPA:

Stercoral perforation can be preventable as it is associated with several risk factors compared to idiopathic perforation which has none. As constipation is associated with a stercoral perforation, prevention of constipation by any means, i.e. dietary, life styles modifications or others can reduce the incidence of stercoral perforation. Additionally, early detection of stercoral colitis can prevent the progression to a perforation.

Idiopathic perforation on the other hand, is not associated with any known risk factor but has a more favorable outcome than a stercoral colonic perforation ¹⁴. This is because of the absence of underlying bowel inflammation or necrosis and also due to the minimum degree of fecal contamination.

Prevention of SP may be achieved by:

1) Health education to increase the awareness of this disease to the medical professionals and the general public.

2) Regular monitoring of bowel habits of the elderly, mentally impaired and, bed ridden patients. This includes abdominal examinations, careful rectal and manual evacuation of stool to stimulate bowel motions.

3) Dietary modification: high fiber diet, increase fluid intake, decrease flour, red meat and oily fatty foods

4) Limiting the use of drugs that reduce intestinal motility especially in chronically constipated and high risk patients.

Conclusion:

Spontaneous colonic perforation is an extremely rare disease. Due to its rarity, the exact etiology and pathogenesis has not been elucidated. Stercoral and idiopathic perforations are not same as spontaneous perforation rather these are subtypes of spontaneous perforation. Both of them have distinct macroscopic and microscopic features. Early diagnosis, resuscitation and prompt surgical intervention can often lead to a favorable outcome for most patients with SCPA. Types of surgery will depend on patients underlying comorbid conditions, hemodynamic status, and degree of fecal peritonitis. Constipation should be prevented or minimized to reduce the future risk of perforations in elderly patients with co-morbid conditions. Further research on the ultrastructure of the colon is required to yield more information about SCPA.

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