

Case Study

Solitary Rectal Ulcer Syndrome – A Potential Diagnostic and Management Dilemma for the Surgeon

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ABSTRACT

Occult mucosal prolapse syndrome, also known as the solitary rectal ulcer syndrome (SRUS) is uncommon. Due to its rarity, a misdiagnosis of rectal cancer is occasionally made as the clinical features may closely mimic those of rectal malignancy. We hereby report a case of SRUS in an elderly Malay gentleman who had primary symptoms of rectal bleeding with associated anaemia and anorectal pain. Even though the clinical features and specific investigations suggested the presence of rectal cancer, preoperative histological analysis failed to confirm this. In view of the intractable symptoms and rectal bleeding, a low anterior resection was performed. A detailed examination of the resected specimen intraoperatively, together with the histological report and awareness of this condition avoided the performance of an abdomino-perineal resection. Incidentally coexisting malrotation of the sigmoid colon to the right side was discovered during surgery. This finding, which may be coincidental, has not been reported thus far in the medical literature. The patient's symptoms improved postoperatively with a subsequent uneventful recovery. A brief literature review supplements this case report.

Keywords: Mucosal prolapse syndrome, solitary rectal ulcer syndrome, rectal cancer

INTRODUCTION

Solitary rectal ulcer syndrome (SRUS) or occult mucosal prolapse syndrome is a rare disorder characterised by disturbed defaecatory behaviour with passage of blood and mucus. Endoscopic findings of erythematous ulceration of the rectal wall is usually associated with typical histological features.^[1] The condition is however often misdiagnosed as a malignant ulcer or an ulcer associated with inflammatory bowel disease.

The term solitary rectal ulcer syndrome is a misnomer as the ulcers are often multiple in approximately one third of cases. Furthermore, there exists a pre-ulcer polypoid phase and similar lesions have been reported in the anal canal and sigmoid colon.^[2,3] Delayed or incorrect diagnosis of SRUS has been the subject of discussion in numerous reports.^[1] We add a further case to this clinical conundrum involving an elderly Malay gentleman who was also discovered to have a malrotated sigmoid colon, an association which has not been reported in the medical literature thus far.

THE CASE

A 60-year-old Malay patient was seen at the surgical outpatient clinic with symptoms of fresh rectal bleeding of a few months duration associated with tenesmus and perineal pain. Associated symptoms included alteration of bowel habit over an 18-month period. Rectal examination confirmed the presence of a mass involving the posterior wall of the lower rectum, located 5 cm from the anal verge.

Full blood count demonstrated a Hb of 8gm/dL. The rest of the haematological investigations were normal as was a chest x-ray and serum CEA. Colonoscopy confirmed the presence of a rectal mass, appearances of which resembled a neoplasm. Initial biopsy of the rectal lesion demonstrated inflammatory changes with no malignancy. A repeat biopsy was performed and the histological report on this occasion indicated a possible diagnosis of a solitary rectal ulcer and that malignancy could not be ruled out. A staging computed tomography of the abdomen and pelvis demonstrated a circumferential thickening of the lower rectum suggestive of a neoplasm with no evidence of local or distant metastasis except for a hypodense lesion in segment 7 of the liver.

In view of the high diagnostic suspicion of a rectal neoplasm even though there was uncertainty from a histological perspective, an elective anterior resection was planned. Informed consent included a detailed explanation of the possible pathological findings, especially of a benign nature. Intraoperative findings were of a malrotated sigmoid colon to the right side with minor tethering of the lower rectum posteriorly. No malignant lesion was found. There were no other abnormalities noted. A low anterior resection was performed with a covering loop ileostomy. The distal resection margin was approximately 0.5 cm from the lesion but this was accepted as intraoperative examination of the specimen revealed features that were more in keeping with a benign pathological process.

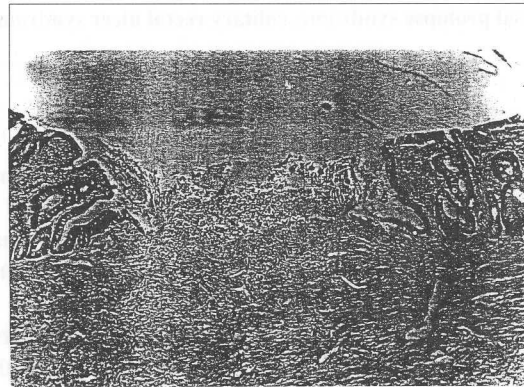


Figure 1. Histology of resected specimen (4x magnification) demonstrating an ulcerated area within the rectal wall

Histological analysis of the resected specimen confirmed the diagnosis of a solitary rectal ulcer syndrome (*Fig. 1*). The patient subsequently made an uneventful postoperative recovery and has remained asymptomatic since.

DISCUSSION

Although first described in 1829, the clinico-pathological features of SRUS were defined and published only in 1969.^[3] Localised colitis cystica profunda and inflammatory cloacogenic polyp are closely allied conditions with solitary rectal ulcer syndrome that have been linked to bowel prolapse.^[2] Affected patients often demonstrate abnormal function of the anal and pelvic floor musculature during defecation that leads to rectal mucosal prolapse or even intussusception.^[3] The resulting trauma is thought to cause the clinical symptoms and pathologic changes.

Patients with mucosal prolapse syndrome range in age from 10 to 83 years with the majority presenting in the third and fourth decade of life. The condition occurs more commonly in women.^[2] The rarity of this disorder has resulted in general lack of awareness of this syndrome.

The cause of SRUS is unknown. It is generally agreed that occult or overt rectal prolapse and paradoxical contraction of the pelvic floor muscles are among the factors involved in the development of SRUS. Congenital abnormalities have not been implicated thus far in the development of this condition, which suggests that the concomitant finding of a malrotated sigmoid colon in our patient was incidental. This statement however has to be balanced by the fact that few patients undergo a formal exploratory laparotomy and surgical resection that would confirm the presence of a congenital anomaly. The decreasing trend in barium enema evaluation of the large bowel could further hamper the discovery of such isolated anatomical abnormalities. The possible role of ischaemia in the pathogenesis of SRUS is highlighted by the association between the use of ergotamine suppositories and the development of the syndrome.^[4] Solitary rectal ulcer has also been reported to occur as a result of direct trauma from rectal digitation to aid evacuation and following radiotherapy.^[5] A disturbance of toileting behaviour, as an expression of psychological problems, appears to be an important pathogenic factor in some patients.^[5]

Table 1. Clinical presentations of patients with the solitary rectal ulcer syndrome
(Modified from references 1,2,3,5)

Feature	Relative Frequency
Difficulty with defaecation	100%
Rectal bleeding	68 - 91%
Passage of mucus	43 - 68%
Diarrhoea	19 - 27%
Pruritus ani	21%
Intermittent bowel habits	6 - 28%
Anorectal pain	9 - 24%
Abdominal cramps	12 - 16%
Constipation	12%
Feeling of rectal fullness or a sense of bearing down	8%
Incomplete rectal evacuation	6%
Overt rectal prolapse	4%
Faecal Incontinence	2%

The diagnosis of mucosal prolapse syndrome can be difficult because the history often suggests primary inflammatory or ischemic bowel disease. Table 1 outlines the various clinical presentations of mucosal prolapse syndrome along with their relative frequencies. The diagnosis can be made predominantly on the basis of symptoms, endoscopic appearance and supported by histopathological examination. Radiological and physiological investigations are of limited value. In addition, mucosal prolapse is often covert requiring special techniques for its demonstration. Patients often present long after the onset of symptoms with reported duration of symptoms before diagnosis as long as 5.3 years.^[1]

The clinical impression in patients with mucosal prolapse syndrome is often incorrect and includes idiopathic ulceration, Crohn's disease, non-specific proctitis, carcinoma, or villous adenoma.^[1] More importantly, patients with solitary rectal ulcer syndrome (SRUS) frequently present with a mass that can be misinterpreted as cancer apart from the presence of other symptoms that can mimic malignancy. In our patient, a provisional diagnosis of a rectal cancer was made as the age of the patient, symptomatology and investigations performed suggested the presence of a sinister pathology.

Characteristic findings on both transrectal and endoanal ultrasonography have been described,^[5] the former revealing thickening of the muscularis propria and the latter, occasional marked thickening of the internal sphincter. A definitive diagnosis usually requires demonstration of characteristic changes in the biopsy specimen as the proctoscopic appearance can often be misleading while sigmoidoscopic appearances vary from patient to patient.^[2,3] Ulcers are not universally present as demonstrated by its reported prevalence of 57% in one large series.^[5] When present, ulcers typically occur on the anterior or anterolateral wall of the rectum, usually between 5-10cm from the anal margin and centred on the rectal fold. In patients without ulceration, the mucosa appears polypoid (25%), roughened or erythematous (18%).

The characteristic histopathologic changes are found in the mucosa adjacent to the ulcer or in the polypoid areas and consist of fibromuscular obliteration of the lamina propria associated with mucosal architectural distortion often with a regenerative hyperplasia or villiform appearance.^[2,6] Inflammation is typically mild or absent. On occasion, colonic glands may be misplaced into the muscularis mucosae or submucosa, a histology referred to as "localized colitis cystica profunda". This can be associated with dissecting mucous pools and be easily mistaken for invasive mucinous adenocarcinoma.^[6]

There is no specific cure for SRUS. Symptoms may improve with limited treatment modalities but it is uncommon to achieve endoscopic and histological normality. The few studies with long-term follow up report a chronic but stable appearance of the lesions.^[2,3] The majority of mucosal prolapse syndrome patients tolerate their symptoms following reassurance that they do not have cancer. Addition of dietary fibre, biofeedback and behavioural retraining to reduce straining and avoiding digital manipulation have all been reported to produce variable success in symptomatic improvement.^[5] The rare patient with severe bleeding or obstruction requires excisional therapy. Patients with severe symptoms have been treated by rectopexy, resection which has included abdomino-perineal resection, diverting colostomy, or rectal prolapse repair.^[5] The results of surgery for this condition are varied since various surgical procedures have been described and comparisons are difficult, especially with small series of patients.

Rarely massive rectal bleeding may dictate the choice of operation, which includes sclerotherapy, circumferential suturing and even anterior resection should local therapy fail.^[7] Our patient had an anterior resection performed due to the underlying symptoms of rectal bleeding requiring transfusion, apart from the need to definitely rule out the presence of a rectal cancer. We stress the importance of discussing the possibility of a benign eventual finding with patients due to undergo resections for rectal lesions where equivocal pre-operative histological reports have been obtained. It is also prudent to examine specimens intraoperatively in such cases to avoid performing an abdomino-perineal resection for a benign entity.

CONCLUSION

The term SRUS is used to describe a rare disorder which covers a spectrum of clinico-pathological abnormalities which can affect both adults and children, often with characteristic clinical and investigative findings. It can be misdiagnosed as a malignant ulcer and the pathogenesis is likely to vary in different patients. Behavioural techniques directed towards defaecatory habits help a substantial proportion of patients and should be considered as the first line of management. Surgery has a minor role and should be reserved for those with intractable symptoms and had failed conservative therapy.

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