ABSTRACT

Solitary rectal ulcer syndrome is a multifactorial pathology, which entails a variety of clinical, histologic and endoscopic aspects that needs step-wise logical approach for management especially in relapsing refractory cases. Apart from the diagnostic dilemma that may be faced due to similarities of presentation with inflammatory bowel diseases or colorectal neoplastic lesions, the syndrome also overlaps with dyssynergic defecation syndrome, health anxiety disorder, obsessive compulsive disorder, and latent mucosal rectal prolapse, a systematic composite treatment modality including psychological, pharmacological, physiological and possibly surgical interventions are sometimes essential. Selecting appropriate treatment in this condition not only affects clinical outcome but also patients' experience and further stigma of SRUS life-long. In this review, we will discuss the detailed pathophysiology, diagnostic and therapeutic approaches in dealing with solitary rectal ulcer syndrome.

KEYWORDS:
Solitary rectal ulcer syndrome, Inflammatory bowel diseases, Colorectal neoplastic lesions

INTRODUCTION

A 32-year-old woman came to the outpatient gastrointestinal clinic and complained of intermittent abdominal pain, rectal bleeding, straining, and a sense of incomplete evacuation. She did not notice any involuntary weight loss. There was no personal and family history of colorectal cancer and other bowel diseases. She was worried about her diagnosis. What should be done for evaluation and management?

Cruevihier first described unfamiliar cases of rectal lesions in a series of patients in 1820s and a more than a century later, the term “solitary ulcer of the rectum” was introduced in the medical literature.¹,² Now a renowned medical condition, solitary rectal ulcer syndrome (SRUS) is a chronic infrequent disorder in gastroenterology. Its exact prevalence is not clearly known but a figure of 1:100,000 per year is usually reported³-⁵ and there seems to be no sexual or age preference.⁶-⁸

The syndrome is characterized by painful and difficult defecation, a sense of incomplete evacuation, and sometimes lower gastrointestinal bleeding.⁹ Although the term may be misleading, patients with SRUS do not necessarily have ulcers and if present, they are not confined to the rectal mucosa and may differ in size and form, from a patchy erythematous lesion to polyps, and well-developed ulcers.⁸,¹⁰ Hence, it is the combination of endoscopic findings, histologic properties and patients’ complaints that will lead physicians to the diagnosis of SRUS.
The vague definition of SRUS, its wide range of endoscopic findings and overlapping symptoms, make it hard to distinguish this somehow rare condition from more serious diseases such as inflammatory bowel disease. The present article will review different aspects of SRUS, providing thorough insight into the disease, which might help improving diagnosis and treatment of the condition.

Pathogenesis
The underlying mechanisms leading to SRUS are not certainly recognized. However, various factors are considered to have a role. Here we propose four main categories known to be involved. First, prolonged straining during defecation, which might end in direct trauma to the vulnerable rectal mucosa. Patients who have constipation or anatomical anomalies are more prone to suffer from straining and at higher risk for developing rectal ulcers. It is also reported that patients with solitary rectal ulcers are more likely to have increased resting and squeezing pressure than normal individuals. This may end in higher overall pressure burden bearded upon rectal mucosa and higher risk of traumatic injuries.

Second, direct instrumentation or digital trauma inflicted by patients as a mean to take out impacted fecal material will lead to traumatic injuries to rectum. This is far more common, if not limited to patients suffering from constipation. Though reasonable, this can’t be the only contributing factor since there are reports of patients having lesions much further than a finger can reach.

Disorganized contraction of puborectalis muscle in response to attempt on defecation is a well-proposed etiology, which result in higher pressures in anorectal area. During defecation, the perineum would descend to straighten the canal. Inappropriate puborectalis contraction during this process or defecation in squatting position will lead to traumatic compression of rectal wall against anal canal. On the other hand, the contradictory force of this paradoxical movement causes mucosal ischemia and renders the area more susceptible to traumatic injury and ulceration.

The excessive pressure generated as the aggregate result of constipation, straining and puborectalis aberrant contraction may end in rectal prolapse and intussusception in the long run. Even the first tiny areas of intussusception can cause vascular injury and further compromise blood supply of the mucosa, which is the last part of ulcer formation and development.

Apart from these explanations, some unknown factors may also be involved. For example, there is a case report of a solitary rectal ulcer in a woman resolving during pregnancy and recurring after delivery, which may point to a hormonal factor. Failure of corrective surgery to resolve symptoms of SRUS brings about another uncertainty in understanding possible factors involved in the pathogenesis of this condition. Overall, it seems that chronic mucosal injury and ischemic trauma are the main mechanisms though further studies may shed more light on the process.

Clinical presentation
Solitary rectal ulcer syndrome (SRUS) may typically present with anal pain, rectal bleeding and constipation. Accompanying symptoms also include tenesmus, excessive straining during defecation, mucus discharge, pelvic discomfort and incomplete evacuation. Up to one fourth of the patients may be asymptomatic. More than half of patients suffer from constipation; however diarrhea is seen in 20-40%.

The most common symptom is rectal bleeding, the amount of which is different according to patients’ condition. Hematochezia may range from blood streaks over stool to a gross hemorrhage requiring transfusion or emergency diagnostic workup. Remained untreated, rectal prolapse would be the final presenting symptom. Psychologic problems, especially obsessive-compulsive disorder, may be present in some SRUS patients.

SRUS, as the name would express, is usually acknowledged as a single rectal ulcer but the lesion may actually vary from an erythematous patch to multiple well-developed polyps. This presentation along with the various symptoms mentioned above is sometimes misleading to a diagnosis of inflammatory bowel disease and needs special attention. Digital manipulation to remove impacted stool may also cause mucosal injury and induce mucosal breaks.

Diagnosis
The diagnosis of solitary rectal ulcer syndrome is based on clinical, endoscopic and histopathologic aspects. Since the two former categories are not so specific, the diagnosis should be confirmed by hist-
Endoscopy
The endoscopic findings vary from simple mucosal erythematous patch to a solitary or multiple ulcers. However, endoscopy may also reveal non-ulcerative polypoid or mass lesions. Ulcers range in size from 0.5 to 4 cm and most of them are located on the anterior rectal wall, up to 10 cm from the anal verge. Rarely there might be some ulcers located in the anal canal or even the sigmoid colon.

Defecography
Uncoordinated contraction of puborectalis muscle and defecation problems are one of the main aspects of SRUS. In patients with defecation disorders, defecography can detect the functional anorectal components involved in pathogenesis of SRUS. In one study of 43 patients with solitary rectal ulcer syndrome, only two patients had normal defecogram with the most common finding being intussusception. In another study, 55% of SRUS patients had abnormal defecography including anterior or posterior rectocele, non-relaxing puborectalis muscle, prolonged contrast retention, rectal intussusception and megarectum.

Defecation dys-synergy should be diagnosed based on three measures: Presence of constipation, inappropriate pattern of defecation on manometry or electromyography, and other evidence of colorectal dysfunction including abnormal defecography, delayed metallic marker elimination or abnormal balloon expulsion test. The latest ROME-IV criteria for diagnosis of dyssynergic defecation is shown in table 1.

Endoscopic ultrasound (EUS)
EUS findings in patients with solitary rectal ulcer syndrome include thickening of rectal wall and internal anal sphincter as well as intussusception. Multiple submucosal cysts, hyperechogenic bands of fibrosis in the submucosal layer without disruption of the outer hypoechogenic layer (muscular layer) and regional lymph node infiltration are other possible findings. Patients with SRUS and abnormal balloon expulsion test may have thicker internal anal sphincter on EUS. The ratio of external anal sphincter to internal anal sphincter thickness is usually reduced in patients with SRUS.

Histopathology
Typical features of rectal ulcers in SRUS are fibromuscular obliteration in lamina propria along with hypertrophic and disrupted muscularis mucosa and distorted crypt structure. Villiform structures are apparent in polypoid lesions and gland entrapment in submucosa is sometimes reported, a condition called colitis cystica profunda. Histologic findings of a study on samples from 115 patients showed superficial ulcers, cryptic distortion, and inflammatory properties in 59%, 17%, and 33%, respectively.

The observation of collagen infiltration in lamina propria is a key feature differentiating SRUS from IBD or chronic colon ischemia. Since many malignant lesions may initially present as a single superficial ulcer or polypoid lesion, biopsies should be taken to rule out neoplastic infiltration. Though, the two might also be seen synchronously.

Differential diagnosis
It is important to distinguish solitary rectal ulcer syndrome from other disorders which may have similar clinical presentations and endoscopic appearance. The differential diagnosis of SRUS includes rectal cancer, idiopathic inflammatory bowel disease, infectious diseases, rectal endometriosis, and drugs.
**Treatment**

Treatment of SRUS is based on pathophysiology, the severity of symptoms, type of SRUS and presence of rectal prolapse.\(^5\)\(^,\)\(^26\) Patient education and behavior modification are the first and main steps in the treatment of SRUS. Behavior therapy such as biofeedback therapy (BFT) teach you how you can relax your pelvic floor muscles and external anal sphincter (EAS) during bowel movement.\(^28\)

Asymptomatic patients benefit from BFT and lifestyle changes. These may include high-fiber diet, drinking enough water (non-carbonated and caffeine-free beverages) during the day, regulation of toilet time, treatment of psychological problems and prevention of straining and anal digitation.\(^45\) BFT and lifestyle changes in combination with stool softeners and bulks laxatives may help patients with mild to moderate symptoms without significant rectal mucosal prolapse.\(^46\) Jarrett and colleagues found that 12/16 (75%) of patients with SRUS had subjective improvement and endoscopic healing after BFT.\(^47\)

Improvement of clinical symptoms do not mean endoscopic healing.\(^48\) People with more severe signs and symptoms may require medical or surgical treatment. Topical treatments such as sucralfate, 5-ASA, sulfasalazine, and corticosteroid enema have been reported to be effective by reducing inflammation and preventing irritant injury.\(^49\)\(^,\)\(^52\) More controlled studies and long-term follow up are needed to confirm topical agent effects on ulcer healing.

Surgery is recommended for refractory cases of SRUS despite lifestyle changes and medical treatment or for the patient with full thickness rectal mucosal prolapse.\(^53\) There are multiple types of SRUS surgery. Rectal prolapse surgery (rectopexy procedure), surgery to remove the ulcer (Delorme procedure) or rectal excision (Altemeir perineal proctectomy). Patients with paradoxical rectal spasm (PRS) and complete internal prolapse were treated by stapled transanal local excision (STARR). diversion colostomy is used in patients who have failed other surgical methods.\(^31\) Careful patient selection for surgery is extremely important and cannot be overemphasized. Most patients do not benefit from surgery and sending the inappropriate patient to surgery may lead to catastrophes.

Scleratherapy by injection into the submucosa or retro rectal space with 5% phenol, 30% hypertonic saline or 25% glucose and perianal cerclage has been also reported to be effective in some patients with rectal prolapse.\(^54\) Endoscopic application of human fibrin sealant is useful in the treatment of SRUS.\(^55\) Therapeutic transcutaneous needle injection of botulinum toxin into the external anal sphincter (EAS) is a novel treatment for difficult defecation and anal fissure and may help to resolve pathophysiology of SRUS.\(^56\)

A summary of a proposed clinical pathway for management of a patients with SRUS is depicted in figure 1.
Case management

No abnormal finding was found in abdominal examination. Digital rectal examination was done with local anesthetics and no mass lesion was detected. Total colonoscopy was performed which revealed normal rectal mucosa and a 1.5 x 1.3 cm ulcerative polypoid lesion was seen in the anterior wall of rectum approximately 9 cm from anal verge. Multiple biopsies were taken.

Histopathology report showed superficial mucosal hemorrhage with fibroblasts and smooth muscle obliterated the lamina propria. Edema and thickening of the muscularis mucosa was also present. No evidence of dysplasia or malignancy reported. All of these characterized a case of SRUS.

Bulk laxatives were started and the patient was advised to follow a high fibre diet with plenty of fluid consumption. Six sessions of biofeedback therapy was scheduled and treatment with 5-ASA was also initiated. She returned to the clinic after 3 months with no improvement in her symptoms. Thus, she was referred to a gastrointestinal surgeon who requested an anorectal manometry. Dysynergic contraction of the puborectalis muscle compatible with type 4 dysynergic pattern of defecation was evident in manometry and she underwent surgery using Delorme procedure.

Her post-operative care was ordinary and she was sent home with emphasis on continuing dietary modifications to avoid excessive strain during defecation. There was no complaint in the 3-month follow-up visit and the ulcer was healed in sigmoidoscopy.

ETHICAL APPROVAL

There is nothing to be declared.

CONFLICT OF INTEREST

The authors declare no conflict of interest related to this work.

REFERENCES