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An Interesting Case of Mistaken Identity

S.B. Umar J.E. Efron R.I. Heigh

Mayo Clinic Arizona, Scottsdale, Ariz., USA

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Solitary rectal ulcer · Inflammatory bowel disease · Defecatory disorders

Abstract
Solitary rectal ulcer syndrome (SRUS) is an uncommon disorder which can present in patients being evaluated for defecatory disorders or which can present as a primary process often involving hematochezia, rectal pain and tenesmus. Unfortunately the diagnosis of this disorder is often delayed due to misdiagnosis and/or physician unfamiliarity with the condition. We present a 24-year-old female who presented with 6 months of bloody diarrhea and weight loss. She had been receiving treatment for a presumed diagnosis of inflammatory bowel disease (IBD) due to an endoscopic picture of rectal thickening, edema and ulceration and had been on prednisone for 2 months prior to presentation without relief of her symptoms. After further testing including repeat endoscopy with biopsies, defecography and anorectal manometry, the diagnosis of SRUS was made and treatment was changed. Medical management was unsuccessful and she ultimately required surgical intervention. This case highlights the difficulty in diagnosing SRUS due to its resemblance to other gastrointestinal diseases and should serve as a reminder that if a patient is not responding to IBD therapy, another etiology should be considered.

Introduction

Solitary rectal ulcer syndrome (SRUS) is an uncommon disorder, with a reported prevalence of 1:100,000 adults [1]. SRUS is characterized by symptoms such as hematochezia, rectal pain, rectal fullness and tenesmus. Additionally, SRUS can be asymptomatic and found secondarily in patients undergoing surgery for defecatory disorders such as rectal intussusception and rectal prolapse. The diagnostic tools used include anorectal manometry, defecography and endoscopy with biopsies of the affected area. Unfortunately, however, the diagnosis of SRUS is often significantly delayed, and in many studies the average time from onset of symptoms to diagnosis was between 4 and 8 years [2–4]. This delay is likely due to the fact that SRUS can mimic other gastrointestinal conditions such as neoplasm and inflammatory bowel disease (IBD) [2].

Sarah B. Umar
Mayo Clinic Arizona, 13400 E Shea Blvd
Scottsdale, AZ 85259 (USA)
Tel. +1 480 342 2000, Fax +1 480 301 8673, E-Mail umar.sarah@mayo.edu

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We present a 24-year-old female with no prior gastrointestinal complaints who complained of 6 months of bloody diarrhea and weight loss. The patient had been given a diagnosis of IBD prior to presentation and had been receiving prednisone and a 5-ASA medication for 2 months with no relief of her symptoms. After testing she was diagnosed with SRUS and medical therapy was started. Unfortunately she continued to have difficulties with hematochezia and ultimately required surgical intervention which resolved her symptoms.

Case Report

A 24-year-old previously healthy female was seen in consultation for complaints of 6 months of bloody diarrhea. She was having between five and ten episodes daily with tenesmus and fecal urgency. There was no fecal incontinence. The stool was mixed with frank blood and clots and on occasions she would pass blood without stool. She had no prior history of gastrointestinal symptoms such as constipation or bloody stools. Her family history was notable for lymphoma and the absence of IBD or other gastrointestinal conditions. Also noted was a 15-pound weight loss since the onset of her symptoms accompanied by fatigue and malaise. She denied fevers or chills and remainder of review of systems was negative.

Prior to being seen in our clinic, she had undergone colonoscopy elsewhere that noted an ulcerated, 'apple-core' lesion at 10 cm from the rectal verge. Biopsies were interpreted as ulceration consistent with an inflammatory polyp; no dysplasia was seen. The remainder of the colon was normal. She was then prescribed oral mesalamine as well as cortisone enemas as treatment for IBD. These therapies did not improve her symptoms and she was ultimately started on prednisone 20 mg p.o. b.i.d. She had been on steroids for approximately two months with no improvement before presentation to our institution.

We repeated a colonoscopy showing her rectum to contain pseudopolyps within a nodular, friable, ulcerated rectal mucosa extending from 5 to 10 cm from the anal verge (fig. 1). Endoscopic ultrasound of the affected area was performed and showed that the area was hypoechoic and diffusely thickened to the muscularis propria. Anorectal manometry was performed which showed no evidence for pelvic floor dyssynergia, but evidence of rectal hypersensitivity was present. A defecating proctogram was performed which showed a nodular prominence posteriorly overlying the sling musculature measuring approximately 2 cm in diameter. This segment appeared to prolapse out of the anal opening on evacuation. Colonoscopic biopsies showed eroded hyperplastic mucosa with fibromuscularization of the lamina propria and hyperplastic glandular changes (fig. 2), findings consistent with SRUS.

Prednisone was discontinued and topical management initiated with hydrocortisone foam and sucralfate enemas. Cholestyramine was used to control her diarrhea. She continued to bleed, however, and required outpatient blood transfusion. Ultimately she was referred to colorectal surgery where a rectopexy was performed. After 8 months of follow-up she was doing well without recurrence of her symptoms.

Discussion

SRUS is an uncommon disorder; one study cited an incidence of 1:100,000 adults [1]. It is largely a disorder of younger adults, more than 80% of patients in the literature are under the age of 50. The name of this disorder is in itself misleading, since often the lesions are neither solitary nor ulcerated. Most lesions occur on the anterior rectal wall within 10 cm of the anal verge. The most common presenting symptoms are rectal bleeding, straining and altered bowel habits. Often a careful history will elicit prior symptoms of constipation and/or description of rectal prolapse. SRUS is unfortunately a disorder that often goes undiagnosed for prolonged periods of time due to misdiagnosis and lack of awareness of the condition by physicians. Tjandra et al. [2] examined 98 patients with SRUS and found that 26% of them had been initially misdiagnosed, with a median duration of incorrect diagnosis of 5 years. The most common misdiagnoses were

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IBD and neoplasm. Nearly one-third of these patients had been on high doses of prednisone for more than 3 months prior to SRUS diagnosis.

The diagnosis of SRUS is made using a combination of diagnostic tools including a careful history and physical exam in concert with endoscopy and biopsy. Anorectal manometry and defecography will often be obtained to support the diagnosis and/or to understand the etiology of the patient's symptoms. Endoscopy can reveal a large variety of findings extending from mild mucosal erythema to multiple, large ulcerations in the rectum. These ulcers can measure up to 4 cm but usually are less than 1.5 cm. Polypoid lesions have also been observed [5]. Histology is important in making a correct diagnosis of SRUS. Characteristic microscopic findings include the presence of mucosal architectural distortion without inflammation; this lack of inflammation differentiates SRUS from IBD. Additionally obliteration of the lamina propria with fibromuscular tissue is a hallmark of SRUS. Defecography can reveal rectal intussusception and/or prolapse. Anorectal manometry is performed to evaluate for concomitant or other defecatory disorders such as dyssynergic defecation.

The pathogenesis of SRUS is poorly and incompletely understood. It is thought that a combination of rectal prolapse and paradoxical puborectalis contraction leads to the generation of high pressures within the rectum, leading to ischemia and, ultimately, ulceration. Additionally, paradoxical puborectalis contraction creates shear forces on the rectal mucosa, and this is thought to be another etiologic factor in this disorder. These are simply theories, however, as not all patients with SRUS have evidence of paradoxical contraction and there have been cases where correction of rectal prolapse does not resolve the symptoms, leading to a supposition that rectal prolapse is not a causative but rather a concomitant process associated with SRUS.

Treatment of the SRUS includes medical, surgical and behavioral options. Medical therapeutic options include systemic and topical medications such as corticosteroid and sucralfate enemas [6, 7], human fibrin sealant [8] and sulfasalazine, both orally and topically [9, 10].

Biofeedback has shown some success in SRUS. Biofeedback improves coordination of the pelvic musculature in combination with anal relaxation techniques to minimize the shear stress and potential for ischemia in the rectum that was due to dyssynergic defecation. Rao et al. [11] performed a prospective trial of biofeedback therapy in 11 patients with medically refractory SRUS. In this population, 82% of patients showed evidence of pelvic dyssynergia as well as higher anal pressures and lesser anal relaxation with straining compared to a population of 15 healthy controls. After a 12-week period of biofeedback therapy, none of these patients re-demonstrated dyssynergic defecation, and anal residual pressures and percentage of anal relaxation both significantly decreased. On repeat endoscopic evaluation, 54% of patients had ≥50% ulcer healing. Biofeedback has also been shown to improve rectal blood flow via Doppler mucosal flowmetry, and this improvement was correlated with subjective improvement in symptoms and endoscopic improvement in ulceration [12].

Unfortunately not all patients respond to medical or behavioral intervention and some will require surgical intervention. Common surgical techniques include transabdominal rectopexy, Delorme procedure (transrectal mucosal stripping with mucosectomy), low anterior resection and occasionally colostomy. Choi et al. [13] reviewed 49 patients with histopathologically diagnosed SRUS and found postoperative symptomatic improvement in approximately 70% of patients. Transabdominal rectopexy and perineal rectosigmoidectomy comprised the majority of surgical procedures performed in this

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series. In another series involving 66 patients undergoing surgery for SRUS, rectopexy was the predominant technique performed with a success rate of approximately 55%. This study also identified fecal incontinence and inability to complete evacuation as preoperative predictors of poor surgical outcome [3]. Other retrospective studies have been performed with similar results, and it is usually recommended that patients undergo a trial of medical management before undergoing surgery [14].

Conclusion

SRUS is an uncommon disorder which is often mistaken for other conditions such as IBD or neoplasm. The diagnosis of SRUS can be significantly delayed. Medical, surgical and behavioral therapies have all been used and it is generally accepted that before proceeding to surgery, patients undergo a trial of medical and, if appropriate, behavioral therapy in the form of biofeedback. Topical therapies include glucocorticoids, sucralfate and mesalamine compounds; however efficacy is limited. If evidence of dyssynergic defecation is present, biofeedback has shown promising results. Physicians should be aware of this diagnosis, especially in patients who are not improving with conventional therapies directed towards IBD.

Fig. 1. Endoscopic appearance of an approximately 5 cm area within the rectum showing pseudopolyps within a nodular, edematous, ulcerated mucosa.

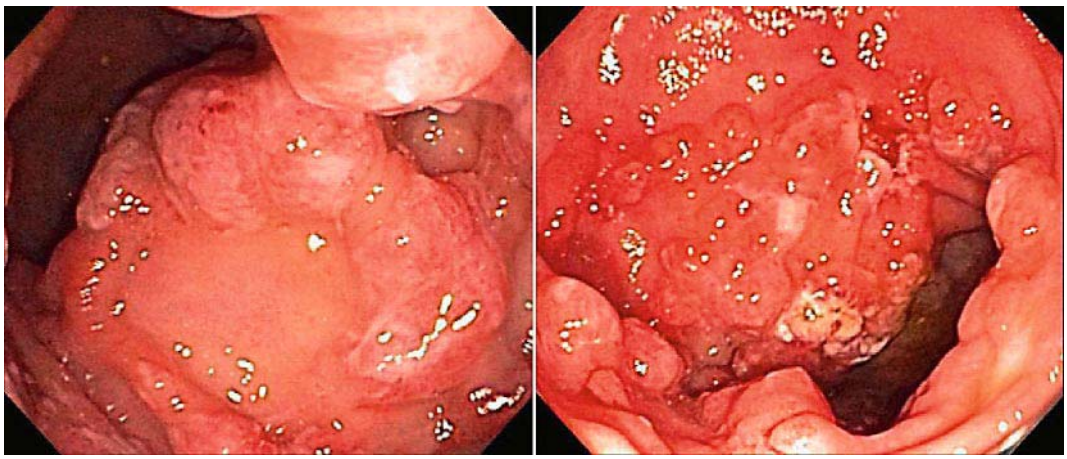

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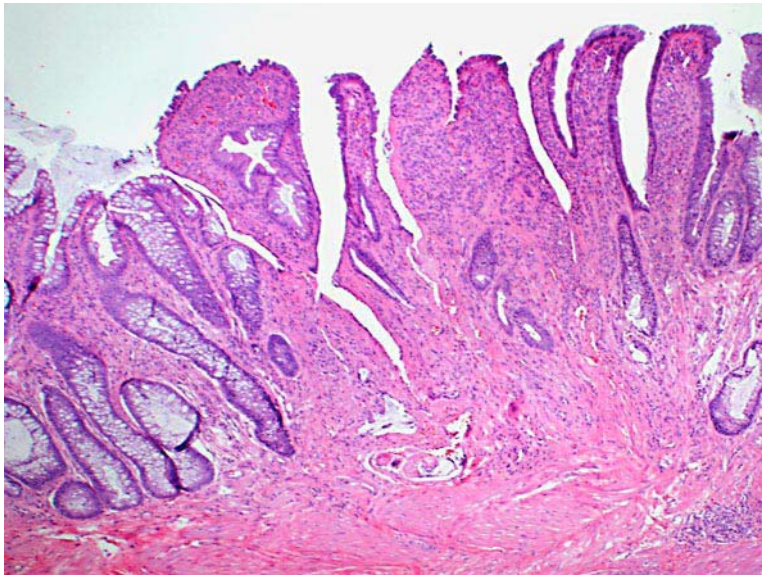
Fig. 2. Pathology showing eroded hyperplastic mucosa with fibromuscularization of the lamina propria and hyperplastic glandular changes.



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