Review Article

Solitary Rectal Ulcer Syndrome: An Unusual Cause of Per Rectal Bleeding in Children

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Abstract

Solitary rectal ulcer syndrome (SRUS) is a relatively uncommon but troublesome chronic benign disorder more in young adults and less in children. It is usually presented as rectal bleeding, constipation, copious mucous discharge, prolonged excessive straining, tenesmus, lower abdominal pain, localized pain in the perineal area, feeling of incomplete defecation and rarely rectal prolapse.The underlying etiology is not well known. It is diagnosed based on symptoms and endoscopic and histological findings.

Key words: *Solitary Rectal Ulcer Syndrome; Children; Rectal Bleeding*

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Introduction

Solitary rectal ulcer syndrome is a rare, benign rectal disorder in children but well recognized in adult populations.[1−5] It usually presents with rectal bleeding and/or mucous discharge associated with prolonged straining during defecation and a sense of incomplete evacuation, constipation, tenesmus, lower abdominal pain and localized pain in the perineal area.[6,7] Cruveilhier in 1829, described four unusual cases of rectal ulcers.[6] The term "solitary ulcers of the rectum" was used by Lloyd-Davis in the late 1930s.[7] The term solitary rectal ulcer syndrome is a misnomer because i) ulcers are found in 40% of patient, others having single or multiple ulcers including hyperemic mucosa to broad-based polypoid/mass lesions ii) 20% of patients have a solitary ulcer, other lesions are different in shape and size and iii) may involve the sigmoid colon.[8,9] The underlying etiology is not well understood.[8,10] It often goes unrecognized or easily misdiagnosed with inflammatory bowel diseases, amebiasis, malignancy and juvenile polyp.[10−12]

1. Epidemiology

Regarding pediatric age group, there are very limited data. From adult study, SRUS is an infrequent, unrecognized or misdiagnosed disorder with an estimated prevalence of about 1 in 100000 persons per year.[1] In children, median age group are 10 years among them maximum are above 8 years and range from 18 months to 18 years. It has somewhat male predominance with male- female ratio 1.4:1.[13]

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2. Pathogenesis

The pathogenesis is incompletely understood. Various factors may be involved in its disease process. Most accepted theories are factors associated with direct trauma or causes of local ischemia. Factors for trauma or local ischemia are (1) Straining: prolonged straining during defecation in the patient who suffers from constipation may result in a direct trauma to the mucosa. (2) Self-induced trauma: Direct trauma during attempts at manual disimpaction (3) Paradoxical contraction of puborectalis muscle: Pressures generated by the rectum during defecation causing rectal mucosa to push downward. The opposing force of the paradoxical contraction of the puborectalis muscle can generate high pressures within the rectum which entrapped the prolapsed mucosa and lead to mucosal ischemia thus predisposing to ulceration. (4) Rectal prolapse and intussusception: Rectal intussusceptions can lead to localized vascular trauma and then onset of solitary local ulceration.[14–16] Rectal hypersensitivity causing persistent desire to defecate and sensation of incomplete evacuation may also have a role in SRUS.[14] A possible hormonal cause has also been proposed.[17]

3. Clinical manifestations

Patient may asymptomatic in up to 26% cases and symptoms were mostly rectal bleeding (56%), straining with defecation (28%) and pelvic fullness (23%) mucous discharge, incontinence, tenesmus, and rectal pain, abdominal pain, constipation, and rectal prolapsed, sense of incomplete evacuation.[8,9,13,18,19,20–22] The amount of blood varies from a little fresh blood to severe hemorrhage that requires blood transfusion. Digital manipulation during defecation invariably reported in patients with SRUS. [23,24]

4. Evaluation

Diagnosis is done via combination of symptom with colonoscopic and histological findings.[25] High index of suspicion for the possibility of SRUS in young children with non-specific clinical picture of proctitis makes diagnosis easier.

Colonoscopy — The microscopic appearance of solitary rectal ulcer syndrome, is supportive for diagnosis but not specific.Macroscopic findings range from mucosal erythema to single or multiple ulcers,small or giant ulcers and broad based polypoid/ mass lesions of different sizes.[8,27] Ulcers are usually superficial and 1 to 1.5 cm in diameter, but may 0.5 to 4 cm (Fig 1). Lesions usually are located in the anterior rectal wall within 10 cm of the anal verge, but they can also be located in the anal canal or the sigmoid colon. SRUS typically appears as shallow ulcerating lesions on a hyperemic surrounding mucosa.[8,27]

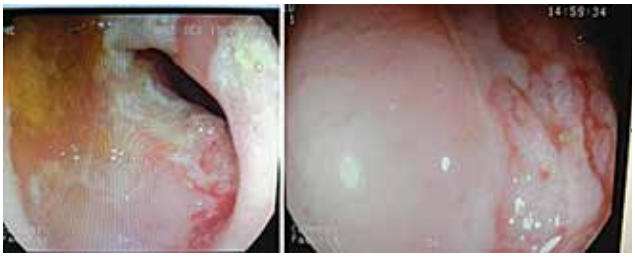

Fig 1. Colonoscopic findings of SRUS

Microscopy: On histology, fibro muscular obliteration of the lamina propria that leads to hypertrophy and disorganization of the muscularis mucosa, streaming of fibroblasts and muscle fibers between crypts, branching and distorted glandular crypts and diffuse collagen infiltration of the lamina propria are

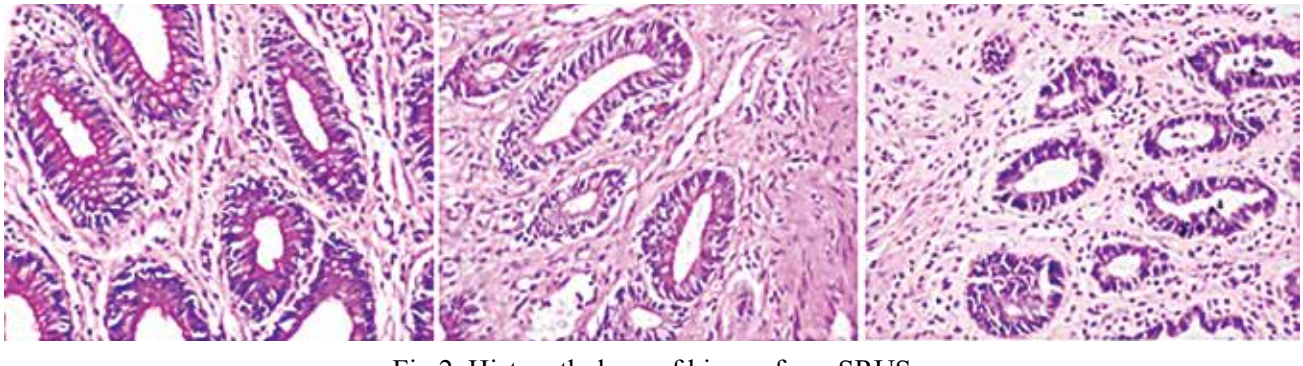

Fig 2. Histopathology of biopsy from SRUS

diagnostic finding (figure 2).[7] In polypoid cases, the mucosa has a villiform configuration, and in some cases, the glands may be trapped in the sub-mucosa (colitis cystica profunda).[28]

Defecography — with persistent symptoms despite initial management, defecography is a useful method for determining the presence of intussusception or internal or external mucosal prolapse and can demonstrate a hidden prolapse, a non-relaxing puborectalis muscle and in complete or delayed rectal emptying.[29] The most common findings are internal and external rectal prolapse and delayed rectal emptying.[30]

Anorectal manometry should be performed in all children with SRUS to define the primary patho-physiological abnormality and to select the most appropriate treatment protocol. Barium enema may be normal or may have nonspecific findings shows granularity of the distal rectal mucosa, polypoid lesion, rectal stricture, rectal ulcer ratio and thickened rectal folds.[26–29]

5. Differential diagnoses

Inflammatory bowel disease, amebiasis, lymphogranuloma venereum, chronic ischemic colitis, colitis cystic profunda, malignancy, anal fissure and juvenile polyp.[13,31,32]

6. Management

Treatment of SRUS is difficult and mostly practiced management includes general measures, bio feedback therapy, pharmacotherapy and surgery for selected cases.[33–37]

General measures: All patients should first be treated conservatively. Conservative treatment is effective in patients with mild to moderate symptoms and absence of significant mucosal prolapse. Must give reassurance that the lesion is benign and advice to avoid straining or anal digitation, regulation of toilet habits, decrease time on the commode, consumption of a high-fiber diet and bulk laxatives.[17,33,32]

Bio feedback: If general measures fails to relieve symptom then biofeedback therapy for symptomatic improvement.[1,38,39] Biofeedback is known as a variety of behavioral changes that acts by correcting pelvic floor dyssynergia, reduces excessive straining associated with defecation and stopping the use of suppositories and laxatives.[39]

Pharmacotherapy:In children, primary medical treatment is advised for most cases. Topical application of sucralfate enema can be effective for treatment of SRUS in some patients.[40,41] Medications that are useful in inflammatory bowel disease patient also have been tried in those with SRUS, such as sulfasalazine and topical glucocorticoids, and mesalamine has been described in small series of patients with varying responses.[10,33–35]

Surgery: Surgery is reserved for patients with severe symptoms that are unresponsive to initial management. Surgical options include excision of the ulcer, treatment of rectal prolapse or rectal intussusception, and defunctioning colostomy.[26]

Refractory or recurrent symptoms after surgery: should be offered biofeedback again, who fail to respond to biofeedback again may be offered salvage surgical treatment depending on the severity of symptoms. Mucosal sleeve resection with coloanal pull-through or fecal diversion is reserved for patients who have failed other surgical modalities. Patients with intractable rectal pain and/or bleeding require proctectomy.[32]

Solitary rectal ulcer syndrome significantly impacts quality of life. Diversity of the clinical presentation of SRUS requires a high index of suspicion for diagnosis. Most patients with SRUS in childhood have a satisfactory outcome using a simple behavioral modification approach. Ongoing follow-up to reinforce behavioral modificationis important and may avoid long-term, treatment resistant disease into adulthood.

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