

RESEARCH PAPER

Solitary Rectal Ulcer Syndrome – An Uncommon Cause of Rectal Bleeding in Children

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Abstract

Background: Solitary rectal ulcer syndrome (SRUS) is a benign chronic disorder rarely found in children commonly presents with rectal bleeding, mucorrhea, straining during defecation, tenesmus, feeling of incomplete defecation, rectal prolapse and constipation. The pathogenesis is not clearly understood but several factors such as trauma, rectal prolapse, ischemia, behavioral disorders such as excessive straining during defecation and rectal manipulation, sexual abuse and disharmony of the pelvic floor muscles during defecation may be involved. SRUS is diagnosed on the basis of clinical symptoms and endoscopic and histological findings. Various treatment protocol for SRUS have been described such as conservative management such as family reassurance, regulation of toilet habits, avoidance of straining, encouragement of a high-fiber diet, topical treatments with salicylate, sulfasalazine, steroids and sucralfate, and surgery.

Methods: A comprehensive search is done using PubMed, Scopus, Google scholar and MEDLINE.

Results: The result of this review will be published in a journal.

Conclusion: This study discusses about the pathogenesis, clinical features, diagnosis and various treatment strategy of solitary rectal ulcer syndrome in children.

Keywords: Rectal bleeding, Solitary rectal ulcer, Colonoscopy, Children, Constipation.

Introduction

Solitary rectal ulcer syndrome (SRUS) is a benign chronic disorder which is found rarely in children. The syndrome is relatively more common in adult with an estimated incidence of 1 in -100,000.¹

In year 1829, the first case of solitary rectal ulcer syndrome was reported by Cruveilhier, where he described it as an unusual rectal ulcer.² Later on in 1930, Lloyd-Davis used the term “solitary ulcers of the rectum” and in 1969 Madigan et al. reported total 68 cases of SRUS and the disease became well documented.^{3,4}

The pathophysiology of SRUS is not completely understood. In children, this syndrome is associated with chronic mechanical and ischemic trauma, pelvic floor disorder, mucosal prolapse.⁵

Due to variable clinical presentations which has similarity with other rectal diseases, it can be easily confused with other causes of rectal bleeding such as juvenile polyp, inflammatory bowel disease, amebiasis, vascular ectasia.⁶

As SRUS is an unrecognised disorder, it requires high index of suspicion for diagnosis in young children.

There are multiple opinions regarding treatment of this troublesome condition ranging from conservative management and topical preparations to surgical procedures such as rectopexy.⁷

SRUS is a rare disease of childhood, very limited number of studies are done so far and most of them are case reports.

In this review article, risk factor, pathophysiology, various clinical presentations and treatment methods of SRUS in children will be focused on.

Pathophysiology: Although the exact pathophysiology is not known exactly, but several factors have been identified in different studies such as trauma, pelvic floor disorder, mucosal prolapse, nonrelaxing pubo-rectalis muscle that could be the cause of

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ulceration in the rectum.^{8,9,10} In a study conducted by Dehghani et al reported that more than 90% patients of SRUS had history of straining during defecation.¹¹

This goes in favour of the postulated mechanism that excessive straining during defecation causes high intra-abdominal pressure leading to anterior rectal wall prolapse into the contracting pubo-rectalis muscle and then anal canal, which subsequently leads to entrapment of rectal mucosa causing congestion, edema, and then hypo-perfusion, ischemia, ulceration.¹²

Digital manipulation causing trauma to rectal mucosa may also be predisposing factor for development of SRUS.¹³

De la Rubia et al have noticed that ischemia may be the reason of rectal ulceration. Persistent contraction of puborectalis muscle during defecation may cause an occult rectal prolapse which causes the ischemia.¹⁴

According to Womack et al the pressure gradient between the intra-abdominal pressure and intra rectal pressure may have a role in development of SRUS. The high pressure gradient due to prolapsed rectum causes rupture of submucosal vessel and mucosal necrosis.¹⁵

However, none of the above mentioned hypothesis can fully explain the development of SRUS.

Clinical Manifestations

In children, as the clinical presentation varies from patient to patient, proper knowledge and awareness is required for diagnosis of SRUS.

Although there is no significant male or female predominance found in adult patient with SRUS, but in pediatric age group, 75%-80% of children with SRUS are boys.¹⁶

The most common age of presentation is after 5 years. In most of the study, the average time interval found between onset of symptoms to diagnosis is 5 years (ranging from 1.2 to 5.5 years).^{17,18}

Patient may remain asymptomatic in up to 25% of cases which is diagnosed incidentally.¹⁹

Fresh rectal bleeding, mucorrhea, straining during defecation, tenesmus, feeling of incomplete defecation, rectal prolapse and constipation are the major symptoms.^{20, 21} However obtaining a detail history regarding prolonged straining, manual evacuation of stool is very crucial for diagnosis of SRUS.²¹

Occasionally patient may present with severe bleeding which may require blood transfusion.^{22, 23}

In a study conducted by Dehghani et al in 55 children with SRUS also found similar clinical features like rectal bleeding in 98% cases, excessive straining in 91% cases.¹¹ In a large cohort study of 140 children with SURS done by Poddar et al, the predominant manifestations were found rectal bleeding (93.6%), with features of dyssynergic defecation such as prolonged sitting in the toilet (93.6%), excessive straining (98.6%), and a feeling of incomplete evacuation (92.8%).²⁴

Diagnosis

SRUS is easily misdiagnosed condition of childhood. So high index of suspicion is required for both the clinician and the pathologist and diagnosis is done on the basis of symptomology in combination with endoscopic and histological findings.²⁵

Despite of being a benign condition, it needs prompt diagnosis to avoid long term consequences such as anemia secondary to massive rectal bleeding, poor appetite or distress to other family member.³

It should be differentiated from other causes of rectal bleeding such as juvenile polyp, inflammatory bowel disease, amebiasis, infectious proctocolitis, intussusception, hemorrhoids, prolapsing rectal polyp, or sexual abuse.^{26, 27}

In pediatric age group, detail investigations for SRUS are done in few cases.^{28, 29}

Colonoscopic examination with histopathology is the "gold standard" in the diagnosis of SRUS and it can clearly differentiate SRUS from IBD (more inflammation and less of fibrosis) and tumor.²⁵ The typical colonoscopic finding is shallow ulcerating lesions on a erythematous surrounding mucosa, most often located on the anterior wall of the rectum at 5 to 10 cm from the anal verge, ranging from 0.5 to 4 cm in diameter.^{18, 19} But as SRUS is a disease of misnomer, the lesion is solitary only in 20 % of patients, in other cases it can be multiple or circumferential, it may not be ulcerated rather polypoid or hyperaemic in nature, ulcers are found only in 40% cases and not always involve the rectum but may involve the sigmoid colon.^{19, 20}

In the largest case series conducted by Poddar et al, revealed 72% patients of SRUS had ulcer, among them 83% was solitary and all were present in rectum.²⁴

The typical histological finding of a patient with SRUS are fibromuscular obliteration of lamina propria that is lamina propria is replaced by smooth muscle and collagen which leads to hypertrophy and disorganization of muscularis mucosa, streaming of fibroblasts, and muscle fibers between crypts, branching and distorted glandular crypts, surface ulceration with minimal inflammation.²¹

Additional diagnostic procedures such as defecography, transrectal ultrasonography, dynamic MRI, or endoscopic ultrasound have recently been used for diagnosis of SRUS. Both defecography and dynamic MRI can be used for detecting intussusception or internal or external mucosal prolapse, pelvic floor dysfunction, rectocele, incomplete or delayed rectal emptying. Temiz et al have recommended that defecography and anorectal manometry should be performed in all children to determine the most suitable treatment strategy.³⁰

Ano-rectal ultrasound is a useful diagnostic procedure for assessing internal anal sphincter thickness which is highly predictive of high-grade rectal prolapse and intussusception in patients with SRUS.^{31, 32}

Treatment

There is no consensual agreement about the treatment of SRUS in pediatric population. Very limited number of studies have been conducted so far regarding management of SRUS. Different treatment regimens, both conservative and surgical have been used with various responses.

Stepwise approach about management should be attempted in children with SRUS.²⁴

Initial stage of the treatment strategy includes reassuring the parents about the benign nature of the disease, behavioural modification in the form of avoiding or reducing time for straining, biofeedback therapy, high fiber diet with stool softeners and bulking laxatives.³³

In a study conducted among 8 children with SRUS, Blackbourn et al. have noticed that behavioural modification such as avoidance of straining and digital evacuation plays an important role for relieving symptoms. In their study, rectal bleeding, mucoid stool and tenesmus improved in 88% patients.²¹ They also mentioned about the importance of maintaining compliance with behavioural modification for prevention of long term morbidity.

In another study, Dehghani et al recommended behavioural changes and dietary modification as the first line therapy of SRUS.¹⁶

Pharmacotherapy such as laxatives and stool softeners with high fiber diet can be used in SRUS if patient has hard stool.¹¹ In a case series, Thirumal et al shown that 75% of children responded to conventional treatment of laxative along with toilet training.³⁴

If the initial conservative treatment fails to resolve the symptoms, then biofeedback therapy may have an effective outcome in children. Some clinicians proposed biofeedback therapy as part of routine management of SRUS, specially in refractory cases. It includes bowel habit training, avoidance of excessive straining, and normalization of pelvic floor coordination. Biofeedback therapy is proven to improve the defecation dyssynergia and reform defecation dynamics.³⁰

If patient is refractory to first line therapy, topical agents such as Mesalamine enema, Sucralfate and steroid enema can be used in patient with SRUS.²⁴ In a prospective study among 12 children, Dehghani et al found that 58.3% patients improved with sucralfate enema.¹¹

Some studies reported the success of topical steroid if rectal ulcer is present. According to study done by Poddar et al and Kowalsk-Duplaga et al, topical application of steroid can be a good therapeutic option if ulcer is found.^{13, 24}

Both oral and topical mesalamine are also used in children with SRUS. Though their long term success rate is controversial.³⁵ Outcome of different treatment modalities is very difficult to assess, as no comprehensive study has been conducted so far.

Endoscopic therapy with various agents such as human fibrin sealant, argon plasma coagulation (APC), laser therapy are also used effectively in SRUS.^{36, 37}

Some author suggests botulinum toxin injection into the external anal sphincter as treatment of SRUS specially if the condition is associated with defecation dyssynergia and the therapeutic effect lasts for approximately 3 months which is more satisfactory than biofeedback therapy.³⁸

In children, the results of surgical treatment is still controversial.³⁰ Surgery is recommended if patient is refractory to medical therapy or presents with rectal

prolapse or who has polypoid lesions.³⁰ Before surgery, defecography should be done to detect pelvic floor dysfunction, rectocele, and rectal intussusception or prolapse.¹¹ Surgical options are rectopexy, excision of ulcer, and sometimes colostomy.^{39, 40} In children multiple ways of rectopexy have been tried, and 90% success rate has been observed in postero-sagittal rectopexy.⁴¹

Long term follow up is necessary for reinforcing behaviour modification and avoiding long- term, treatment resistant disease into adulthood.

Conclusion

SRUS is a misdiagnosed condition of childhood. There are clinicopathologic similarities between SRUS and IBD or constipation as most of the child presents with rectal bleeding and dyssynergic defecation. The diagnosis is often delayed so increased awareness of both endoscopists and pathologists are needed for early diagnosis. More prospective studies are required regarding clinical and therapeutic outcome for establishment of treatment protocols in children.

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