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CASE REPORT

Solitary Rectal Ulcer Syndrome in a Teenage Girl: A Case Report

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Introduction

Solitary rectal ulcer syndrome (SRUS) is rare and uncommon disorder of rectosigmoid region which is mostly reported in adults and is very rare in children. This disorder is diagnosed by clinical findings, colonoscopy findings and histopathological changes.[1] The cause of this syndrome is still not known and is usually caused by chronic constipation, which can be associated with excessive straining during defecation, rectal manipulation, trauma and ischemia. The condition was first described by Cruveilhier in 1830 and detailed clinical and histopathological features were reported by Madigan and Morson in 1969.[2] It occurs most commonly in the 3rd decade in men and in the 4th in women, with only a few reported paediatric cases.[3,4] The annual incidence rate of SRUS is 0.001%.[5] SRUS usually presents with rectal bleeding, constipation, mucous discharge, prolonged straining, tenesmus, and lower abdominal pain.[6] The term "SRUS" is a misnomer, and is sometimes referred to as "the three-lie disease,"as the lesion is not always solitary (it may be multiple), is not ulcerative (it may be polypoidal /nodular or affecting the erythematous mucosa only) and is not restricted to the rectum (it may also involve the sigmoid colon).[7] Histological examination is the gold standard for the diagnosis of SRUS.

Case report

A fifteen years old girl was admitted in the department of Paediatric Gastroenterology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh in middle of July, 2019 with the complaints of per rectal whitish discharge for twenty days. She also complained of per rectal bleeding and abdominal pain for same duration. The girl had history of constipation and weight loss with this illness. She was quite alright two years back. Then she developed occasional per-rectal bleeding. At the same time she suffered from intermittent lower abdominal pain. For this illness she was admitted previously in several institutions but no significant improvement occurred. Previously she was undergone colonoscopy for two times which showed nonspecific histopathological results. As Bangladesh is a tuberculosis prone country, so she was treated with anti-tubercular drug for six months. Though she had no contact with tubercular patient previuosly. Initially some improvement occurred with anti-tubercular drug but not totally subsided. Few days later she had again full blown symptoms along with mucorrhoea. She was admitted in BSMMU for second time colonoscopy. She was diagnosed as inflammatory bowel disease at that time and treated accordingly. She was transfused with two unit of blood as her hemoglobin (7.6 gm/dl) dropped down at that time. She was on tab. mesalamine and tab. steroid for one month with mild improvement. But gradually she developed further deterioration of her health. Per rectal bleeding, passage of mucous, abdominal pain became more pronounced along with gradual weight loss. In the recent admission, she was again evaluated with third time colonoscopy. Routine laboratory test results

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including blood cell counts, coagulation, hepatic function, C-reactive protein levels, and the erythrocyte sedimentation rate were normal. A stool examination for bacteria and parasites was negative, Macroscopically the whole colon including terminal ileum (Fig. 2,3) was normal but rectal ulcerations, erythema and sloughing exudates (Fig. 1) were found (5 cm from the anal verge) which was easily bled after light touching. Several biopsies were obtained from both the lesion and the ileal mucosa. Ileal biopsy was normal. A histopathological examination of rectal biopsy revealed chronic inflammatotry cell infiltration in the lamina propria and surface erosion. The crypts were diamond shaped. The muscularis mucosa was thick and thin bands of smooth muscle fibers were present in between the crypts. We took proper history of constipation and habit of straining during defecation. So, on the basis of clinical suspicion, colonoscopy and histology finding, we made a rare diagnosis of this illness as 'Solitary Rectal Ulcer Syndrome'. We managed the girl with sucralfate enema. As sucralfate enema is not available in our country, we prepared it locally by adding two tablet of sucralfate in 10 ml distilled water. Then introduced this solution per rectally twice a day for 14 days. Management of constipation with polyethelene glycol and lactulose went on side by side. All the symptoms (per rectal bleeding, passage of mucus, abdominal pain) subsided in just two weeks with this treatment. Now the girl's condition is much improved. She is on polyethelene glycol and lactulose for last two months. She was advised to take high fiber diets and also advised not to strain during defecation. She was routinely followed-up and assessed for further bleeding. As further bleeding did not occur repeat colonoscopy was postponed.

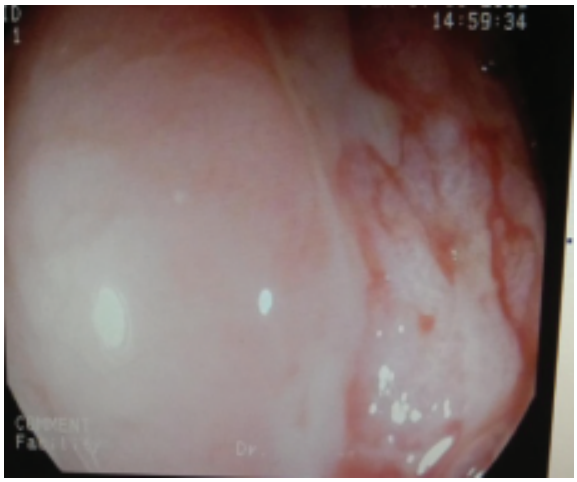

Fig 1 *Rectal ulcerations, erythema*

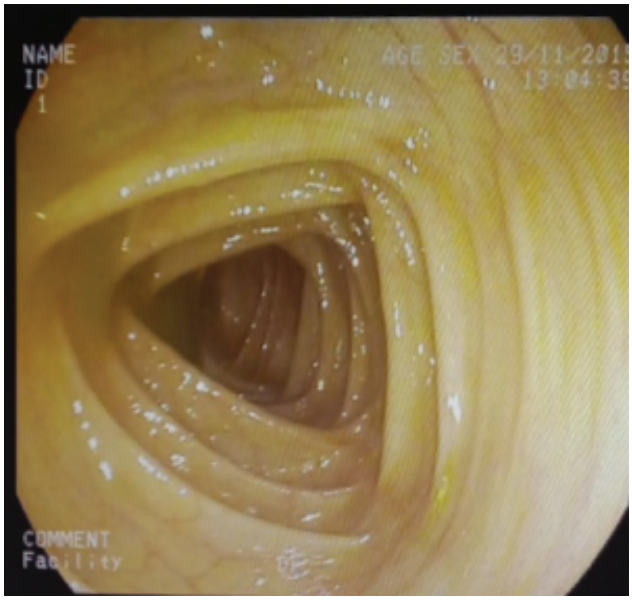

Fig 2 *Normal colon*

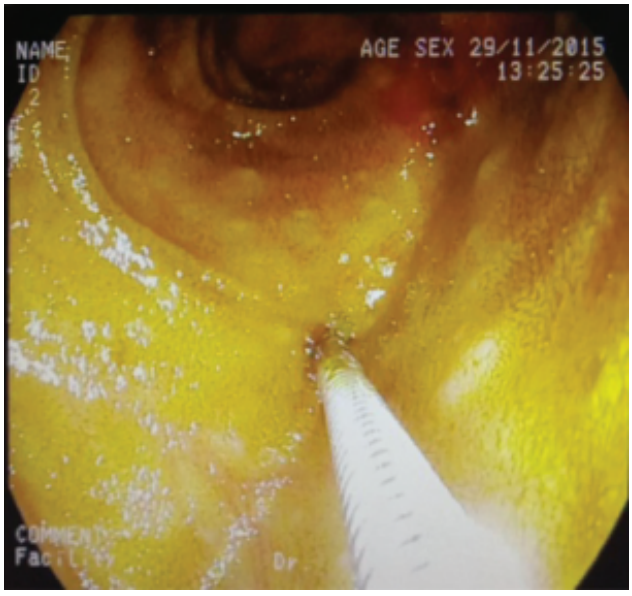

Fig 3 *Normal terminal ileum*

Discussion

SRUS is a rare benign condition of children where individuals experience difficult defecation. It is characterized by rectal bleeding, constipation, passage of mucus, mucosal prolapse, feeling of incomplete evacuation, rectal pain (tenesmus) or intermittent lower abdominal pain. The patient in the current case experienced abdominal pain, passage of mucus and rectal bleeding. The etiology of SRUS may be associated with certain lifestyles such as enema use or insertion of suppositories, self

digitation, staying in the toilet for a long time having the habit of straining during defecation, use a stream of water, or cleaning the inside part of rectal as far as possible. The patient of the current report had the habit of straining during defecation. Her diet was lack of fruits and vegetables which is rich in dietary fibers and had constipation for last few years, which is a risk factor for SRUS. Ischemic injury of rectal mucosa is recognized as the main mechanism of SRUS. The reasons for ischemic injury of rectal mucosa include paradoxical contraction of the pelvic floor, external anal sphincter, and abdominal pressure. The youngest reported patient with SRUS was a 1.5-year-old child but the majority of cases are children older than 8 years.[8] The average time from the onset of symptoms to diagnosis is 5 years, ranging from 1.2 to 5.5 years.[8] The average time from the onset of symptoms to diagnosis is 3.2 years, ranging from 1.2 to 5 years in children, which is shorter than in adult patients (5 years; range, 3 months to 30 years).[9] It was 2 years in our patient. It has been reported that 75% to 80% of children with SRUS is male.[9] Our patient is a teenage girl.

Anemia is not typically present in SRUS.[10,11] A lengthy period of misdiagnosis may cause anemia due to prolonged bleeding, but blood transfusion in SRUS is rare.[12] Our case had anemia, received two unit of blood transfusion. The gold standard of diagnosis of SRUS is the histopathological findings. The histology of solitary rectal ulcer has a distinct feature including thickening of the mucosal layer with disrupting crypts structure, infiltration of the lamina propria with fibroblasts, muscle and collagen fibers that lead to hypertrophied and disrupted muscularis mucosa which look like fibromuscular obliteration.[8] Our patient's histology report revealed chronic inflammatotry cell infiltration in the lamina propria and surface erosion. The crypts were diamond shaped. The muscularis mucosa was thick and thin bands of smooth muscle fibers were present in between the crypts. This finding was consistant with SRUS. Macroscopically, SRUS typically appears as shallow ulcerating lesions on a hyperemic 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 but usually are 1 to 1.5 cm in diameter.[13] The polypoid variant is very rare among the cases reported in children.[14] Our case had shallow ulceration (4 cm in diameter) in rectal wall (5 cm from anal verge). Repeat endoscopies were not routinely carried out unless patients had persistent symptoms.[7] We did not perform colonoscopy during follow up as symptoms were resolved in our patient. Though SRUS is benign, the patient's morbidity is significant. As symptoms persist over a periods of time requires multiple admissions.[15] Our patient had multiple admissions in different institutions for this illness.[16-18]

At the time of diagnosis, patients should be counseled about taking high-fiber diet and bulk laxatives. They also need to be trained for avoidance of straining and anal digitation. The toilet habits (time spent in the toilet) should be adjusted. Dietary and behavioral changes, especially in patients with mild to moderate symptoms, can be dramatically effective in the absence of mucosal prolapse, which can help in the improvement and prevention of disease progression.[19]

The current treatment protocol is the use of enemas containing sucralfate, salicylate, corticoste-roid, sulphasalazine, mesalazine, and topical fibrin sealant.[3] Surgery is indicated in cases not responding to conservative treatment.[7] In the study by Dehghani et al[9] conservative treatments, behavioral and dietary changes were recommended as the initial treatment. In that study, 58.3% of the patients (7 out of 12 patients) had the complete recovery of symptoms after treatment with sucralfate enema and recommend this as a suitable treatment for children. One of their patients responded to Salicylate enema, 1 to corticosteroid enema, 2 to corticosteroid injection and 1 of the patients were finally treated with rectopexy.[9] Our patient responded well with bulk laxative and modified sucralfate enema. Though duration of treatment is not properly mentioned in any literature, we gave sucralfate enema for 14 days.

Conclusion

Solitary rectal ulcer syndrome (SRUS) is a rare benign and reversible condition in children. Few reported cases have undergone detailed investigations, treatments have been extremely varied and outcome poorly reported. The clinical features of SRUS is easily confused with other common diseases such as inflammatory bowel disease, infectious proctocolitis. So paediatric gastroenterologists and pathologists should have a high index of suspicion about SRUS and be alert about

the patient if he/she has gastrointestinal hemorrhages specially when other diagnosis is not matched. It is noteworthy that conservative management, patient education, fiber consumption, and bulk laxatives are the first strategies at initial stage. Surgical management is preserved for late stages.

References

1. Zhu QC, Shen RR, Qin HL, Wang Y. Solitary rectal ulcer syndrome: Clinical features, pathophysiology, diagnosis and treatment strategies. *World J Gastroenterol* 2014; **20**:738-44.
2. Chiang JM, Changchien CR,Chen JR. Solitary rectal ulcer syndrome: An endoscopic and histological presentation and literature review. *Int J Colorectal Dis* 2006;**21**:348-56.
3. Dehghani SM, Malekpour A, Haghighat M: Solitary rectal ulcer syndrome in children: Literature review. *World J Gastroenterol* 2012;**18**:6541-45.
4. Perito ER, Mileti E, Dalal DH, Cho SJ, Ferrell LD, McCracken M, Heyman MB: Solitary rectal ulcer syndrome in children and adolescents. *J Pediatr Gastroenterol Nutr* 2012;**54**:266-70.
5. Dehghani SM, Bahmanyar M, Geramizadeh B, Alizadeh A, Haghighat M. Solitary rectal ulcer syndrome: Is it really a rare condition in children? *World J Clin Pediatr* 2016;**5**:343-48
6. Suresh N, Ganesh R, Sathiyasekaran M. Solitary rectal ulcer syndrome: A case series. *Indian Pediatr* 2010;**47**:1059-61.
7. Urgancý N, Kalyoncu D, Eken KG. Solitary rectal ulcer syndrome in children: A report of six cases. *Gut Liver* 2013;**7**:752-55.
8. Abreu M, Alves RA, Pinto J, Campos M, Aroso S: Solitary rectal ulcer syndrome: A paediatric case report. *GE Port J Gastroenterol* 2017;**24**:142-46.
9. Dehghani SM, Haghighat M, Imanieh MH, Geramizadeh B. Solitary rectal ulcer syndrome in children: Prospective study of cases from southern Iran. *Eur J Gastroenterol Hepatol* 2008;**20**:93-95.
10. Martin de Carpi J, Vilar P, Varea V. Solitary rectal ulcer syndrome in childhood: A rare, benign, and misdiagnosed cause of rectal bleeding. Report of three cases. Dis Colon Rectum 2007; 50:534-39.
11. Pi RR, Mathai AM, Magar DG, Tantry BV. Solitary rectal ulcer syndrome in childhood. *Trop Gastroentaerol* 2008;**29**:177-178
12. Ertem D, Acar Y, Karaa EK, Pehlivanoglu E. A rare and often unrecognized cause of hematochezia and tenesmus in childhood: Solitary rectal ulcer syndrome. *Pediatrics* 2002;**110**:e79. Doi:10.1542/peds.110.6.e79.
13. Tjandra JJ, Fazio VW, Church JM, Lavery IC, Oakley JR, Milsom JW. Clinical conundrum of solitary rectal ulcer. *Dis Colon Rectum* 1992;**35**:227-34.
14. Saadah OI, Al-Hubayshi MS, Ghanem AT. Solitary rectal ulcer syndrome presenting as polypoid mass lesions in a young girl. *World J Gastrointest Oncol* 2010;**2**:332-34
15. Chong VH, Jalihal A. Solitary rectal ulcer syndrome: Characteristics, outcomes and predictive profiles for persistent bleeding per rectum. *Singapore Med J* 2006;**47**:1063-68.
16. Keshtgar AS. Solitary rectal ulcer syndrome in children. *Eur J Gastroenterol Hepatol* 2008;**20**:89-92.
17. Ertem D, Acar Y, Karaa EK, Pehlivanoglu E. A rare and often unrecognized cause of hematochezia and tenesmus in childhood: Solitaryrectal ulcer syndrome. *Pediatrics* 2002;**110**:e79.
18. Ruan M, Zhang Y, Feng Z, Qu B, Han M, et al. Solitary rectal ulcer syndrome in a young patient: A case report. *Iran Red Crescent Med J* 2017;**19**:e15106. doi:10.5812/ircmj.15106.
19. Sharara AI, Azar C, Amr SS, et al. Solitary rectal ulcer syndrome: Endoscopic spectrum and review of the literature. *Gastrointest Endosc* 2005;**62**:755-62.