Solitary rectal ulcer syndrome in children

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The solitary rectal ulcer syndrome (SRUS) is an unusual disorder in childhood. Although well recognized in adult literature, the pediatric experience with this condition is limited, so SRUS often goes unrecognized or misdiagnosed. There are very few pediatric case reports in the English literature.

This report describes four patients who presented with rectal bleeding, constipation, mucous discharge, and lower abdominal pain, with a diagnosis of SRUS. The diagnosis was made by rectoscopy, defecogram, anorectal manometry and histopathological evaluation. In two patients, defecogram showed a rectocele with both, the sphincter failed to relax to voluntary squeeze pressure on anorectal manometric examination. the histopathological finding in all patients was fibrous obliteration of the lamina propria with disorientation of muscle fibers. all of the patients responded well to conservative therapy, which included defecation training, laxatives, sulfasalazine, and application of rectal sucralfate enema, and remained asymptomatic on the follow-up.

Although rare in the pediatric population, SRUS should be relatively easy to recognize in the child with rectal bleeding, after elimination of other causes. If suspected, the diagnosis of SRUS may be made at endoscopy and confirmed by rectal biopsy.

Key words: solitary rectal ulcer syndrome, rectal bleeding, constipation, children.

Although, solitary rectal ulcer syndrome (SRUS) was described as early as the 19th century by Cruveilhier[1], the first definitive descriptions were made by Madigan and Morson in 1969[2]. The condition is rare, with an estimated annual incidence of one per 100,000 per year in the adult population[3]. The average age at presentation is around 30 years, but this condition can occur from adolescence to late middle age. It is even more unusual in childhood. The incidence of SRUS in children is unknown. We found only seven patients reported separately in the English literature. The clinical features of these cases are summarized in Table I[4-8]. Herein, we describe the clinical, radiological, and pathological features of four cases of SRUS occurring in children.

Case Reports

Case 1

An 11-year-old girl was referred because of intermittent rectal bleeding for four months. She had a history of straining during defecation, lower abdominal pain, and passage of mucus on defecation. Digital examination of the rectum revealed an enduration on the posterolateral rectal wall. test for intestinal parasites, including amebiasis, were negative. Sigmoidoscopy revealed an ulcer 2 to 6 cm in diameter on the posterolateral rectal wall 5 cm from the anal verge with well demarcated edges. Subsequent colonoscopic examination up to the cecum revealed no other lesions. An air contrast barium enema was normal, but evacuation of the barium was difficult. Anal manometry showed failure of he sphincter to relax to voluntary squeeze pressure (Fig. 1). Defecogram showed occult internal prolapse as well as a small posterior rectocele (Fig. 2). The first biopsy was reported as a non-specific granulation tissue of the rectum. The child was started on stool softener. A month later, a second sigmoidoscopy was performed because of the persistence of symptoms, and serial biopsies were repeated. These biopsies were reported as a solitary rectal ulcer (Fig. 3). After treatment with laxatives,

sulfasalazine, and defecation training for three months, the patient was asymptomatic, although the rectoscophic appearance remained unchanged. For the subsequent six months, she underwent reinvestigation of rectoscopy every month. During this period, although she was asymptomatic, a limited healing was observed by rectoscopy. Consequently, she was started on sucralfate enema in glycerine 10%, three times per day. After one month of local sucralfate application, the ulcer had shrunk significantly on endoscopic examination. After an additional three months of therapy with sucralfate enema, the ulcer had completely healed.

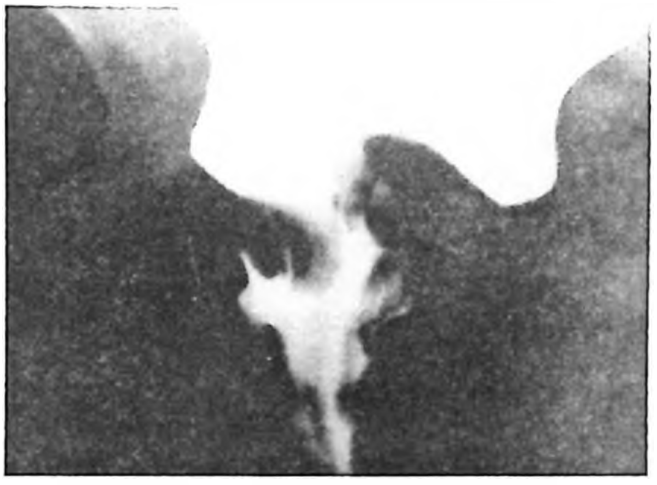

Fig. 1. Defecogram. Note the internal rectal intussusception and enterocele.

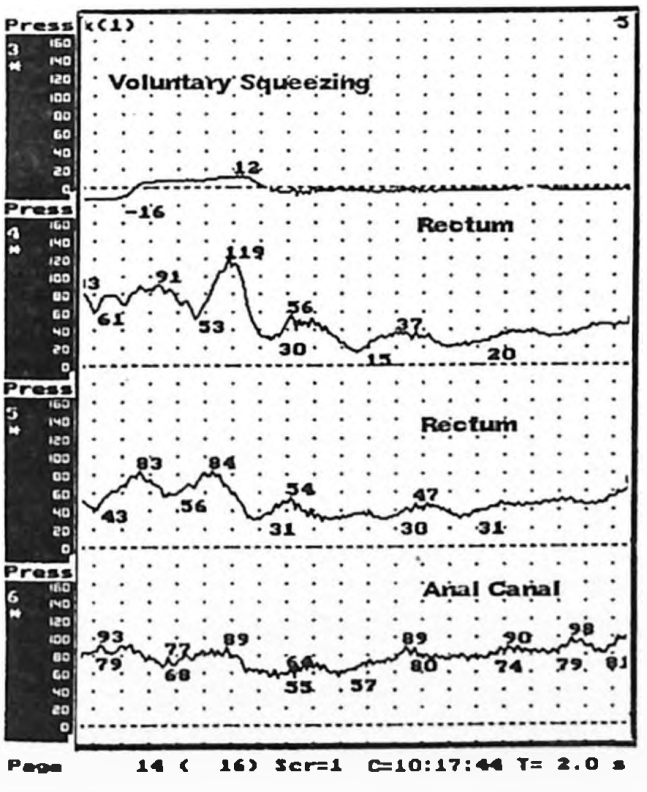

Fig. 2. Computerized anorectal manometry: no relaxation of pelvic musculature during squeezing.

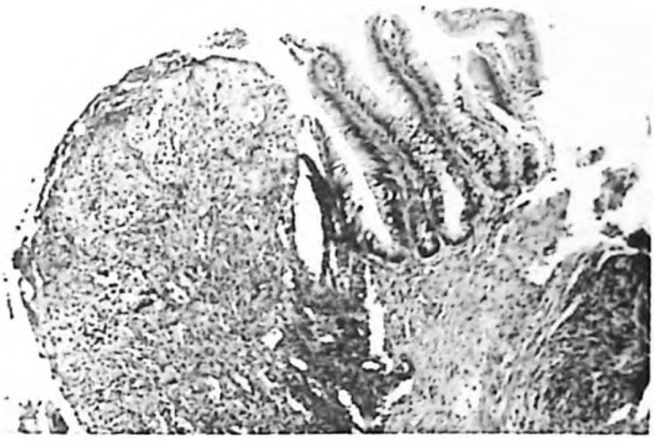

Fig. 3. Histological appearance of SRUS. The lamina propria is replaced by fibromuscular tissue. There are distorted crypts in the submucosa and superficial ulcerationn (H & E, x200).

Case 2

An eight-year-old girl presented with a small amount of rectal bleeding after defecation and lower abdominal pain. Previous complaint of the patient was chronic constipation for three months, which had been treated with defecation training and laxatives in another clinic. At the beginning of rectal bleeding, she was transferred to our institution. She was otherwise asymptomatic. Digital rectal examination was normal. Sigmoidoscopy showed an ulcer 0.5 to 1 cm on the posterolateral rectal wall 5 cm from the anal verge. Defecogram and anal manometric evaluation were normal. Histopathological examination revealed fibrosis of the lamina propria, ulceration and inflammatory cells infiltration. The ulcer healed three months after the initial episodes of bleeding, following treatment with laxatives and a high-fiber diet.

Case 3

An eight-year-old girl was referred because of constipation, rectal bleeding and rectal and lower abdominal pain during defecation. The physical examination was normal, except for sensation of the posterior rectal wall on rectal examination. During the rectoscopy, an ulcer 0.5 to 1 cm in diameter was seen on the right posterolateral rectal wall and a biopsy was taken. Defecogram showed an anterior rectocele 3.5 cm from the anorectal junction. The anorectal manometric examination showed that anal canal resting pressure was high and remained unchanged during squeezing. Pathology was

reported as a solitary rectal ulcer. Three months later after a conservative treatment, the ulcer was healed on endoscopic examination.

Case 4

A three-year-old-boy was referred because of a six-month history of constipation and soiling. Recently he had rectal bleeding and mucous discharge. The physical and rectal examinations were normal. Sigmoidoscopy showed an ulcer 0.5 to 1.5 cm in diameter surrounded by a hyperemic halo and covered with a fibrinous membrane on the posterior rectal wall 3.5 cm from the anal verge. Rectal biopsy was taken from the edge of the ulcer, and solitary rectal ulcer was confirmed by histopathological examination. The child was started on a high-fiber diet and laxatives. After one month of therapy, his symptoms disappeared and by the twelfth week the ulcer had almost healed.

Discussion

Solitary rectal ulcer syndrome is an unusual cause of rectal bleeding in children[9]. Only seven separate pediatric cases have been reported[1,4,5-8]. The largest series of solitary rectal ulcer patients, among those published by different authors, contain six extra pediatric cases (under 15-years-old)[2,10-12]. In these published series, none of the pediatric cases have been evaluated separately.

The etiology of the disorder remains obscure. Early report hypothesized that local proctitis or hamartomatous malformation in the rectal wall led to development of local ulcers, but no theory has been generaly accepted[6,13]. Recently, however, the common histological characteristic of the polypoid and ulcer lesions, as well as the characteristic history of straining and prolapse, have suggested that either local trauma or ischemia of the rectal wall is the common cause[10-15]. In normal circumstances, straining induces inhibition of the puborectalis muscle, but Rutter[15] showed increased electromyographic activity in patients with SRUS. Womack et al.[16] also reported increased intrarectal pressures during voiding. The significance of this finding is unknown, but it is stated that it has a traumatic effect on the rectal mucosa. Schweigher and Alexander-Williams[17] suggested that the SRUS is due to a preexisting rectal prolapse which itself may lead to further difficulty in defecation and also to secondary mucosal damage. The latter would result in increased mucus production and bleeding. In addition, patients with SRUS show failure of the sphincter to relax in response to voluntary squeeze pressure on manometric evaluation[4]. Most authors note two main causes of this mucosal lesion: rectal prolapse or intussusception and an abnormal relaxation of the pelvic muscles during defecation[4,16,17]. Two of our four patients had both rectocele and a high anal resting pressure without relaxation of the anal sphincter during squeezing. It is therefore suggested that SRUS is a weakening of the rectal wall caused by rectal prolapse or spastic pelvic floor syndrome, or both, in children. The most frequent symptoms are rectal bleeding, mucous discharge, tenesmus, and pain localized to the perineum or sacral area. It is common to find in these patients a long history of constipation, straining at defecation, and rectal prolapse, but these symptoms are non-specific and may be present with any other anorectal disease. In assessing the seven reported pediatric patients in whom sufficient clinical detail is available, only minor differences from adult patients are apparent[10,12,13]. In our patients, a small amount of rectal bleeding associated with constipation was the initial symptom.

Diagnosis of SRUS is made by rectoscopy, which also permits biopsy. The ulcers vary in size from a few milimeters up to 5 cm. The lesion is usually found on the anterior wall of the rectal mucosa. The posterolateral quadrants are rarely the site of ulcers. The distal point of the ulcers in children varies from 3 to 20 cm from the anal verge[4-8]. In our patients, all ulcers were seen on the posterior or posterolateral rectal wall and the distance of the ulcers from the anal verge varied from 3.5 to 11 cm. The significance of this localization is obscure. The varied macroscopic appearance of the rectal lesions that included mucosal hyperemia, ulceration, and/or polypoid lesion can be confusing. In addition, the rectal lesion can be multiple or circumferential[2,12]. Thus, an unusual-appearing rectal lesion with atypical presentation should arouse the suspicion of SRUS. The presence of rectal prolapse should further suggest a diagnosis of SRUS[4,7,13,16]. In our patients, the rest of the rectal mucosa was quite normal: there was no granular appearance or squamous metaplasia. The most common clinical diagnostic confusion is with inflammatory bowel disease or inflammatory cloacogenic polyp (ICP), which is

a rare polypoid lesion arising in the region of the anorectal transitional zone[14]. The most characteristic histopathological feature of SRUS is obliteration of the lamina propria of the mucous membrane in the region of the ulcer by fibroblasts and muscle fibers derived from the muscularis mucosae[2,12,14]. It is important to distinguish the histopathological appearances of SRUS. from inflammatory bowel disease, infectious proctocolitis and cloacogenic polyp. The major difference between ICP and SRUS relates to the site, but they share similar histopathological features. Furthermore, if a biopsy is taken from the proximal end of an ICP and does not include the squamous eithelium of the anal canal, it can be incorrectly reported as SRUS[14]. Although it is thought that these two clinical entities are manifestations of the same pathogenetic process, i.e., mucosal ischemia secondary to prolapse leading to the decribed histological changes[14,18], this theory has not yet been accepted. In three of our patients, the first rectal biopsy was histologically suggestive of solitary ulcer. In the other case, the first biopsy was reported as a non-specific granulation tissue, but the second biopsy showed classic histopathological findings of SRUS. All biopsies had ganglion cells and none had specific findings of neuronal intestinal dysplasia.

The most appropriate treatment of this conditionhas not yet been determined. Treatment of SRUS is difficult and should be conservative as far as possible. Conservative treatment of SRUS includes a high residue diet, stool softeners, and defecation training[5,6,8,10,11,13]. Most studies have suggested that sulfasalazine and topical steroids are of no significant benefit to these patients[10,12,13]. Sucralfate has been shown to be effective in the treatment of peptic ulcer, esophagitis, and post-polypectomy hemorrhage. Batman et al.[19], and Spiliadis et al.[20] showed that sucralfate enema might be effective in the teratment of SRUS. Symptomatic improvement and healing after medical therapy such as a high residue diet, laxatives, and defecation training were obtained in three of our four patients. Although the other patient was totally asymptomatic after three months of conservative therapy, endoscopic healing of the ulcer was completed after one year of treatment. Sucralfate enema was successfully used in this patient.

Although local excision, insertion of a Thiersch wire, a different kind of abdominal rectopexy and colostomy have been recommended as operative treatment in adults[12,13,17,21], none of these procedures is free of complication. A laser therapy has recently been recommended by some authors[22]. In the pediatric age group, conservative therapy should be considered as a first choice of treatment. We successfully treated our patients using conservative treatment.

The recognition of solitary rectal ulcer is relatively easy in adults because of the lower localization of the lesion and different macroscopic appearance of the SRUS, but diagnosis is somewhat difficult in children. Since the localization of the lesion is important in children, careful endoscopic examination is necessary. If the ulcer does not respond to classical medical therapy, the sucralfate enema may also be used.

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REFERENCES

1. Cruveilhier J. Ulcere chronique du rectum. Anat Path 1870; 2: 4.
2. Madigan MR, Morson BC. Solitary ulcer of rectum. Gut 1969; 10: 871-881.
3. Martin CJ, Parks TG, Biggart JD. Solitary rectal ulcer syndrome in Northern Ireland 1971-1980. Br J Surg 1981; 68: 744-747.
4. Eigenmann PA, Coultre CL, Cox J. Solitary rectal ulcer: an usual cause of rectal bleeding in children. Eur J Pediatr 1989; 152: 658-660.
5. Figueroa-Colon R, Younozasi MK, Mitrops FA. Solitary rectal ulcer syndrome of rectum in children.J Pediatr Gastroenterol Nutr 1989; 8: 408-412.
6. Gündoğdu ZH, Altin MA, Aksoy F. Solitary rectal ulcer in a 12-year old girl: an unusual cause of rectal bleeding. Pediatr Surg Int 1994; 9: 131-132.
7. Rubia LD, Villaespesa AR, Cebrero MC. Solitary rectal ulcer syndrome in a child. J Pediatr 1993; 122: 733-735.
8. Sondheimerr JM, Slagle TA, Bryke CR. Solitary rectal ulcer syndrome in a teenaged boy. J Pediatr Gastroenterol Nutr 1985; 4: 835-838.
9. Balkan E, Kırıştıoğlu İ, Gürpınar A, Özel İ, Sınmaz K, Doğruyol H. Sigmoidoscopy in minor lower gastrointestinal bleeding. Arch Dis Child 1998; 78: 267-268.
10. Brant-Gradel VV, Huibregtse K, Tytggat GN. Treatment of solitary rectal ulcer syndrome with high-fiber diet and abstention of straining at defecation. Dig Dis Science 1984; 29: 1005-1008.
11. Ford MJ, Anderson JR, Gilmour HM. Clinical spectrum of solitary ulcer of the rectum. Gastroenterol 1983; 84: 1533-1540.

12. Tjandra JJ, Fazio VW, Petras RE. Cinical and pathologic factors associated with delayed diagnosis in solitary rectal ulcer syndrome. Dis Colon Rec 1993; 36: 146-153.
13. Niv L, Bat L. Solitary rectal ulcer syndrome: clinical, endoscopic, and histologic spectrum. Am J Gastroenterol 1986; 81: 486-491.
14. Bass J, Souchy P, Walton M. Inflammatory cloacogenic polyps in children. J Pediatr Surg 1995; 30: 585-588.
15. Rutter KR. Solitary rectal ulcer syndrome. Proc R Soc Med 1975; 68: 22-26.
16. Womack NR, Williams NS, Holmfield JH. Pressure and prolapse-the cause of solitary rectal ulceration. Gut 1987; 28: 1228-1233.
17. Schweigher M, Alexander-Williams J. Solitary ulcer syndrome of rectum its association with occult rectal prolapse. Lancet 1977; 1: 170-171.
18. Washington K, Rourk MH Jr. Inflammatory cloacogenic polyp in a child: part of the spectrum of solitay rectal ulcer syndrome. Pediatr Pathol 1993; 13: 409-414.
19. Batman F, Arslan S, Telatar H. Effect of sucralfate enema in the treatment of solitary rectal ulcer. Endoscopy 1988; 20: 128.
20. Spiliadis C, Skandalakis N, Emmanouliadis A. Treatment of solitary rectal ulcer syndrome with sucralfate enema. Gastrointest Endosc 1989; 35: 131-132.
21. Tjanda JJ, Fazio VW, Church JM. Ripstein procedure is an effective treatment for rectal prolapse without constipation. Dis Colon Rec 1993, 36: 501-507.
22. Rau BK, Harikrishnan KM, Krishna S. Laser therapy of solitary rectal ulcers: a new concept. Ann Acad Med Singapore 1994; 23: 27-28.