Original Article

Solitary rectal ulcer syndrome

Clinicopathological Review of a Series of 19 Patients

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Abstract

Background and Objectives: **The etiology, pathophysiology, nomenclature and clinical manifes-tation of solitary rectal ulcer syndrome (SRUS) is poorly understood.**

Aim: To examine the pathology of mucosal changes and clinical features of this syndrome.

Materials and Methods: **The mucosal biopsies of 19 patients with clinical evidence of solitary rectal ulcer has been reviewed in accordance with their clinical complaints and endoscopic findings within three years..**

Results: **SRUS has been seen in both sexes (11 male and 8 female) at the age ranging from 12 to 72 years (mean 29 years). The main clinical complaints were rectal bleeding, mucorrhea and perianal pain. The major rectoscopic findings were ulceration, erythema of mucosal surface, congestion and polypoid pattern. The significant pathologic features were mucosal architectural distortion, very superficial and irregular mucosal ulceration, fibrosis of lamina propria and thickening of the muscularis mucosa with splaying of its fibers and extension of muscularis fibers between the mucosal crypts.**

Conclusion: **Although SRUS is rare but it can be confused clinically with other similar diseases such as inflammatory bowel disease,cloagenic carcinoma and other malignancies. It must be looked in differential diagnoses of pelvic disorders since it is well recognized on rectoscopic and morphologic basis.**

Key words: **Rectom, solitary, ulcer, solitary rectal ulcer syndrome (SRUS)**

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Introduction

Solitary rectal ulcer syndrome (SRUS) is an infrequent disorder often associated with pelvic floor discomfort or rectal mucosal prolapse (1) The nomenclature of solitary rectal ulcer is controversial, since there are cases with no ulceration, patients with several similar ulcers . In addition the lesion can be seen in other parts of the colon and even has been reported in other site of gastrointestinal tract (1-8).Now, however, the entity of SRUS is popular and accepted by authors in medical literatures(6,7)

The clinical symptoms usually manifested at third or fourth decades (1,3,8) but it has been reported in children frequently (9-11) The main symptoms appear as rectal bleeding, mucus discharge, changes in defecation habits and perianal pain. Rectoscopic examination show irregular ulceration in the anterior wall or antero-lateral wall of rectum. Some patients do not show ulcer ,besides having rough and hyperemic polypoid or prolapsed mucosa. The lesion is approximately 4-18 centimeters far from anal margin(1). The most characteristic histopathology features are mucosal fibrosis,hypertrophy of muscularis mucosa with extension of their fibers between the mucosal glands (5,8).

In this report we will discuss the clinical presentations, rectoscopic features and morphologic findings of 19 cases presented in our laboratory. We will briefly discuss the other name of this syndrome including rectal mucosal prolapse which sometimes it is considered as synonym or preferred name for this lesion(8).

Patients and Methods

19 patients were clinically suspected for SRUS have been studied in this series. The have been clinically and rectoscopically evaluated by two surgeons (MV and SD) and proper mucosal biopsies were taken. The sample immediately sent to pathology laboratory in 10% formalin liquid fixative. For each sample several serial thin sections (between 6-12 sections) from paraffin embedded block made. The sections were stained with routine Hematoxylin Eosin(H&E) staining. When necessary ,in addition to H&E staining with Masson Trichrome and Bielschowsky Reticulin for collagen fibers and configuration of the epithelium performed. The slides were reviewed with two pathologists separately (MB and KGM) and the results, after consensus, were recorded.

Results

Between January 2000 to December 2002 ,19 patients with clinical diagnosis of SRUS have been evaluated. Table 1-4 showing the age and sex distribution, clinical presentations ,rectoscopic features and histological findings respectively. Two boys aged 12 years and six patients over 30 years are among our patients. The majority of our cases are young and less than 30 years old. The oldest one was a woman aged 72 year. The mean age was 29

years which the average was 24 years among the men and 37 years among the women. There were eight women and 11 men ,this is contrary to the believe of women predominance.

Table 1. Age and Sex Distribution

Age group(year)	Male	Female	Total	٪
Under 14	2	0	2	10.5
15-30	7	4	11	57.9
31-up	2	4	6	31.6
total	11	8	19	100
percent	57.9%	42.1%	100%	-

Table 2 indicates the clinical manifestations. The most frequent complaints was rectal bleeding, mucus discharge and perianal pain. Other complaints according to frequency were longstanding on toilet, tenesmus, finger extraction and difficulty in defecation. It is plausible that each patient may have more than one problems.

Table 2. Clinical Manifestation

Symptom	number	percent
Rectal bleeding	18	94.7
Mucous Discharge	11	57.8
Perianal pain	8	42.1
Longstanding on toilet	7	36.8
Finger extraction	6	31.5
Frequency	5	26.3
Tenesmus	4	21.o
Diarrhea	4	21.0
Diarrhea/constipation	1	5.2

Rectoscopic examination revealed rectal mucosal ulcer either on the anterior mucosal surface or anterolateral. there were more than one ulcer in some cases. Other findings were erythema or redness of mucosa, sign of inflammation, hemorrhoids and polypoid or prolapsed pattern (table 3). The site of the lesion has been divided as less than six centimeter above anal verge ,6-14 centimeter above anal verge and in four patients either the lesion was diffuse or not known (table3).

Table 3. Rectoscopic findings

Features	No.	٪
Ulceration*	18	94.7
Site :less than 6cm.		
Above anal verge	4	21.0
More than 6cm above		
Anal verge	11	57.8
Not known or diffuse	4	21.0
Mucosal prolapse	3	15.7
Inflammatory polyp	2	10.5
Hemorrhoids	2	10.5
Miscellaneous	4	21.0

* three cases show more than one ulcer

Table 4 presents the histologic findings. Fibrosis of lamina propria was the major histologic features which have been seen in all samples (100%). This was accompanied with fibromuscular obliteration of intercryptic lamina. Thickening and hypertrophy of muscularis mucosa with splaying of its fibers and extension of fibers between the glands have been seen in 18 cases (94.7%). The third in frequency findings were mucosal architectural distortion often with hyperplastic crypts and/or villiform pattern, there was superficial ulceration, ectatic venules hyperemia and congestion. The inflammation was absent or very mild. No cryptic abscess or goblet cell depletion seen.

Table 4 . Histopathologic findings

Morphology	No.	٪
Fibrosis of lamina propria	19	100.0
Hypertrophy/thickening of		
Muscularis mucosa		
With splaying fibers	18	94.7
Intercryptic Fibromuscular		
Obliteration	17	89.4
Mucosal architecture distortion	17	89.4
Superficial ulceration,congestion		
And/or ecstatic venules	12	63.1

In two cases more than one biopsy taken

There were two patients which had more than one biopsy (two times in cases 13 and 17, the first attempt was unsatisfactory and granulation tissue respectively). For practice, we divided the mucosal morphologic changes as major and minor. The major criteria were: fibrosis of lamina propria and/or intercryptic obstruction; muscularis mucosa hypertrophy wit extension of its fibers between the glands; and, mucosal architectural distortion. The minor criteria were: superficial mucosal ulceration; ectatic venules; and, hyperplastic crypts and/or villiform pattern. At least two of the three major and on minor criteria must be present for histologic satisfaction.

Discussion

Solitary rectal ulcer syndrome (SRUS) is a rare disorder manifested by disturbed defecatory behavior, passage of blood and mucus and pelvic pain. On rectoscopic examination the lesion is

characterized as erythematous mucosa and/or with ulceration of the rectal wall, associated with typical histological features (1-6). Morson describe the lesion as: solitary ulcer/mucosal prolapse syndrome (5) Because of polypod features and rectal prolapse the term Prolapsing Mucosal Polyp has been given(12). Other terminology such as Rectal Wall Prolapse (complete or incomplete) (13,14,15), Rectal Internal Mucosal Prolapse (RIMP) (16), Colitis Cystica Profunda (17), Inflammatory Cloacogenic Polyp (8,12), Inflammatory "Cap" polyp (12) and Benign Idiopathic Recurrent Rectal Ulceration (BIRRU) (1) has been given to this disorder. Each of these have their own rectoscopic or histologic pattern which can be linked one way or another to this syndrome. Among these the most frequent term is either solitary rectal ulcer or rectal mucosal prolapse. Since the disorder is seen in other site of colorectal mucosa and occurs in many forms throughout the gastrointestinal tract, and ulcer is not always seen, neither the ulceration is always single but in the majority of cases rectal ulcer is present, hence the term solitary rectal ulcer syndrome has been preferred by the authors (see references).

SRUS has been reported in both sexes and in all ages but they are most frequent in adulthood in the third or fourth decades(1,3,4). SRUS is rare among children and few reported cases are seen in the literatures (8-11)We have seen two cases among our series of 19 patients. Other interesting matter in our series is that our patients are younger from those cases in literature(11,15,16). Approximately 70% of our patient were in the second or third decades. The reason for this younger occurance must be investigated. Often SRUS goes unrecognized or misdiagnosed, this may happen more frequently in children and younger age groups (8,9).In our series the male are more than female(11/8) which is different from other series (14,15).

The clinical presentation varies. They are seen most frequently as an ulcer in the anterior or anterolateral mucosa of rectum. The ulcer is irregular in shape and size but well demarcated 4-18 centimeters far from anal verge. The majority of our cases had only one ulcer but multiple ulcer has been reported (11) Associated internal rectal prolapse,mucosal prolapse, and total rectal prolapse is seen in some patients (6,14,15,8,). Kang et al divided the

SRUS into three groups based on the extent of accompanying rectal prolapse (no prolapse, internal prolapse and external prolapse (18). Many authors consider rectal prolapse and SRUS as one entity, nevertheless there are evidences that these two conditions are different both histologically and clinically (14,15,17) and shall have overlapping.. Kang et al suggested that solitary ulcer patients have a spectrum of clinical and physiological features which may be with or without prolapse. They also suggest a different underlying etiopathophysiology of SRUS from that of complete rectal prolapse. Mackle et al concluded that both conditions are being affecting the rectum but with different pattern. Prolapse tends to occur late in life, which SRUS has a predilection for younger age (15). This syndrome have varieties of clinical ,etiological and physiological features and compose a range of different disease entities (15,9). Typical clinical history, rectoscopy and histology distinguish SRUS from other rectal ulcer-associated etiology (10). Identical mucosal changes are seen in the apex of a complete rectal prolase, the apex of a prolapsing hemorrhoids and the tip of colostomy(5)

The etiology and pathophysiology of SRUS are poorly understood (14). The pathogenesis is likely to vary in different patients. SRUS is essentially due to strain and traumatization of the rectal mucosa.Inappropiate puborectalis contraction, abnormal perianal descent and rectal prolapse have all been cited the possible mechanism of developing SRUS (15,19,20) This syndrome is a consequence of chronic straining during defecation, ischemic changes due to vascular damage, direct digital trauma and

possibly primary neuromuscular pathology (7,11) On defecography failure of pelvic floor relaxation during straining and non-relaxing puborectalis muscle on straining have been reported (11,15) Electromyography showed evidence of pudental nerve damage but incontination is rare or absent in this syndrome(15). In chronic cases changes are analogous to colitis cystica profunda (1,5)

The histolopathology although is characteristic but shows different features at different stage. In early cases sometimes a preulcer polypoid phase can be seen (3). With regard to histologic diagnoses of SRUS,the pathologists should be familiar with this morphology and well trained (21) The biopsy obtained from ulcerated areas usually is nonspecific consisting of fibrinopurulent exudates and granulation tissue. Hence, the rectoscopic biopsy should be obtained from mucosa near the ulcer. The sample may come from operative full thickness rectal wall, or endoscopic rectal mucosal biopsies. All cases in our series came from rectal mucosal biopsies. The pathology in these cases were straightforward and consistent with the findings of other authors. Hyperplasia of crypts ,some of which having a peculiar diamond shape and tending towards villous configuration may lead to misdiagnosis of adenoma (5) In the case of rectal wall surgical specimen in addition to mucosal findings there are thickening of rectal wall by hypertrophy and disorganization of muscular propria, particularly in inner circular muscular layer, nodular induration of inner circular layer and grouping of outer longitudinal layer into bundles, this changes are characteristic of SRUS (14). These changes are not seen in complete rectal prolapse or control specimen. Of histological importance is the misplacement of the glands may be associated with dissecting mucous pool and can be easily mistaken for invasive mucinous adenocarcinoma (1,3)

Though the majority of cases, being familiar with the syndrome,

come to attention at clinical presentations, but there are cases which were misdiagnosed at initial evaluations. These include inflammatory bowel disease, rectal polyps, villous tumor, jevenile polyposis, nonspecific ulceration, proctitis and malignancies. This mis-interpretation is seen most in children cases (2,8,18,19). There are report indicating malignant consequence of the syndrome such as invasive and insitu carcinoma (22,23), but there is no sufficient proof for their relationship.

The treatment of SRUS remains problematic and is less than ideal. Well designed,prospective studies on the efficacy of various procedures for the treatment of SRUS are unsatisfactory and experiential (24,17). Surgical therapy and non-surgical therapy have been proposed, non of these proved to be curative, but, conservative and palliative. Since complete " cure " is uncommon in this syndrome, the goal of therapy should be decrease of clinical complaints. The patient should be aware of his/her illness, patient must know that the disease which is a benign lesion but may recur and the treatment is, according to the stage of disease, for the cessation or minimizing the clinical symptoms (24). Conservative therapy with dietary fiber, bowel retraining and trial of some medications. With this awareness of patients surgical procedure if needed must be performed.Several surgical procedures have been proposed .According to the stage and accompany complication these procedure are performed (17).

Conclusion

SRUS is an uncommon disease that usually present in younger age groups. Clinical presentations are rectal bleeding, mucorrhea, perianal pain and pelvic discomfort. Endoscopic examination and histologic findings are helpful in the diagnoses and management of the syndrome. The clinical management and the goal of therapy needs the cooperation of patients. A stepwise, individualized approach must be employed. These include defecatory training, conservative therapy, high fiber diet with bulk of laxatives, non-surgical medication and surgery.

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