Research Article

The protean clinical endoscopic and histological manifestations of solitary rectal ulcer syndrome

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Abstract

Background: Solitary Rectal Ulcer Syndrome (SRUS) is a relatively uncommon, benign condition which clinically, endoscopically and histologically can mimic many conditions including carcinoma of the rectum and inflammatory bowel disease.

Objectives: Description of the clinical manifestations, endoscopic findings and histological features in a cohort of patients with SRUS.

Method: The clinical histories, endoscopic records and the Haematoxyline & Eosin stained slides of 43 patients diagnosed as having SRUS were analyzed.

Results: The total study population included 43 patients (men-25 and women-18) with a median age of 45 years and age range from 9-80 years. Symptoms were documented in 33 patients and the commonest presentation was altered bowel habits which included tenesmus, mucous discharge, excessive straining and sensation of incomplete defecation seen in 58% (n=19). Other symptoms were rectal bleeding, diarrhoea and constipation in 48% (n=16), 15%(n=5) and 9%(n=3) respectively.

Key words: solitary rectal ulcer, endoscopy, histopathology

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The endoscopic findings were available in 38 patients and revealed a solitary rectal ulcer, polypoidal lesion, erythematous unhealthy mucosa, circumferential ulcers and an indurated area in 37%(n=14), 24% (n=9), 16% (n=6), 13%(n=5) and 11% (n=4) respectively. The predominant histological features seen were crypt distortion, fibromuscular obliteration of the lamina propria and telangiectatic vessels in 100% (n=43), 98% (n=42) and 70% (n=30) respectively. Other histological features included mild mucin depletion, mucosal ulceration, inflammation in the lamina propria, surface serration and cryptitis, seen in 49% (n=21), 40% (N=17), 37% (n=16), 16%(n=7) and 2%(n=1) respectively.

Conclusion

Presence of a wide range of clinical, endoscopic and histological features in SRUS is also seen in this study similar to other studies but with difference in frequencies. Awareness of these features would enable histopathologists to avoid possible pitfalls in the diagnosis.

Introduction

Solitary rectal ulcer syndrome is a known mimicker of many conditions including adenocarcinoma, inflammatory bowel disease, dysplasia, and adenomatous polyp. The name solitary rectal ulcer is also misleading and it is commonly known as the “three-lies disease” since it is not always ulcerated, not always solitary, and not always located in the rectum [1].

The pathogenesis of SRUS is not fully understood. The most widely accepted theory is that it is related to direct trauma or local

ischemia [2]. The most common presentation in SRUS is bleeding per rectum along with discharge of mucus. Another common complaint is anorectal or abdominal pain. Some patients have constipation and a sense of incomplete evacuation [1]. Apart from diverse clinical presentations, the endoscopic findings are also varied and include ulceration, a polypoid appearance, or simply mild erythema [2].

The histological examination is the key to the diagnosis of SRUS. The microscopic features include surface serration, fibromuscular obliteration of the lamina propria, crypt distortion and vessel changes such as ectasia, congestion, and hyalinization [2]. However, these features are not pathognomonic of SRUS.

SRUS is one of the four entities that are grouped together as mucosal prolapse syndrome that includes, a spectrum of benign disorders related to defaecation comprising SRUS, rectal prolapse, proctitis cystica profunda (PCP) and inflammatory cloacogenic polyp. These four entities share similar clinical and pathological features (3).

Since there is a wide spectrum of clinical, endoscopic and histological features, this study was conducted to further characterize the clinical and pathological features of this syndrome in a Sri Lankan population, with the objective of increasing the awareness amongst both clinicians and histopathologists.

Objective

Description of the clinical manifestations, endoscopic findings and histological features in a cohort of patients with SRUS.

Method

Forty-three histologically diagnosed SRUS cases were retrieved from the departmental archives and reviewed retrospectively. The request forms of these cases were then analyzed to find the presenting clinical symptoms and endoscopic findings. Symptoms included altered bowel habits, rectal bleeding, diarrhoea and constipation. The clinical symptoms of altered bowel habits

included tenesmus, mucous discharge, excessive straining and a sensation of incomplete defecation. The endoscopic features were categorized as ulcerative, polypoid/nodular, erythematous inflamed mucosa or an indurated area. The ulcers were again categorized as solitary or circumferential. The histological features noted included crypt distortion, fibrosis in the lamina propria, smooth muscle splaying of muscularis mucosa, telangiectatic vessels, mucin depletion, mucosal ulceration, inflammation in the lamina propria, surface serration and cryptitis. Inflammation in the lamina propria was diagnosed based on the presence of neutrophils and a lymphoplasmacytic infiltrate filling the lamina propria to the base of the crypts. Cryptitis was defined as neutrophils seen within the crypt epithelium.

Results

A total of 43 cases were evaluated for clinical, endoscopic and histological features. Mean age of the group was 45 years (range 9–80 years); There were 25 (58%) men and 18 (42%) women. Symptoms were documented in 33 patients. The most common symptom was altered bowel habits affecting 19 (58%) patients followed by rectal bleeding 16 (48%), diarrhoea 5(15%) and constipation 3(9%) [Table 1].

The endoscopic findings were available in 38 cases and included a solitary rectal ulcer (**Figure-1**), polypoid lesion, erythematous unhealthy mucosa, circumferential ulcers and indurated mucosa in 37% (n=14), 24% (n=9), 16% (n=6), 13% (n=5) and 11% (n=4) respectively. [Table 2].

Table-1: Clinical symptoms in patients with SRUS

Symptoms (N = 33)	N (%)
Altered bowel habits	19 (58%)
Rectal bleeding	16 (48%)
Diarrhoea	5 (15%)
Constipation	3 (9%)

The predominant histological features seen were crypt distortion, fibrosis in the lamina propria with splaying of smooth muscles in muscularis mucosa (**Figure-2**), and telangiectic vessels (**Figure-4**) in 100% (n=43), 98% (n=42) and 70% (n=30) respectively. Other histological features included mild mucin depletion, mucosal ulceration, inflammation in the lamina propria, surface serration (**Figure-3**) and cryptitis seen in 49% (n=21), 40% (N=17), 37% (n=16), 16% (n=7) and 2% (n=1) respectively.

Figure 1: Solitary ulceration with contact bleeding in the surrounding mucosa.

Table -2: Endoscopic findings in patients with SRUS

Endoscopic finding (N= 38)	N (%)
Solitary rectal ulcer	14 (37%)
Polypoidal lesion	9 (24%)
Erythematous unhealthy mucosa	6 (16%)
Circumferential ulcers	5 (13%)
Indurated area	4 (11%)

Discussion

Solitary rectal ulcer syndrome (SRUS) was first described by Cruveilhier, Madigan and Morson in 1969. The pathogenesis of this syndrome was postulated to be due to rectal prolapse caused by excessive straining, during which high intra-abdominal pressure forces the anterior rectal mucosa to be compressed on to

the contracting puborectalis muscle. As a result, the anterior rectal mucosa is frequently forced into the anal canal, becoming strangulated in turn causing congestion, edema and ulceration. These changes are evident histologically, as fibromuscular obliteration of the lamina propria with disorientation of muscle fibers resulting from the chronic mechanical insult, ischemic trauma and inflammation caused by hard stools leading to intussusception of the rectal mucosa [4]. SRUS usually affects young adults and the mean age at presentation is the third and fourth decade of life [5]. However, its variable presentation and chronicity makes it a diagnostic as well as a management challenge.

In our study, majority of patients were men with a male-to-female ratio of 1.4:1.0. The mean age was 45 years and ages ranged from 9 to 80 years.

The mean age at presentation is slightly older than reported in other studies [3,5]. In most studies there was no sex predilection for SRUS. However, in a retrospective study done by Tjandra et al which consisted of 80 patients with biopsy-proven solitary rectal ulcer (SRU) with a median follow-up of 25 months, the female-to-male ratio was 1.4:1.0, and the mean age was 48.7 years (range, 14-76 years) [6].

The commonest clinical presentation in this study was altered bowel habits which was seen in 58% of the study group. Rectal bleeding, the second commonest presentation was seen in 48% of patients. These findings were different from other previously published data which showed that the commonest presentation was the bleeding per rectum in 60% of patients [1,5].

All our patients were symptomatic and a minority had diarrhoea (15%) and constipation (9%). However, in one study, constipation was the main clinical feature seen in 55% of patients and diarrhoea in 20%. 25% were asymptomatic [1]. This wide variation in symptoms has made this clinical entity very difficult to diagnose solely on the basis of symptoms which might be the main reason for

under-diagnoses and mis-diagnoses, reported in literature.

Figure-2: Smooth muscle splaying, (H&E X100)

Figure-3: Surface serration and angulated glands (H&E x100)

Figure-4: Ectatic blood vessels (H&E X100)

Table 3: Histological findings in patients with SRUS

Histological features (N=43)	N (%)
Crypt distortion	43 (100%)
Fibrosis and smooth muscle splaying	42 (98%)
Telangiectic vessels	30 (70%)
Mucodepletion (mild)	21 (49%)
Mucosal ulceration	17 (40%)
Lamina propria inflammation	16 (37%)
Surface serration	7 (16%)
Cryptitis	1 (2%)

The endoscopic appearance of SRUS is also variable ranging from simple hyperemic mucosa to ulcerations and polypoid lesions of variable size and number [1,7]. The endoscopic features in this study, showed a higher rate of solitary rectal ulcers amounting to 37% compared to 20% in another study done in China by Zhu et al [7]. Other endoscopic findings observed included, polypoid lesions, erythematous and unhealthy mucosa, circumferential ulcers and an indurated mucosa, in 24%, 16%, 13% and 11% of the sample respectively.

Due to versatile clinical and diverse endoscopic characteristics, SRUS can be under recognized or misdiagnosed; Thus, it is crucial to have histopathological confirmation for a definite diagnosis of this entity.

SRUS has well-described histopathological features such as obliteration of the lamina propria by fibrosis and smooth muscle fibers extending from a thickened muscularis mucosa. The histological features of diffuse collagen deposition in the lamina propria and abnormal smooth muscle fiber extensions are sensitive markers for differentiating SRUS from other conditions [7]. The other significant histological feature is the presence of crypt distortion which takes the form of angulated or “squared off” crypts, a feature which is useful in the differentiation from inflammatory bowel disease where crypt branching, shortening and variability in crypt size leads crypt distortion.

In this study, we found that the predominant histological feature was the crypt distortion due to angulated or “squared off” crypts seen in all the cases included in the study (100%). Nevertheless, smooth muscle splaying was also observed in nearly all cases (98%) except in one which showed extensive hyalinization / collagenization of the lamina propria. Similarly, Al-Brahim and colleagues demonstrated that all 13 of their patients had crypt distortion and surface serration [1]. In slight contrast, Abbasi et al reported fibromuscular obliteration in all subjects while crypt distortion was observed in a lesser proportion [5]. Therefore, fibromuscular obliteration and crypt distortion can be regarded as consistent findings in SRUS.

The unavailability of specialized investigations such as ano-rectal physiological testing and defaecography at our institution was a major drawback. Hence, the presence or absence of prolapse and the degree of prolapse was not assessed. Another limitation was the unavailability of information regarding management strategies and follow-up details of the disease progression.

Conclusion

This study highlights that SRUS is a disorder of the middle age group with a male predominance and diverse clinical and endoscopic presentation. Histological features, such as fibromuscular obliteration of the lamina propria and the typical crypt distortion pattern are consistent histological findings and sensitive markers that differentiates it from other mimickers. Misdiagnosis of SRUS has been reported in one fourth of adult cases and as a result the correct diagnosis has been usually delayed approximately 5 to 7 years [4]. A high index of suspicion is needed to differentiate SRUS from other benign conditions as well as other potentially devastating disorders such as carcinoma.

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