

Solitary Rectal Ulcer Syndrome - The Masquerader

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Abstract- Introduction: Solitary rectal ulcer syndrome (SRUS) is a great masquerader and misdiagnosis has serious implications for the patient. **Methods and materials:** We reviewed 21 patients with histopathologically proven SRUS at our tertiary care teaching hospital in South India. **Results:** Bleeding per rectum and constipation with passage of mucous were the common symptoms at presentation. Endoscopy revealed a single ulcer in 16 (76.19%), multiple ulcers in 5 (23.8%), polypoidal lesion in 1 (4.7%) and circumferential ulcer in 2 (9.5%). 3 (14.28%) patients had rectal ulcer resembling malignancy on endoscopy such as everted edge 1 (4.7%) and hard growth 2 (9.5%). Endoscopic biopsy from one such patient was reported as adenocarcinoma for which abdominoperineal resection was done. The final histopathological report of the resected specimen was SRUS. **Conclusion:** SRUS is a misnomer with a potential for misdiagnosis as carcinoma. Under reporting may result in progression of disease and over reporting is disastrous for low rectal lesions.

Index Terms- Bleeding per rectum, Carcinoma, Constipation, Solitary rectal ulcer syndrome.

I. INTRODUCTION

T Solitary rectal ulcer syndrome is a benign condition with chronic ischemia as the most plausible of the several speculated aetiologies^[1].

- The clinical presentation is neither distinctive nor specific to the condition. The symptoms are varied and include rectal bleeding, constipation, straining at stools, diarrhoea and abdominal pain. The endoscopic features range from a spectrum of solitary ulcer to polypoidal lesions. 10% have more than one lesion^[2]. The distinctive but not pathognomonic histologic feature of SRUS is colitis cystica profunda i.e. mucous filled glands misplaced into the sub mucosa or muscularis mucosae and lined with normal colonic epithelium (Fig. 1).
- SRUS is a great masquerader of other serious conditions including adenocarcinoma, inflammatory bowel disease, dysplasia and adenomatous polyp. To error in diagnosis has the potential for treatment disasters, especially if surgical options are considered for low rectal lesions.
- Conservative treatment includes bulking agents, sphincter bio feedback training^[3] and topical mesalamine. Coagulating the lesion using Argon plasma^[4] has also been tried.

Surgical procedures are reserved for non responders with transanal Delorme mucosal resection^[1], stapled transanal rectal resection (STARR)^[5] and abdominal prolapse repair being the most frequently performed. Despite treatment, up to 30 % of patients with SRUS complain of persistent disturbance to anorectal function and significant compromise in their quality of life.

- This study presents the diagnostic and therapeutic dilemma in management of SRUS.

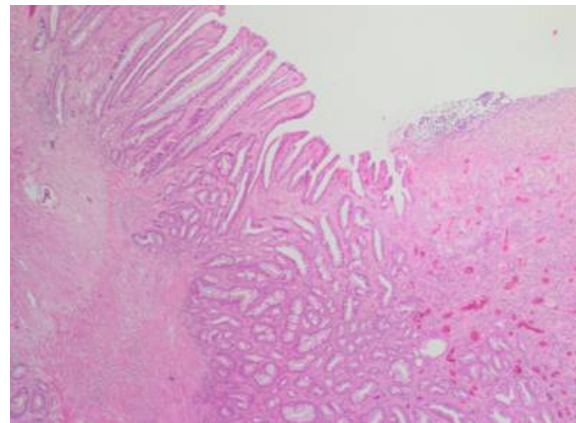


Figure 1: Colitis cystica profunda in solitary rectal ulcer syndrome (H & E staining x 10)

II. OBJECTIVE

- To study clinical, endoscopic, histological and treatment spectrum of solitary rectal ulcer syndrome and to analyze the diagnostic and therapeutic dilemma in its management.

III. METHODS

- This is a retrospective analysis of 21 patients with histopathologically proven SRUS, diagnosed at Kasturba Hospital, Manipal a tertiary care referral hospital in the West coast of South India.
- Clinical records of patients diagnosed between 2011 January to 2012 December were retrieved from the computerized filing system of the medical records department.
- The details of clinical presentations, endoscopic findings, treatment given and

progress during follow up was compiled using a proforma.

IV. RESULTS

- During the study period we had 21 patients with histopathologically proven diagnosis of SRUS. 7 (33.3%) were less than 20 years of age and 4 (19%) were above 60 years of age. The mean age was 48.5 years. 14(66.6%) were males and 7(33.3%) were females. 15(71.42%) had bleeding per rectum as the presenting complaint, constipation in 5(23.8%), passage of mucous in 5(23.8%), abdominal pain in 6(28.57%), diarrhoea in 5(23.8%) and mass per rectum in 2(9.5%). Associated hemorrhoids were seen in 3(14.2%) and rectal prolapse in 1(4.7%).
- The endoscopic findings are summarized in table 1. SRUS resembling malignancy on endoscopy was noted in 3(14.28%) of which 1(4.7%) had the biopsy consistent with malignancy. The aforementioned patient was a 28 year old female, who presented with bleeding per rectum for 3 months. Sigmoidoscopy revealed a circumferential indurated ulcer 4 cm from the anal verge, biopsy being reported as adenocarcinoma. She underwent abdominoperineal resection (Fig.2) with an uneventful post operative period. Final histopathology showed an ulcerated lesion extending into the submucosa with features suggestive of solitary rectal ulcer with no focus of carcinoma in situ or invasive carcinoma.



Figure 2: Resected specimen of the above patient.

Table 1: Endoscopic findings of patients with SRUS (n = 21)

Number	Solitary	16(76.19%)
	Multiple	5(23.8%)
Pattern	Ulcerative	20(95.23%)
	Polypoidal	1(4.7%)
Location	Anterior	1(4.7%)
	Posterior	2(9.5%)
	Circumferential	2(9.5%)
	Data not available	16(76.19%)
Distance from anal verge	<5 cm	3(14.28%)
	5 – 10 cm	5(23.8%)
	>10 cm	13(61.9%)
Diameter	<1 cm	3(14.28%)
	1 – 3 cm	10(47.61%)
	>3 cm	8(38.09%)
Features suggestive of malignancy	Everted margin of ulcer	1(4.7%)
	Hard growth	2(9.5%)
Features suggestive of inflammation	Edematous mucosa	2(9.5%)

- Bulk laxatives alone was prescribed for 9(42.85%), mesalamine for 5(23.8%) and argon plasma coagulation (APC) was tried in 5(23.8%) patients. Low anterior resection was done in 1(4.7%) and abdominoperineal resection in 1(4.7%) patient.
- Nine patients were followed up for 18 months. Others were lost for follow up. Out of 9 patients on follow up, 3(33.3%) had no recurrence of symptoms, 2(22.2%) patients had symptom free period of 1 year, 3(33.3%) patients had symptom free period of 6 months, 1 (11.1%) patient did not have any relief of symptoms(Table 2).

Table 2: clinical outcome and treatment modalities (n = 9)

Clinical Outcome	No Of Patients	Treatment Modalities
No Recurrence	3	Anterior resection (1), APR(1), Bulk laxative(1)
Recurrence in 1yr	2	APC with mesalamine (2)
Recurrence in 6 months	3	Bulk laxative (1), mesalamine (2)
No Improvement	1	Bulk laxative (1)

V. DISCUSSION

- The aetiology for SRUS is ill understood and of the several postulates, chronic straining at stools with ischaemic ulceration seems the most plausible. SRUS has also been associated with high anal pressure due to altered ability to completely evacuate the contents of rectum i.e., obstructed defecation of varying degrees, in paradoxical puborectalis contraction, internal anorectal intussusception and descending perineal syndrome [6]. Mechanical trauma due to self digitation to assist evacuation has also been implicated.
- In the current study SRUS was seen to occur in all age groups with a slight predominance in young, with wide range between 6 to 91 years. Other series have also reported such a variance in age at presentation suggesting that no age predilection exists. Our study revealed a male predominance 14(66.6%) though no definite sex predilection is discernible when all other published series are considered [1, 2, and 7].
- Rectal bleeding was the most common presenting symptom in 15(71.42%) patients which is in agreement with other studies [2, 7]. SRUS presenting as massive rectal bleeding have been reported. Other symptoms reported were constipation, mass protruding through anus, passage of mucous, abdominal pain, tenesmus and diarrhoea but these are neither specific nor distinctive. Rectal prolapse was seen in only 1(4.7%) patient in the paediatric age group and reflects the lack of paediatric patients in our series. In other series that include a larger proportion of paediatric patients [8] rectal prolapse was a common association. The presence of overt rectal prolapse in children may be attributable to diastases of levator ani.
- Endoscopically, solitary lesions were recorded in 16(76.19%) patients and multiple lesions in 5(23.8%) patients. Other series [7] report a marginally higher incidence of multiple lesions. It is evident that SRUS does not always imply the presence of only a single lesion as there can be multiple, polypoidal or circumferential lesions as well. Ulcers are usually placed anteriorly¹ because anterior rectal mucosa is more mobile and the initial part of an intussusceptum, as it descends downward into the anal canal [9].
- The size of the lesion varied from 0.5 to 5 cm in its greatest dimension. The significance of size of the ulcer is yet to be established. 1(4.7%) patient had a polypoidal lesion in our study. Other series have also reported a similar incidence with the exception of Ortega AE who reported 74% polyp or mass [3]. Distance of ulcer from anal verge ranged from 4cm to 20cm as has been seen with other series [1, 2]. It can be present in the sigmoid colon also. In the current study, 2(9.5%) patients had associated polyps in the terminal ileum; both were less than 20 years of age. It may be an incidental finding, which needs further evaluation to establish any co-relation. No other reports exist regarding this finding, most series having no comment on the ileum. Either the ileum was not evaluated or the finding was not correlated with SRUS.
- SRUS may mimic a malignant ulcer with a hard growth and everted edge. The current study had 3(14.28%) patients with endoscopic features suggestive of carcinoma. Other series and case reports have suggested that SRUS may appear as malignant on endoscopy [9]. Cornerstone histological features of SRUS are fibromuscular obliteration of lamina propria with splaying of muscularis between the crypts and diffuse collagen infiltration of lamina propria. They are not pathognomonic of SRUS but most consistent with it. Decussation and thickening of the two muscle layers, nodular induration of the inner circular layer and grouping of outer longitudinal layer are unique to SRUS. 'Diamond shaped crypts' is a salient feature of in SRUS [10]. Biopsy and histopathology may not settle the diagnostic dilemma nor provide clarity regarding further management. Colitis cystica profunda is characterized by presence of benign columnar epithelium and mucous cysts residing deep to muscularis mucosae, this histologic pattern may be confused with invasive adenocarcinoma [11].
- When severe dysplasia is reported, the surgeon may suspect under reporting because the clinical picture is suggestive of malignancy. The pathologist may even be overly cautious in reporting malignancy. The patient then becomes a candidate for abdomino perineal resection with permanent colostomy. In the current study 2 out of 3 patients had their slides send to other centers for a second opinion, due to discordance between the clinical findings and the histopathology report. The diagnosis of SRUS was confirmed and a debilitating surgery like abdominoperineal resection was avoided. However, one patient underwent abdominoperineal resection based on the diagnosis of adenocarcinoma in the endoscopic biopsy and a clinical picture suggestive of malignancy; only to find that the final histopathological report showed SRUS. This event is exceptional and rare, but makes it mandatory to get a second opinion of slides when SRUS is considered even a remotely possible diagnosis.
- A school of thought which believes that SRUS has the potential to progress to malignancy exists. It is supported by loss of hMLH1 gene expression in several cases of SRUS [10].
- Patients presenting with obstructed defecation or bleeding per rectum may require colonoscopy and rectal biopsy, followed by defecography and dynamic MRI of pelvis. The former can demonstrate recto rectal intussusceptions and the latter can reveal anorectal redundancy, lack of mesorectosacral fixation and mild to severe pelvic floor descent. Anorectal manometry, balloon expulsion test, electromyography, nerve stimulation, transit studies and endoultrasound may all contribute to diagnosis [12]. Anal endoultrasound shows marked thickening of internal sphincter, which is a striking feature of SRUS. A study using endorectal ultrasound and MRI was done

preoperatively to stage patients diagnosed to have rectal cancer based on endoscopic findings. Eight of them were diagnosed as malignant, but final biopsy was reported as SRUS^[13]. The chances of misdiagnosis are high.

- A variety of medical treatment^[7] options exist. In the current study mesalamine was used to treat SRUS in 5(23.8%) patients, bulk laxatives in 9(42.85%) patients and argon plasma coagulation (APC) in 5(23.8%) patients. Patients opting for APC may be required to undergo 4-8 sessions to achieve alleviation of symptoms^[4]. Response of patients on laxatives is varying from complete remission to no improvement of the symptom. Laxatives may not help patients with rectal prolapse or internal intussusceptions. Laxatives do not treat the aetiology. One study stated that oral salicylate or topical mesalamine and steroids are not effective^[14]. Behavioural modification or bio feedback therapy, which includes bowel habit training, avoiding excessive straining and normalization of pelvic floor co-ordination may improve symptoms, anorectal function and also facilitate healing of SRUS. Randomized control trials have established that SRUS caused by dyssynergic defecation can be effectively treated by bio feedback therapy^[3]. Behavioural modification requires follow-up reinforcing to have long term remissions. Injection of botulinum toxin into external anal sphincter is effective for a period of three months in treatment of SRUS and constipation associated with dyssynergia of defecation^[14]. Overall 66.6% of patients in our study had at least a minimum symptom free period of 6 months with conservative measures but recurrence is common.
- Surgery is restricted to non responders with full thickness rectal prolapse and intractable bleeding despite conservative management^[1]. Stapled transanal rectal resection (STARR), rectopexy or Delorme's procedure are reported to have the best success rates. 1(4.7%) patient in our series who underwent low anterior resection was completely relieved of all symptoms and had no recurrence. Laparoscopic ventral mesh rectopexy (LVMR) for internal and external prolapse causing SRUS has shown to heal the ulcer and improve the quality of life^[15].
- Defaecography and anorectal manometry were not done in the current study, hence limiting the aetiology oriented treatment. A large volume prospective study is required to draw definitive conclusions.

VI. CONCLUSION

- Solitary rectal ulcer syndrome is a misnomer because lesions may be multiple and even occasionally polypoidal.
- There is no true syndromic association except that its etiopathogenesis is ill understood and multi factorial. Though bulk laxatives, biofeedback sphincter training, topical mesalamine

and argon plasma coagulation are all useful in conservative therapy for SRUS, persistence of symptoms and relapses are common. Surgical treatment is reserved for non responders.

- SRUS may on occasion simulate malignancy both in macroscopic appearance at endoscopy and on histology of endoscopic biopsies. Hence, it is important to carefully review the endoscopic biopsy specimens to avoid performing inappropriate radical surgery with attendant major morbidity.

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