SHORT COMMUNICATION

Solitary Rectal Ulcer Syndrome: A Case Series

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A retrospective analysis of the clinical profile, endoscopic features and management of 22 children (age 18 months – 18 years) diagnosed as solitary rectal ulcer syndrome is presented. The majority (81.8%) were ≥8 years of age. Rectal bleeding was the presenting feature in all the children. Mucorrhea, constipation, tenesmus and rectal prolapse were observed in 77.3%, 63.6%, 59% and 13.6% children, respectively. Colonoscopy showed classical single rectal ulcer in 68.2% and multiple ulcers in 22.7%. Polypoidal and erosive lesions were documented in 4.5% each. The medical management comprised of bowel training and high fibre diet for all children. The other modalities included oral 5-amino salicylate (59%), sucralfate enema (4.5%) and rectal mesalamine in 9%. 64% children recovered and 13.6 % had recurrence of symptoms.

Key words: *Colonoscopy, Lower gastrointestinal bleed, Rectal ulcer.*

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Solitary rectal ulcer syndrome (SRUS) is an uncommon disorder of evacuation that affects all ages but is less common in children compared to adults. SRUS is characterized by rectal bleeding, mucorrhea, tenesmus, incomplete evacuation with characteristic colonoscopic and histopathologic features(1). SRUS is occasionally referred to as "the three- lies disease" since the lesion is not always solitary or ulcerative or restricted to the rectum(2). The incidence of SRUS in adults, is 1 in 1,00,000(3) whereas in children only few case series have been reported. We report a series of 22 cases, probably the largest ever reported from a pediatric tertiary referral center in Chennai.

METHODS

This is a retrospective study of children diagnosed as SRUS based on colonoscopic findings and confirmed by histopathology during the period May 2001-August 2009 in the Gastroenterology department of our hospital. Case records of children who underwent colonoscopy during the study period were reviewed. Only those children diagnosed as SRUS by colonoscopy and confirmed by histopathology were included in the study. The age, sex distribution, presenting symptoms, endoscopy features, histopathology and treatment details were obtained from the records and analyzed. Prior to colonoscopy all children undergo a detailed clinical evaluation of all systems including rectal examination and also investigations, including complete blood count, stool examination for parasites and ova and coagulation profile.

RESULTS

During this eight-year period, 325 children less than 18 years underwent colonoscopy for various indications. Bleeding per rectum was the most 6.7% (22 children), polyps in 27% and anal fissure in 15%. The age of children diagnosed as SRUS ranged from 18 months to 18 years (median age 10 years) of which 81.8% were ≥8 years of age. The male to female ratio in this group was 1.4:1. Chronic, intermittent rectal bleeding was the presentation in all the patients with duration between 2 to 6 months in 60% and more than 6 months in 40%. Overt rectal

prolapse was present in 3 (13.6%), mucorrhea in 17 (77.3%), straining during defecation in 14 (63.6%), digital evacuation in 6 (27.2%), constipation in 14 (63.6%) and abdominal pain/tenesmus in 13 (59%). None had evidence of liver disease or receiving medications that may cause constipation.

Full length colonoscopy was done in all the 22 children and single rectal ulcer was documented in 15/22 (68.2%) and multiple in 5 (22.7%). Ulcer size ranged from 0.2-3 cms and all were located with in 5 to 10 cms from the anal verge. In one child the lesion was polypoidal, and erosions with surrounding erythema was seen in another child. One child had, in addition, a rectal polyp situated 6 cm above the ulcer for whom polypectomy was done and confirmed as juvenile polyp. All the children had biopsy of the lesion and the histopathology revealed the diagnostic hallmark of SRUS *viz* fibromuscular obliteration of the lamina propria stroma with misorientation of smooth muscle cells (***Fig.* 1**).

The medical management comprised of bowel training and high fibre diet which was offered to all patients. 5-Amino Salicyalate (ASA) was prescribed for 2 months in 13 (59%), topical applications such as sucralfate enema in 1 (4.5%) and rectal mesalamine in 2 (9%). The youngest child presented with overt rectal prolapse and frank bleeding per rectum. This child was diagnosed earlier as probable cow's milk allergy based on the history and finding of nodular lymphoid hyperplasia on sigmoidoscopy and was referred because symptoms did not subside with withdrawal of milk proteins. Repeat endoscopy showed a large ulcer in the rectum which was treated medically; however, there was recurrence of symptoms and worsening of rectal prolapse. During the second admission, Meckel's scan and ultrasound of abdomen was done. The child required blood transfusion and underwent surgery (rectopexy) for the rectal prolapse. The majority of children improved symptomatically except three who came with recurrence of bleed. The median follow up was 6 months (range 4-24 months). None of the children had other specialized investigations such as anorectal manometry, endoultrasound or defecography.

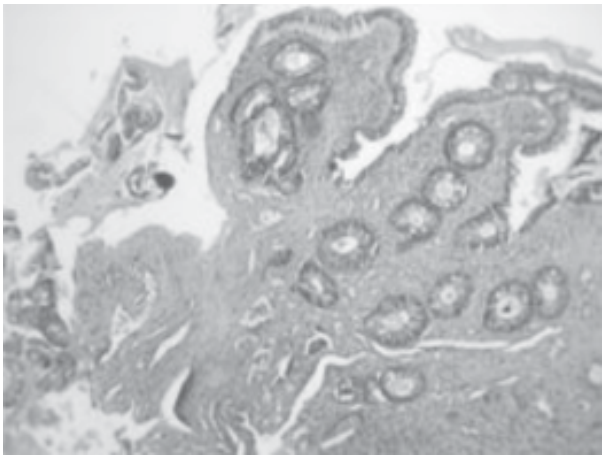

FIG. 1 *Histopathology section showing fibromuscular obliteration of the lamina propria (10 X).*

DISCUSSION

SRUS is a benign rectal disorder of defecation which is a well recognized entity in adults but often misdiagnosed or under-diagnosed in children(4). The youngest child with SRUS reported so far was 4.5 yrs(5); however, in this study, the youngest child was 1 year and 6 months. The etiopathogenesis of SRUS is not well understood but is probably secondary to ischemia and trauma to the rectal mucosa and paradoxical contraction of pelvic floor(4). The excessive straining generates a high intra-rectal pressure which pushes the anterior rectal mucosa into the contracting puborectalis muscle resulting in pressure necrosis of rectal mucosa. In addition, the anterior rectal mucosa is frequently forced into the closed anal canal causing congestion, edema and ulceration(6). Rectal bleeding was the most common presentation, observed in all 22 cases. However, blood transfusion in SRUS is rare(7). Colonoscopy in SRUS usually reveals rectal ulcer about 0.5 to 5 cm in diameter on the anterior wall(2) and similar findings were also noted in this study. Atypical features such as multiple ulcers in 30%, polypoidal lesions in 25% or lesions situated beyond the rectum have been reported and when present belies the term SRUS(2). Similar atypical features were seen in this study including confirmed as SRUS on histopathology. Rectal polyps have been reported as part of the spectrum of SRUS(8). In this study one child had a juvenile polyp proved histopathologically in addition to rectal ulcer, an observation that has not been reported in literature so far. This spectrum of presentation in SRUS stresses the importance of full length colonoscopy in children

WHAT THIS STUDY ADDS?

- Solitary Rectal Ulcer Syndrome should be considered in children presenting with rectal bleeding, mucorrhea and excessive straining during defecation.

presenting with bleeding per rectum. The differential diagnosis of SRUS includes inflammatory bowel disease, inflammatory cloagenic polyps and ulcers related to topical medications and infection, which can be differentiated by histopathology(8). The characteristic histologic features of SRUS include mucosal thickening with elongation and distortion of the glands, edema and fibrosis of the lamina propria and extension of smooth muscle fibres upward between the crypts(2,4). Rectal prolapse, either occult or overt is well documented in SRUS varies from 15 to 59 %(9) and was a feature in 13.6% of this group.

Various therapeutic regimes have been tried and the one universally recommended is high fibre diet and bowel training(2). Oral ASA and topical agents such as steroids and mesalamine have not been found effective(10). Sucralfate enemas and human fibrin sealant have shown benefit in some patients(11). Argon plasma coagulation has been utilized to treat disturbing hemorrhage(1). Behavioral modification or biofeedback therapy improves both rectal blood flow and symptoms in more than 50% and includes bowel habit training, avoiding excessive straining and normalization of pelvic floor coordination. Surgery is indicated in those with persistent bleeding per rectum not amenable to medical therapy and includes rectopexy, excision of ulcer and rarely colostomy(1).

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