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Mucosal prolapse syndrome presenting as rectal polyposis

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Abstract

Mucosal prolapse syndrome comprises a variety of clinical and histopathological entities, with mucosal prolapse as the underlying pathogenic mechanism. Due to variable clinical, endoscopic and histopathological presentation and rareness of symptomatic mucosal prolapse, misdiagnosis resulting in delayed or inappropriate treatment is frequent. This report describes a patient initially diagnosed with a colorectal polyposis syndrome consisting of multiple rectal hyperplastic and adenomatous polyps. But after careful review of medical history and histopathology, the patient was found to have a rare variant of solitary rectal ulcer syndrome presenting as rectal polyposis. The recognition of rectal polyposis as a manifestation of solitary rectal ulcer syndrome/mucosal prolapse syndrome will improve diagnosis and treatment and prevent inappropriate management of this condition.

Mucosal prolapse syndrome comprises a variety of clinical and histopathological entities, with mucosal prolapse as the underlying pathogenic mechanism. Disorders considered part of this condition include rectal prolapse, solitary rectal ulcer syndrome (SRUS), proctitis cystica profunda (PCP), inflammatory cloacogenic polyp, inflammatory cap polyps and inflammatory myoglandular polyps.^{1–3}

Clinically, SRUS is the most common manifestation of rectal prolapse. Patients often present with passage of mucus or blood per rectum on defecation. In addition, they may report tenesmus, straining, altered bowel habits, partial incontinence and a sensation of incomplete emptying. Constipation is frequently reported, and about 30% of patients acknowledge rectal digitation.⁴ Patients may also be asymptomatic.⁵

Endoscopically, SRUS can appear as shallow ulcerating lesions on a hyperaemic surrounding mucosa, but also as multiple ulcerative lesions, patchy erythematous normal mucosa or as a polypoid lesion.^{6,7} The macroscopic appearance of SRUS can be classified into ulcerative (55%), polypoid (24%) and flat (21%).⁴ Madigan and Morson reported multiple lesions in 30% of SRUS cases.⁸ Another study found that only 35% of SRUS cases presented with a solitary ulceration, whereas 22% presented with multiple ulcers, 18% with patchy granular hyperaemic rectal mucosa and 25% with a broad-based polypoid lesion.⁹ Others found polypoid SRUS in 44% of cases.⁵

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Histologically SRUS/mucosal prolapse is characterised by fibromuscular obliteration of the lamina propria with upward extension from hypertrophic and splayed muscularis mucosae. Glandular crypt abnormalities are almost always present. Secondary changes modifying the SRUS/prolapse-induced lesions include surface erosion with or without pseudomembranes, inflammation, haemorrhage, ectatic and congested vessels, submucosal fibrosis, deep cyst formation, or misplaced glands in the submucosa. The inflammatory infiltrate is usually mild.⁴ Other non-specific changes include reactive epithelial atypia and hyperplastic and distorted crypts that can cause difficulty in differentiating SRUS from inflammatory bowel disease or even cancer.

Frequently, patients with SRUS are initially misdiagnosed clinically and pathologically since SRUS is not necessarily solitary or ulcerated.^{4–6,10,11} SRUS presenting with rectal ulcers or hyperaemia is often labelled Crohn disease or mucosal ulcerative colitis, and polypoid SRUS is often diagnosed as a neoplastic polyp.¹¹

Awareness of the characteristic histology of SRUS/mucosal prolapse and the variable clinical and endoscopic presentations should improve diagnosis of this condition. Here we present an unusual case of solitary rectal ulcer syndrome presenting with multiple rectal polyps, initially misdiagnosed as a colorectal polyposis syndrome.

Case Report

A middle-aged female underwent screening colonoscopy and approximately 70 polyps were noted in the rectum. No polyps were noted in the remainder of the colon. During that procedure, a third of these polyps were endoscopically removed. During a second colonoscopy another 30 polyps were removed, but numerous others remained (fig 1A). At presentation, the polyps were sessile to semipedunculated, ranged in size from 0.1 to 1 cm, and were initially histologically diagnosed as a mixture of hyperplastic and tubular adenomatous polyps. Because of the diagnosis adenomatous polyposis, the patient was referred to a specialised colorectal cancer risk assessment clinic for further assessment.

The patient had no family history of adenomatous polyposis, or colorectal, uterine or ovarian cancer.

The patient reported a 20-year history of constipation and frequent rectal digital manipulation to aid in defecation. For at least a decade, she had been taking prune juice and using rectal suppositories and had a history of a rectocele. Otherwise, she denied haematochezia, epidermoid cyst, supernumerary teeth, and pigmented ocular fundic lesions. Her past medical history included hysterectomy and sarcoidosis. The physical examination was unremarkable.

Histopathological review of the biopsies by an expert gastrointestinal pathologist revealed polyps with prominent lamina propria smooth muscle and epithelial serrated changes (fig 1B and C). This appearance raised the possibility of hyperplastic/serrated polyposis syndrome, but the sampled polyps lacked the architectural pattern of sessile serrated adenoma. The prominent smooth muscle was regarded as the result of prolapse since smooth muscle invested individual glands rather than clusters of glands; this is a feature of Peutz–Jeghers polyposis. The polyps were classified as inflammatory myoglandular polyps.

The combination of clinical and histopathological characteristics, including smooth muscle in the lamina propria, the absence of adenomatous change, the confinement of polyps to the rectum, the history of constipation, rectal manipulation and rectocele, strongly suggested that these polyps were an expression of a variant of rectal prolapse/solitary rectal ulcer syndrome.

Discussion

Mucosal prolapse syndrome/solitary rectal ulcer syndrome can have multiple and variable expressions. Here an unusual case of mucosal prolapse syndrome/SRUS with multiple rectal polyps mimicking a hyperplastic or adenomatous polyposis syndrome of the rectum is presented.

Only few reports of multiple prolapse-induced polyps exist (table 1). One described seven patients with 2–12 prolapse-induced polyps² and another a patient with 30 polyps.³ Also, two children with rectal prolapse presenting as rectosigmoid polyposis were initially diagnosed as having a colonic polyposis syndrome.¹² Another case of SRUS/mucosal prolapse syndrome was initially erroneously diagnosed as Peutz–Jegher polyposis.¹³

Careful histopathological examination of the polyps is important in the diagnosis of mucosal prolapse syndrome. Characteristic features of mucosal prolapse are thickening and disruption of the muscularis mucosae and muscularisation of the lamina propria resulting in characteristic fibromuscular obliteration of the lamina propria with disorientation of muscle fibres.⁸ The lamina propria may be oedematous and often shows increased numbers of fibroblasts.^{2 14} Also, mild disruption of the normal architecture with elongation, displacement and distortion of glands and ulcerations are often accompanied by chronic inflammation.^{7 8}

Hyperplastic and serrated changes of the mucosa, including exaggerated serration within the lower crypt compartments, crypt branching, hypermucinous appearance of the epithelium, and horizontal extension of crypt bases along the muscularis mucosae have been reported in 38–66% of cases of mucosal prolapse.^{7 14 15} In particular, the polypoid variant of SRUS shows regenerative hyperplastic changes, such as a relatively high incidence of mucous cell proliferation, dilatation of glands, and serrated change.¹⁶

Additional histological features of mucosal prolapse/SRUS include diamond-shaped crypts and mucosal elastin that can be delineated with Elastic van Gieson staining.¹⁷ Also, demonstration of diffuse excess of mucosal collagen (H&E with added saffron) and mucin stain to identify glands in the muscularis mucosae or submucosa can be helpful in making the diagnosis.¹⁸ The presence of colonic crypts with cystic change in the muscularis mucosae or a muscularised submucosa is referred to as “localised colitis/proctitis cystica profunda”.⁷ Lastly, microvascular abnormalities, including the presence of fibrin in the vessel wall and thickening of mucosal vessels, have been described in mucosal prolapse with ulceration.¹⁹

Take-home messages

- Mucosal prolapse syndrome/solitary rectal ulcer syndrome (SRUS) is regularly misdiagnosed, resulting in delayed or inappropriate treatment.
- Endoscopically, SRUS may appear as ulcers, erosions or polypoid lesions; these are not necessarily solitary but can be multiple or present as rectal polyposis.
- Careful histopathological review and clinical history are important in establishing mucosal prolapse syndrome/SRUS diagnosis.
- Fibromuscular obliteration of the lamina propria is the most characteristic histopathological feature of mucosal prolapse syndrome. Also, disruption of the normal crypt architecture, hyperplastic and serrated changes of the crypts, and chronic inflammation, can be seen.

- Clinical features suggestive for mucosal prolapse syndrome/SRUS are incomplete defecation, excessive straining, rectal digitation, partial incontinence, constipation, diarrhoea and rectal prolapse.

Together one or more of the histopathological features described above are present in many (polypoid) inflammatory lesions of the colorectal and anal regions. These characteristics may be seen in entities such as SRUS, PCP, inflammatory cloacogenic polyp, inflammatory cap polyps and inflammatory myoglandular polyps.^{1–3,20} Prolapse is regarded as the unifying pathogenic mechanism accounting for all these lesions, and polypoid change with or without ulceration is dependent on the severity of the prolapse. Variations in histological pattern are likely to represent different stages of temporal development and are not considered sufficient for separate categorisation of these entities.³

The polyps in the present case are most consistent with inflammatory myoglandular polyps. These lesions are regarded as a manifestation of mucosal prolapse syndrome by most authors,³ although some have suggested that they are a distinct entity from mucosal prolapse syndrome.²¹

In addition to careful histopathological review, clinical history is important in establishing the diagnosis. Almost all patients have difficulties with defecation such as incomplete defecation, excessive straining, rectal digitation, partial incontinence and altered bowel function including constipation and diarrhoea.⁴ Rectal prolapse is found in about a third of individuals. About a quarter of patients are asymptomatic.⁵

In summary, we have described a patient with SRUS/rectal prolapse presenting with multiple inflammatory myoglandular polyps, initially diagnosed as a hyperplastic and/or adenomatous polyposis syndrome. However, after careful re-review of the histopathology and the patient's medical history, the rectal polyposis is consistent with a variant expression of mucosal prolapse syndrome instead of a premalignant polyposis syndrome. This case illustrates that rectal polyposis can be a rare expression of mucosal prolapse/SRUS.^{2,3,12,13} Knowledge of the clinical and histopathological variations of this condition will prevent misdiagnosis, and inappropriate management of these patients.

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Figure 1.

Endoscopic and microscopic pictures of the rectal polyps presented in this case report. (A) Endoscopic picture of the rectum following the second colonoscopy, showing multiple sessile rectal polyps. (B) Low-power magnification showing hypermucinous changes with crypt dilatation (asterix) and subtle epithelial serration reminiscent of that in hyperplastic polyps (arrow); these are common features in prolapse lesions. (C) High-power magnification showing smooth muscle proliferation in the lamina propria. Note that the smooth muscle proliferated among individual glands, replacing the lamina propria, rather than separating groups of glands in an arborising pattern (arrow).

Table 1

Patients with rectal mucosal prolapse syndrome presenting as polyposis

	Age (years)	Gender	No. of polyps	Histology	Histopathological differential diagnosis
Daniel <i>et al</i> ¹²	14	M	Multiple	Inflammatory (cap) polyps	Inflammatory/juvenile/hamatomatous polyposis
Daniel <i>et al</i> ¹²	13	M	Multiple	Inflammatory pseudopolyps	Adenomatous/hyperplastic polyposis
Jansen <i>et al</i> ¹³			NS	NS	Peutz-Jeghers polyp
Tendler <i>et al</i> ²	33	M	2	NS	NS
Tendler <i>et al</i> ²	45	M	6	NS	NS
Tendler <i>et al</i> ²	54	M	5	NS	NS
Tendler <i>et al</i> ²	46	M	6	NS	NS
Tendler <i>et al</i> ²	60	F	3	NS	NS
Tendler <i>et al</i> ²	61	F	12	NS	NS
Tendler <i>et al</i> ²	71	M	4	NS	NS
Chetty <i>et al</i> ³	48	M	30	Inflammatory cap polyps and inflammatory myoglandular polyps	NS
Present paper	51	F	70	Inflammatory myoglandular polyps	Adenomatous/hyperplastic polyposis

NS, not stated.