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CLINICAL NOTE

"The three-lies disease": Solitary rectal ulcer syndrome

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RESUMEN

El síndrome de la úlcera rectal solitaria es una entidad benigna poco común cuyos síntomas más frecuentes son la rectorragia y el dolor anal. Los hallazgos anatomopatológicos son típicos. Se suele evidenciar engrosamiento de la mucosa, elongación y distorsión de las glándulas, una lámina propia edematosa con gran cantidad de colágena y engrosamiento de la muscularis mucosae. El diagnóstico se realiza mediante endoscopia con toma de biopsias. No siempre se trata de lesiones ulceradas. Se suele localizar preferentemente en la cara rectal anterior y/o lateral, aunque hasta un 30% de las lesiones son múltiples, existiendo casos de afectación del colon sigmoide y descendente. Por tanto, la presentación puede ser heterogénea y este es el motivo por el que esta entidad es también conocida como la "enfermedad de las tres mentiras". Presentamos un caso de síndrome de la ulcera rectal solitaria manifestado endoscópicamente como una placa eritematosa localizada en la cara lateral izquierda del recto.

Palabras clave: Síndrome de la úlcera rectal solitaria. Hemorragia digestive baja. Molestias anales.

ABSTRACT

Solitary rectal ulcer syndrome is an uncommon benign condition characterized by rectal bleeding, passage of mucus, and pain. Histological features are well established as obliteration of the lamina propria by fibrosis and smooth-muscle fibers extending from a thickened muscularis mucosa to the lumen. Diagnosis can usually be made on sigmoidoscopy, and biopsies should always be taken. Ulceration is not universally present, and polypoid, non-ulcerated lesions and erythematous areas are also seen. The lesion or lesions are most often found on the anterior or anterolateral wall of the rectum, although they can also be located in the left colon and be more extensive or even circumferential. Lesions are multiple in 30 percent of cases. These are the reasons why this entity is also known as "the disease of three lies". We report a case of solitary rectal ulcer syndrome presenting at endoscopy with an erythematous area on the left side wall of the rectum.

Key words: Solitary rectal ulcer syndrome. Lower digestive hemorraghe. Anal discomfort.

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INTRODUCTION

Solitary rectal ulcer syndrome (SRUS) is a chronic benign disorder with characteristic clinical, endoscopic, and histologic manifestations (1,2). In 1969, Madigan and Morson defined its clinicopathological features for the first time after a review of 68 cases (3).

SURS is an infrequent entity with an estimated incidence of 1-3 cases per 100,000 persons per year (4,5). The condition is rare and so misdiagnosis with other digestive diseases such as cancer or inflammatory bowel disease is common. In one study with 25 patients the average time from symptom onset to definitive diagnosis was 5 years. Patients with patches of hyperemic mucosa were misdiagnosed with ulcerative colitis or Crohn's disease whereas polypoid lesions were confused with neoplastic polyps or even cancer (6).

We report a case of SRUS that presented as an erythematous area located on the left side of the rectum. Biopsies confirmed the diagnosis of SRUS although the macroscopic appearance at colonoscopy initially suggested inflammatory bowel disease or stercoral ulcer.

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CLINICAL NOTE

A 42-year-old woman with a history of mild rectal bleeding at the end of defecation, feeling of incomplete emptying, and anorectal pain was referred to our Gastroenterology Unit for evaluation in January 2005. She had no history of previous illness and there were no important disease in her family. She denied using suppositories or any other medication. Physical examination was normal except for the rectal exam, which revealed a firm zone on the left side of the rectum. Complete blood count and basic biochemical parameters were within normal ranges. Colonoscopy showed two small rectal polyps, internal hemorrhoids (one of them prolapsed), and a 2 x 2-cm patch of erythematous mucosa located on the left side of the rectum with well-delimited borders. These findings initially suggested inflammatory bowel disease, stercoral ulcer, or solitary rectal ulcer syndrome. Rectal biopsy showed a fibrous obliteration of the lamina propria, which was replaced with smooth muscle and collagen, and a thickened muscularis mucosa with distortion of crypt architecture, all hallmarks of solitary rectal ulcer (Figs. 1 and 2). Finally, anorectal manometry was performed. A reduced maximum tolerable rectal volume and a prolonged balloon expulsion time were detected.

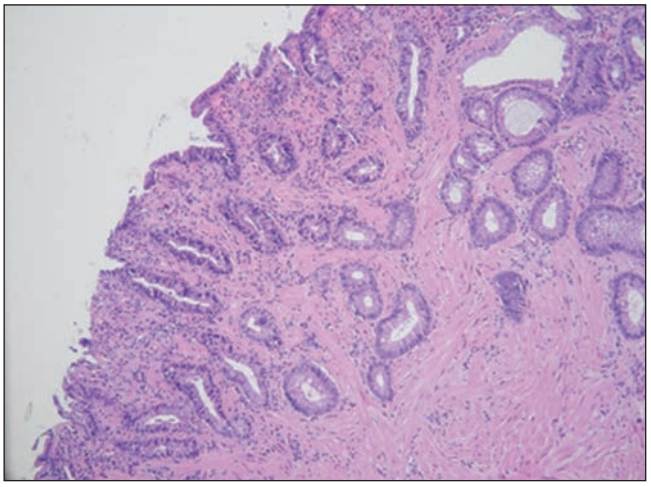

Fig. 1. Small magnification. A fibromuscular obliteration of the lamina propria can be observed.

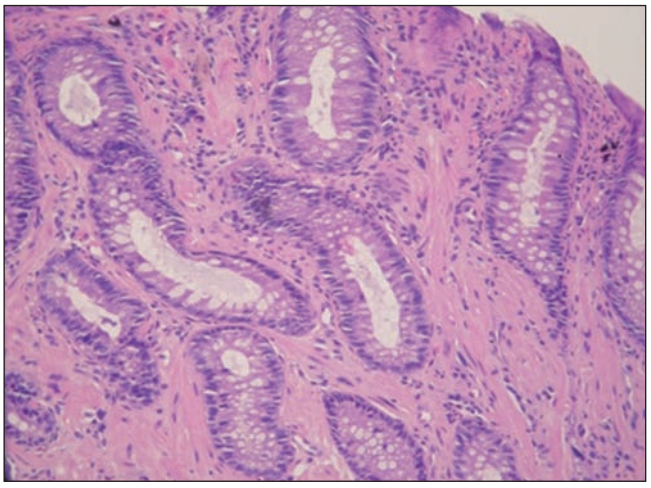

Fig. 2. Great magnification. A thickned muscularis mucosae and the extension of smooth-muscle fibres from the muscularis mucosae vertically upwards between crypts may be observed.

RESULTS

Treatment with high-fiber diet and 2-g sucralfate enemas twice daily was tried for three months. Biofeedback sessions were initiated simultaneously with pharmacological treatment and maintained for 9 months. The patient became free of anorectal pain, and the amount of bleeding was significantly reduced after two months of treatment. In October 2005 a new colonoscopy was performed. A well-delimited hyperemic and edematous patch of mucosa was found. New biopsies were obtained and the previous diagnosis was confirmed. At the moment (February 2007) the patient is asymptomatic even though endoscopic findings still remain.

DISCUSSION

Solitary rectal ulcer syndrome (SRUS) is a chronic benign disorder with characteristic clinical, endoscopic, and histological manifestations (1). The most common symptom is the passage of small amounts of red blood on defecation (60%). Passage of mucus is reported in 18% of cases and constipation in 55%. A sense of incomplete defecation is common, and patients usually have a feeling of unsatisfied defecation. Another common complaint is anorectal or abdominal pain. Diarrhea is seen in 20% of patients, and up to 25% can be asymptomatic, incidentally discovered while investigating other diseases (7). Macroscopically, SRUS typically appears (57% of cases) as an isolated shallow ulcerating lesion (rounded, oval, or linear) on a hyperemic mucosa, most often located on the anterior or anterior-lateral wall of the rectum, that may range from 0.5 to 5 cm in diameter. Up to 30% of cases consist of multiple ulcers and can be located in the sigma or descending colon (8). In all, 25% of SRUSs may appear as a polypoid lesion, and 18% may appear as patchy mucosal erythema. We report a case of solitary rectal ulcer syndrome presenting at endoscopy as an erythematous area with well-delimited borders –the less frequent presentation of SRUS. Because of the wide endoscopic spec-

trum of SRUS (not always ulcerated, not always solitary, and not always located in the rectum) this disease is also known as "the three-lies disease". It is crucial to take biopsy specimens from the involved area for a positive confirmation of the diagnosis. These lesions are sometimes underestimated during endoscopic examination and are not biopsied; moreover, our variety is only manifestated as a small patch of hyperemic mucosa. They can be confused with minor trauma by the colonoscope on the colorectal mucosa, or attributed to the type of colon preparation (more frequent with the use of phosphate-derived solutions). The histological appearance of SRUS has been well established (Fig. 1 and 2). The mucosa is elongated with distorted glands, especially at the base. The lamina propria is edematous and contains a proliferation of fibroblasts ("fibromuscular obliteration"). There is a thickening of the muscularis mucosae and an extension of the smooth muscle fibres from the *muscularis mucosae* upwards between crypts. (8). Anorectal funtion tests usually do not help in establishing the diagnosis or predicting therapeutic response. SRUS can be occasionally misdiagnosed as colorectal adenocarcinoma. SRUS is accepted as a chronic, benign disorder. Tsuschida et al. reported the first case of SRUS accompanied by dysplasia and well-differentiated adenocarcinoma invading the submucosal layer, so they speculate whether the carcinoma originated in the SRUS-involved mucosa (9). SRUS is also difficult to distinguish from inflammatory bowel disease, especially ulcerative colitis. Uza et al. reported SRUS associated with long-standing ulcerative pancolitis. A follow-up colonoscopy for this patient revealed the presence of a protruding lesion in the anterior wall of the lower rectum with a well-demarcated and longitudinal ulcer on the proximal side from this mass. The biopsy specimen from this site revealed proliferated smooth muscle fibers in the lamina propria, thus resulting in a diagnosis of SRUS. Yearly surveillance with colonoscopy and biopsy was performed, and revealed no obvious changes in the lesion (10). Other conditions we must take into account for differential diagnosis include infectious colitis, ischemic colitis, deep cystic colitis, stercoral ulcer, and traumatic ulcer. The cause of SRUS is unknown, and histopathological studies suggest a spectrum of disease, raising the possibility of a variety of causes. It is generally agreed that rectal prolapse and paradoxical contraction of the pelvic floor muscles are among the factors involved in the development of SRUS (11). A prolapsed mucosa may also become directly traumatized against a closed anal canal, resulting in isquemic trauma and ulceration. Digital evacuation in persons with constipation may lead to direct trauma to the mucosa and ulcer formation (12). Also, an infectious etiology has been proposed. An atypical case of this syndrome due to *Mycobacterium chelonae* has been reported (13). Treating SRUS is difficult, and there are no definite recommendations for SRUS therapy. A useful approach would consist of patient reassurance and education aiming at long-term cooperation. Several treatment options can be used: Behavioral modification, topical agents, biofeedback, and surgery. Anyway, symptoms may be improved by treatment, but achieving endoscopic and histological normality is uncommon (as occurred in the case we report). Our patient was treated with high-fiber diet and sucralfate enemas associated with biofeedback. Once the diagnosis is established, patients should be instructed on a high-fiber diet and the use of bulk laxatives. These dietary and behavioral modifications are especially effective in patients with mild to moderate symptoms, and in the absence of significant mucosal prolapse. Several topical agents, sometimes the same used for the treatment of inflammatory bowel disease, have been used with variable success rates. Zagar et al. conducted an open-label study and reported clinical improvement with the use of 2-g sucralfate enemas twice daily for 6 weeks, with a mean follow-up of 8 months. They performed colonoscopies both before and after treatment, and demonstrated a macroscopic healing of ulcers in all patients (14). Topical glucocorticoids and sulfazalazine enemas are not effective (15). Encouraging results from the use of behavioral therapies for defecation disorders have led to studies exploring whether some patients with SRUS may benefit from biofeedback retraining. Biofeedback has been used both as the sole therapy and as an adjunct to surgical therapy. This non-invasive therapy with no side effects improves symtomps by altering the gut's autonomic pathways (16). It is estimated that one third of patients are refractory to conservative treatment and will need surgery. About 50% of these patients have rectal prolapse. Usually mucosal resection or rectopexy is performed (17).

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