Pediatr Pol 2022; 97 (2): 156-158
DOI: https://doi.org/10.5114/polp.2022.116815
Submitted: 07.02.2022, Accepted: 25.03.2022, Published: 10.06.2022

PEDIATRIA Polska
Polish Journal of PAEDIATRICS

CASE REPORT

An unusual aetiology of solitary rectal ulcer syndrome

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ABSTRACT

Solitary rectal ulcer syndrome (SRUS) is a rare condition, usually affecting young adults, with few cases in children. Solitary rectal ulcer syndrome appears in the mucosa of the large intestine, typically the rectum, and has the form of singular or multiple lesions – most commonly ulcerations. Its aetiology remains unclear. However, there are some theories explaining its onset. The disease can manifest, e.g., as a rectal bleeding, mucoid discharge or abdominal pain. There is a particular constellation of features in the histopathological examination that confirms the diagnosis of SRUS. In spite of the fact that there are many pharmacological and non-pharmacological forms of treatment, managing SRUS remains unsatisfactory.

The paper describes a case of a patient with a very unusual cause of SRUS – tailgut cyst. It is reported to raise the awareness of such a disease and emphasize the importance of further testing when the treatment fails.

KEY WORDS:

constipation, rectal bleeding, gastroenterology, paediatrics, ulcers.

INTRODUCTION

Solitary rectal ulcer syndrome (SRUS) is often called 'the three lies disease': despite what its name suggests, the syndrome affects not only the rectum, but also the sigmoid colon; moreover, there is not necessarily only a single lesion and ulceration is not always present. Clinically, it usually manifests as rectal bleeding – mucoid discharge from the rectum or tenesmus [1–5].

The pathophysiology of SRUS remains unclear, and there are several conditions that can explain its onset. The case presented below appears to confirm the ischaemic theory of SRUS [2, 3, 6, 7]. Our paper emphasises that the presence of one rare disease does not rule out the presence of another. Moreover, the relationship between them can be crucial for choosing the strategy of treatment.

CASE REPORT

A 7-year-old boy was admitted to the Department of Paediatric Gastroenterology and Nutrition with suspicion of mechanical obstruction and anaemia. He had been presenting with faecal incontinence, enlarged abdominal circumference and periodic abdominal pain for approximately a year. He had reported progressive weakness for the previous 2 weeks. An X-ray revealed distention of the colon with air-fluid levels. Blood tests showed decreased haemoglobin (8.2 g/dl) and ferritin (8.0 µg/l). Serology was performed to rule out celiac disease. The masses of stool were suspected to be the cause of the obstruction; thus, an enema was performed. Iron supplements and laxatives were prescribed, and the patient was discharged from the hospital.

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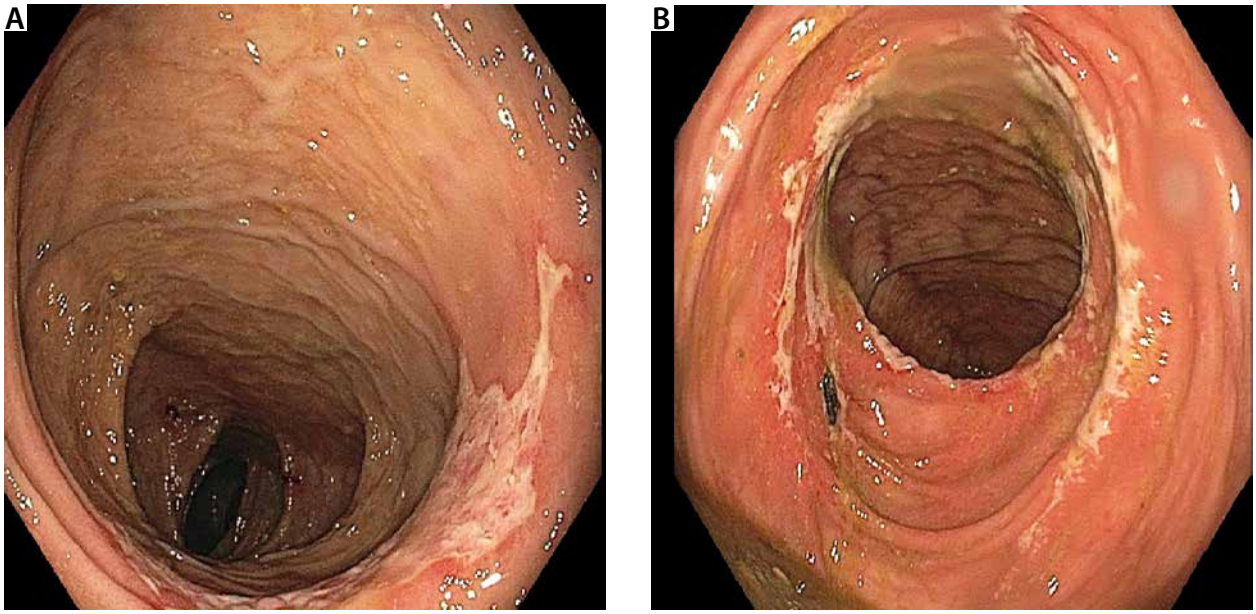

FIGURE 1. **A**) Solitary rectal ulcer visualised during the first colonoscopy, showing the increase in size. **B**) Solitary rectal ulcer visualised during the second colonoscopy, showing the increase in size

Two years later, the patient returned to the hospital with signs and symptoms similar to those present previously. A faecal occult blood test was positive; thus, a colonoscopy was performed. The examination showed a solitary rectal ulcer, which was also confirmed in the biopsy. Treatment with sucralfate, sulfasalazine, and prednisone was initiated. The next colonoscopy, performed 4 months after the first, revealed that the ulcer had increased in size (Figure 1), so argon plasma coagulation therapy was introduced. The patient received a total of 8 sessions in 5 months; nevertheless, no improvement was observed. Accordingly, an magnetic resonance imaging (MRI) scan was performed, revealing a solid-cystic mass (Figure 2). The mass was localised in the retrorectal space and seemed to press the rectum from the outside in the same area where the ulcer was revealed. Surgical consultation resulted in the decision to excise the tumour. Histopathological analysis confirmed the diagnosis of a tailgut cyst. Unfortunately, excision of the tumour did not cause improvement of the symptoms.

DISCUSSION

The aetiology of SRUS is not well understood. Several possible mechanisms of ulceration have been described, one of which is excessive straining during defecation with paradoxical contractions of the pelvic floor muscles. This leads to increased intra-abdominal pressure, which push-

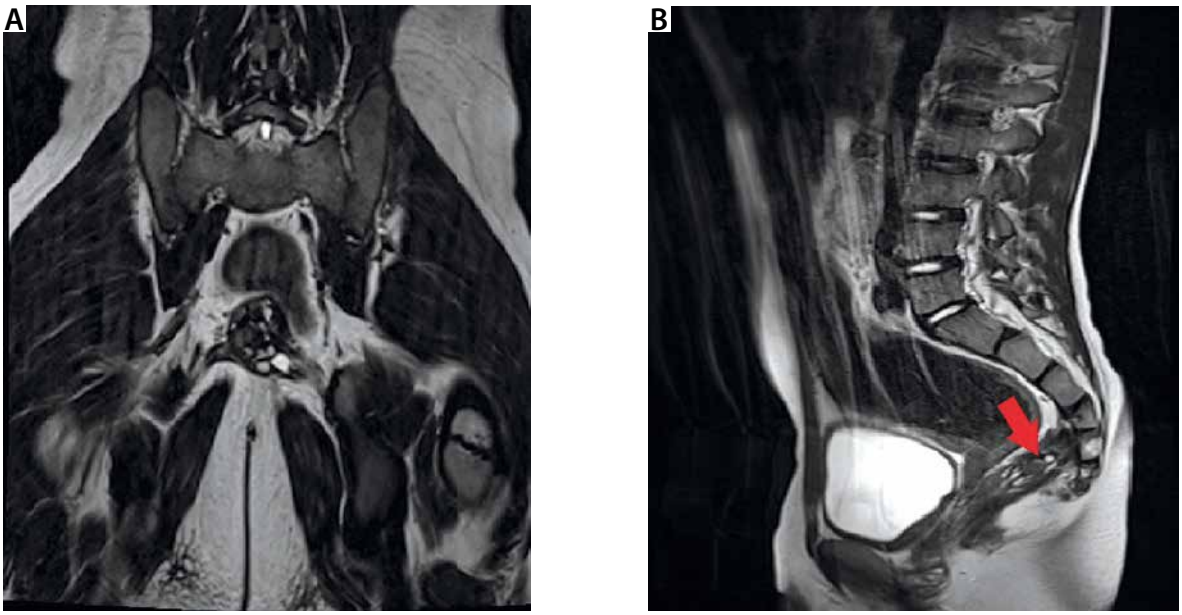

FIGURE 2. Magnetic resonance imaging scan showing a solid-cystic mass (indicated with an arrow) in the retrorectal space

es the mucosa of the anterior rectal wall into the contracting puborectalis muscle and consequently into the closed anal canal. This causes hyperaemia and oedema, then hypoperfusion and ischaemia, and finally mucosal ulceration. In the case presented herein, the tumour compressing the intestine wall caused ischaemia and formation of the lesion [2, 3, 6, 7].

Other possible causes include a malfunction of the external anal sphincter, which may create a reverse pressure gradient in the rectum and cause abnormal defecation [7]. It is also possible for local intussusceptions to develop during excessive straining. Solitary rectal ulcer syndrome can also be caused by direct trauma, mostly repetitive self-digitation or as a result of sexual abuse [2, 3, 6].

The incidence of SRUS is approximately 1 in 100,000 persons per year, and it is less common in childhood [3]. Among adult patients, there is a slight predominance in women [3], but 75–80% of children with SRUS are boys [3–5]. Among children < 18 years of age, the median age at diagnosis is 10 years [3].

Manifestations of the disease vary, and may include rectal bleeding (most common), mucoid discharge, straining during defecation, abdominal or perineal pain, tenesmus, diarrhoea, sensation of incomplete defecation, constipation, rectal prolapse, and rectal digitation [3, 7].

Solitary rectal ulcer syndrome may present as an ulceration, hyperaemic mucosa, or polypoid fragment of a tissue. It is usually localised on the anterior wall of the rectum and measures 0.5–4 cm in diameter [1–5].

Diagnosis of SRUS should be confirmed with a histological examination, which can show features such as fibromuscular obliteration and deposits of collagen in the lamina propria, spreading of fibroblasts and muscle fibres between the crypts, the presence of distorted crypts, and hypertrophy of muscularis mucosa. In addition, erosions, inflammation, and atypical cells can be seen in the epithelial layer. These findings are important in differentiating SRUS from other diseases like inflammatory bowel disease or tumours [3–5, 7].

Recent studies show that defecography, barium enema, anorectal manometry, dynamic MRI, and ultrasounds can be useful in recognising and evaluating the condition, but they are not required for diagnosis [3, 7].

Treatment of SRUS remains unsatisfactory. We know especially little about the management of SRUS in children. Methods of managing the disease include appropriate education, implementing a high-fibre diet, using stool softening medications, and avoiding excessive straining during defecation [3, 7]. Enemas can be helpful in alleviating the symptoms of constipation [3, 5].

Pharmacological options include drugs used for the treatment of inflammatory bowel diseases, such as sulfasalazine, mesalamine, or corticosteroids. However, response to these drugs is usually suboptimal [3, 7]. Other options include substances that form protective barriers at the surface of an ulcer, such as sucralfate (with simultaneous enema) or endoscopically applied human fibrin sealant [3, 5, 7]. Botulinum toxin injections into the external anal sphincter can also be helpful in patients with dyssynergic defecation; however, its effect is temporary, lasting up to 3 months [3].

Among the invasive methods of managing SRUS, argon plasma coagulation therapy is a safe haemostatic modality [7]. Surgical approaches include rectopexy, excision, and Delorme's procedure [3]. The approach chosen depends on the severity of symptoms.

One of the newest treatment strategies for SRUS is biofeedback, which can improve pelvic floor muscle coordination and consequently relieve symptoms [3, 7].

CONCLUSIONS

In the presented case, a tailgut cyst led to the development of SRUS. Tailgut cyst is a rare lesion that is usually found among adults, with few cases reported in children. It is probably a remnant of the embryonic tailgut, which usually regresses over time. Fifty per cent of these tumours are asymptomatic, while others lead to symptoms of a pelvic mass. Differential diagnosis includes teratomas, rectal cysts, chordomas, or meningocele. Surgical excision is the method of treatment [8].

Our paper highlights the importance of proper diagnosis and the need for further testing in cases of treatment failure, such as the case we present herein.

DISCLOSURE

The authors declare no conflict of interest.

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