

Rectal Prolapse as an unusual presentation of celiac disease: report of two cases

Prolapso rectal como presentación inhabitual de enfermedad celiaca: presentación de dos casos

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Received: 20-03-2017; Accepted: 22-08-2017

Abstract

Introduction: Rectal prolapse (RP) is related to an increase of intra-abdominal pressure, pelvic floor disease or anal sphincter. The most common causes of RP are constipation, cystic fibrosis, whooping cough and dysenteric diarrhea. However, celiac disease is not considered among the pathologies related to RP. **Objective:** To present a scarcely described association between RP and celiac disease. **Clinical Cases:** We presented 2 preschoolers in whom the reason for consultation was RP, whose study was focused on as prolonged diarrhea, due to the antecedent of pasty consistency of stools. The tests showed elevated anti-tissue transglutaminase (anti-tTG) antibody titers, and duodenal biopsies with villous atrophy and increased intraepithelial lymphocytes, consistent with celiac disease. Both had an excellent response to the gluten-free diet, with rapid normalization of depositions, without presenting any episodes of RP after treatment. Both with normal staturo-ponderal development and anti-tTG-negative controls at the annual 5-year follow-up. **Conclusions:** Although the association between RP and celiac disease has not been described yet, it should be considered in diagnosis and treatment.

Keywords:

Rectal prolapse,
celiac disease,
chronic diarrhea

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Introduction

Rectal prolapse (RP) is a clinical condition that occurs mainly in children younger than 5 years old^{1,2} due to factors or pathologies that involve an increase in the intra-abdominal pressure and/or weakness of surrounding tissues. The anorectal region anatomy in pediatric age increases the susceptibility to RP³, since the rectal mucosa is adhered loosely to the muscle layer⁴, and the rectum of the children is more vertical than the adult rectum^{5,6}.

The associated pathologies described in the literature are severe constipation of functional or organic origin (Hirschsprung's disease)^{7,8}; acute diarrhea, especially those with dysentery⁹; respiratory pathologies related to severe cough that produce a significant increase in the intra-abdominal pressure (Bordetella pertussis infection, cystic fibrosis)^{10,11}; malnutrition¹²; neurogenic bowel and Ehlers-Danlos' Syndrome^{8,13}.

The diagnostic approach must be symptomatically oriented, considering that constipation is the main cause of RP in developed countries^{8,14}.

Traditionally, therapeutic measures have been conservative, since most of the cases are self-limiting¹⁴. The therapeutic approach is focused on a proper constipation management, which includes a high-fiber diet to improve defecation habit, and the use of osmotic substances, such as polyethylene glycol¹⁵. In case of a triggering pathology such as the mentioned above, it must be studied and treated accordingly. Patients with recurrent prolapses that do not respond to the constipation treatment, and without any other pathologies that could explain it, can be candidates for a surgical procedure³.

Regarding other possible causes of RP, there are only two reports of association between the celiac disease and the RP. The first one describes the case of a 73 years-old woman with chronic diarrhea and RP¹⁶. More recently a clinical case of this association was reported in a pediatric patient¹⁷.

The objective of this report is to describe an association between RP and the celiac disease in two pediatric patients, without pre-existing pathology, whose reason for medical consultation was recurrent RP.

Clinical Cases

Case 1

A male patient, 3 years and 7 months old at the time of consultation, second child of non-blood relative healthy parents. He received exclusive breastfeeding until 6 months of age, and then non-dairy food were added, including gluten. There was no relevant morbid history; the patient had a normal growth.

The mother consulted the pediatric gastroenterologist after two episodes of RP related to defecation, which required an active reduction by the mother.

The patient did not have a respiratory disease history, such as recurrent bronchitis, pneumonia or sinusitis that could indicate cystic fibrosis, underweight or decline of the growth curve. The defecation habit of the patient did not catch the attention of the parents, however, after some questions, the parents agreed that for the last 12 months, the child defecated 3 to 4 times a day, alternating between normal and pasty stools, associated with mild meteorism, without remarkable smell or oily aspect, and also, there was no presence of blood or mucus. The child had no abdominal pain, loss of appetite, vomiting or any other gastrointestinal discomfort. The parents reported a temporal relation between the onset of the symptoms and an increase in the intake of candies, however, the symptoms slightly improved after the interruption of the intake.

The patient, at the date of consultation, had a normal nutritional evaluation of weight and height, according to the WHO reference¹⁸ (weight: 16 kg and height: 1mt). In the physical examination, atopic dermatitis and mild meteorism were found, with no other findings. The anal and lumbosacral exams were normal.

The study focused on chronic diarrhea due to the frequent pasty consistency of the stools and the 12-months period of evolution. Before proceeding with other studies, the possibility of the celiac disease as a first diagnosis was dismissed. The titer of anti-tissue transglutaminase antibodies (anti-tTG Ab) was very high, 276 EU/ml (normal value: < 20) (Table 1), which led to an upper endoscopy, which showed an atrophic duodenal mucosa. The histological study of the duodenal biopsies showed an acute villous atrophy and an increase in the intraepithelial lymphocytes (> 30 lymphocytes/100 enterocytes), this is compatible with an enteropathy-type 3b in the Marsh-Oberhuber classification for celiac disease¹⁹ (Figure 1).

After the confirmation of the diagnosis, a regime without gluten or lactose during the first month was indicated, considering the damage in the duodenal mucosa, it was highly possible that the patient would have an associated transitory malabsorption of lactose, which can be recovered after the stabilization of the intestinal mucosa. In this episode, no other causes of RP were investigated, since the patient did not have respiratory symptoms that could indicate cystic fibrosis. The onset of diarrhea was at the age of 2 years and 7 months old and not right after the birth, without previous history of constipation or any abnormal lumbosacral examination that could indicate an innervation disorder.

Table 1. Laboratory test of the two patientes before treatment

Laboratory test	Pacient 1	Pacient 2	Reference
Erythrocyte count	33.5%	37%	35-45%
Hemoglobin	11.7 g/dL	12.5 g/dL	11.5-14.8 g/dL
MCV	77 fL	73 fL	77-95 fL
Leukocytes	8.260/mm ³	7.100/mm ³	4.500-13.500/mm ³
Platelets	285.000/mm ³	308.000/mm ³	150.000-450.000/mm ³
CRP	1 mg/L	1 mg/L	< 10 mg/L
Albumin	4.2 gr/dL	4.6 gr/dL	3.8-5.4 gr/dL
AST	42 U/L (< 52)	34 U/L	5-52 U/L
ALT	22 U/L (< 39)	26 U/L	5-39 U/L
Alkalyne phosphatase	195 U/L	125 U/L	80-269 U/L
IgA	92 mg/dL	221 mg/dL	19-119 mg/dL
Anti-transglutaminase Ab	276 EU/ml	118 EU/ml	< 20 EU/ml

MVC: mean corpuscular volume. CRP: C reactive protein. AST: aspartate amimotransferase. ALT: alanine aminotransferase. IgA: Immunoglobulin A.

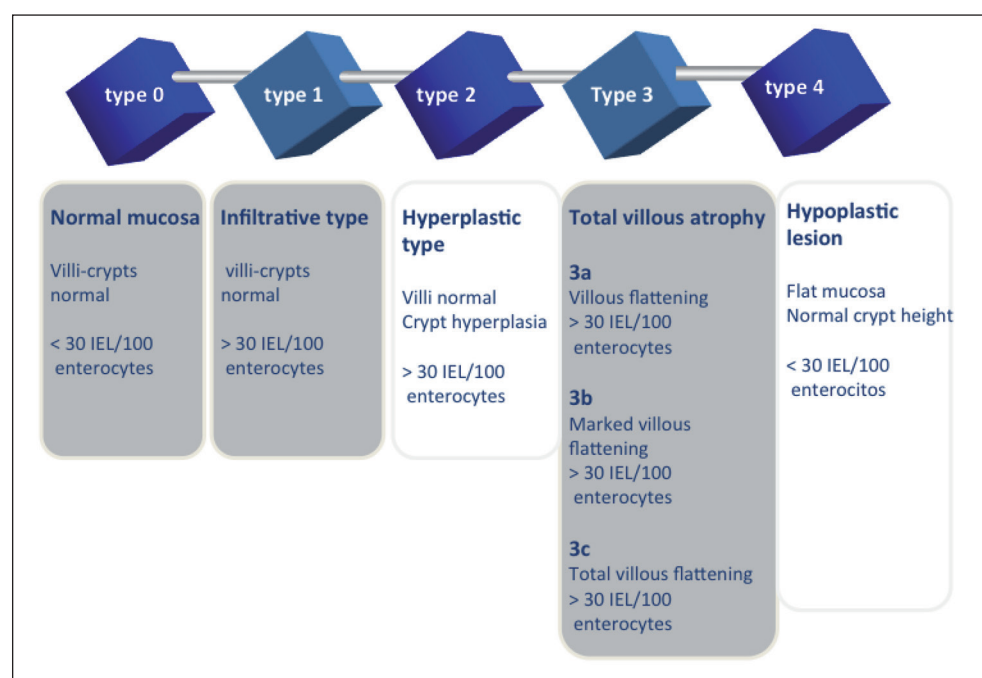


Figure 1. Marsh Oberhuber Classification in Celiac Disease. IEL: intraepithelial Lymphocytes.

The patient showed an excellent response to the diet, with a quick stabilization of stools, and relief from meteorism, thus the parents reintroduced lactose in his diet after 30 days. After 5 years of follow-up, the patient has not presented RP again, his growth has been normal and the Ab anti-tTG have been negative in the annual checks.

Case 2

A female patient, 1 year and 9 months old at the time of consultation, first daughter of non-blood relative and healthy parents, without relevant personal or familiar history. She received exclusive breastfeeding until 4 months of age, then she received infant formula. The mother added non-diary food at the 6th month

and gluten at the 7th month. The patient kept a growth curve within the normal range, according to the WHO references (weight/age < -1 standard deviation (SD) < -1 SD, weight/height normal) (18). This height was attributed to the low familiar height.

The patient was 20 months old when she presented RP twice, which were spontaneously reduced, this was the reason for consultation. During the interview, the mother reported the presence of pasty stools since the first administration of non-dairy food, which was not considered. The patient did not show obstructive bronchitis, pneumonia or otitis that could indicate cystic fibrosis as a possible cause of RP. During the physical examination, the patient was eutrophic, with mild abdominal meteorism, without other relevant findings. The anal and lumbosacral exams were normal.

The study focused on chronic diarrhea with high titers of anti-tTG Ab (Table 1), which led to an upper endoscopy with normal result. The duodenal biopsies showed an acute villous atrophy and an intraepithelial lymphocytes increase, which was compatible with enteropathy-type 3c in the Marsh-Oberhuber classification¹⁹ (Figure 1).

After the diagnosis of the celiac disease, a diet without gluten was indicated and the patient had an excellent response to the treatment. The stools stabilized and the meteorism disappeared. The patient did not present RP after the diagnosis, the growth curve changed from -1SD to normal and the weight increased to +1SD, thus no more studies were necessary. After 5 years of follow-up, where the patient had a strict diet without gluten, the patient has not presented RP again.

Discussion

In this report, two patients with an unusual form of manifestation of the celiac disease are presented. In both patients common causes of RP were dismissed (infectious diarrhea, constipation, respiratory disease, malnutrition, neurogenic bowel and ligament laxity)⁸. Despite the fact that the association between cystic fibrosis and RP is widely described^{10,11}, no diagnostic studies of cystic fibrosis by sweat test were performed, since there was no nutritional involvement or respiratory symptoms that supported this diagnosis.

Both patients had recurrent RP, which was the reason for consultation, and in both cases the chronic diarrhea was unnoticed. Both patients were diagnosed with celiac disease through a duodenal endoscopic biopsy, and then a diet without gluten was indicated. The RP did not recur once started the diet, which was the only therapeutic intervention in both patients. This

observation could suggest a possible causal association between the celiac disease and the RP.

There are only two reports that refer to this association, the first one in 1978¹⁶. It describes a 73-years old woman with chronic diarrhea, with nutritional involvement and RP. This case was studied in many occasions, but there was no duodenal biopsy, however, a gluten-free diet was indicated, which significantly relieved diarrhea, and also, the RP did not recur. The authors concluded that the RP was due to mechanical causes in the context of a non-treated celiac disease. However, in the absence of a duodenal biopsy, it is only possible to suppose that the patient had the celiac disease, and non-celiac intolerance to gluten cannot be dismissed as another possible cause.

The second report, in 2016¹⁷, describes the case of a child whose reason for consultation was the RP, associated with oily and pasty stools, who was also positive for celiac disease. Once started the gluten-free diet, there was a stabilization of stools and the RP did not recur.

Regarding the possible factors that could explain the RP in celiac patients, it could be considered that the physiopathology is similar to the one in cystic fibrosis. Because of the malabsorption, there would be an increase in the intestinal content and meteorism due to the excessive gas production. The patient must eat a large amount of food, probably more than the age requirements, in order to be eutrophic. The excessive intestinal volume and the increased gas production would contribute to a rise in the intra-abdominal pressure, and the presence of chronic diarrhea, increased push and straining would explain the increased risk of PR.

In both cases, the reason for consultation was the RP. In addition, both patients had a history of meteorism and irregular stools, which did not catch the attention of the parents and were not considered in previous medical evaluations, since both patients were eutrophic and looked healthy. The only symptom that made doctors suspect of the celiac disease was chronic diarrhea. Both patients had high titers of anti-tTG Ab, over 100 EU/ml. The duodenal biopsy showed some degree of atrophy and presence of intraepithelial lymphocytes, which were compatible with the classification 3c in the Marsh-Oberhuber scale. Based on these elements, in both cases, the celiac disease was objectified without any doubt in the diagnosis.

Conclusion

Despite the fact that the celiac disease is a common pathology, present in 1% of the general population²⁰, the RP would rarely be the main presentation symp-

tom of this pathology, however, performing an inverse analysis, if there is a case of RP in an infant without an apparent cause that explains it, such as chronic constipation, cystic fibrosis, acute infectious diarrhea, pelvic floor dysfunction or neurogenic pathology, it should be considered the celiac disease in the possible causes of RP.

The association between RP and the celiac disease reported in both patients of this report encourages to specifically look for the celiac disease in patients whose reason for consultation is the RP.

Ethical responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Financial Disclosure

Authors state that no economic support has been associated with the present study.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

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