Pneumatosis cystoides intestinalis: An unusual cause of distal constipation

La pneumatose kystique intestinale : une cause inhabituelle de constipation

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Available online 27 May 2010

Pneumatosis cystoides intestinalis is characterized by the presence of gas-filled cysts within the intestinal wall below either the mucosal or the serosal membrane. This rare and benign disease, first described in 1730 by Du Vernois [1], occurs predominantly between 40 and 50 years of age in both men and women [2]. Cysts are located along the entire gastrointestinal tract from the esophagus to the rectum [3] but are usually predominant in the small as well as the large bowel [2,4]. The pathophysiology of the disease remains unknown. The diagnosis is made during colonoscopy with the visualization of cysts appearing as round heightenings in the colonic wall membrane, although a laparotomy is the only way to diagnose cysts located below the serosa [5].

In most cases, cysts are totally asymptomatic. When symptoms are present, they are not specific, i.e., abdominal pain, intermittent rectal bleeding, diarrhea or constipation. We report a case where constipation was the predominant symptom. In this patient, the symptomatic evolution after cyst treatment suggested that cysts were in fact responsible for functional distal constipation.

Case report

A 57-year-old man consulted for a recent constipation. Previous medical history included pneumatosis cystoides intestinalis diagnosed in 1998 by colonoscopy which was decided due to intermittent rectal bleeding. Endoscopic examination revealed a disease which extended up to 35 cm from the rectum to the left colon. The patient primarily complained of stool frequency with only of one to two stools per week and very episodic sensation of urge to defe cate. Constipation was associated with intermittent rectal bleeding and thin tools. The only objective finding during clinical examination was the presence of stools in the rectal ampulla. This was confirmed on abdominal plain film X-ray. This worsened transit suggested a further colonoscopy that showed typical characteristics of pneumatosis cystoides intestinalis with cysts up to 40 cm from the anal margin but primarily involving the rectum. Although rectosigmoid mucosa bled easily, no stricure was observed (Fig. 1a). During this procedure, the largest cysts were flattened with a needle. This endoscopic treatment was followed by a transient improvement of constipation. Based on the clinical presentation of constipation which appeared to be distal, an anal manometry was performed. The examination revealed numerous spontaneous rectoanal inhibitory reflexes (RAIR) (Fig. 2), while no RAIR could be
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Figure 1  Endoscopic appearance of the rectosigmoid mucosa: a: before hyperbaric oxygen treatment; b: after hyperbaric oxygen treatment.

Figure 2  Rectoanal manometric pattern before hyperbaric oxygen treatment during a resting period.

triggered by the inflation of a rectal balloon (Fig. 3a). The patient was unaware of any rectal distension even when the distending volume was 50 mL (Table 1).

As the effects of the endoscopic cyst flattening were only transient, hyperbaric oxygen therapy was started (two sessions per day with 2.5 ATA during 60 mins) [6]. Constipation improved after the 10th session. Then transit became normal (one stool easy to pass daily) until the end of the 30 scheduled therapeutic sessions. Subsequently, a second anal manometry demonstrated normal findings: absence of spontaneous RAIR, onset of RAIR after each rectal balloon inflation from 10 to 50 mL, increase of both RAIR magnitude and duration when the inflating volume increased (Fig. 3b), perception of distension even for a distending volume of 10 mL. A second colonoscopy revealed the disappearance of practically all the cysts while no luminal stenosis was observed (Fig. 1b). Maintenance treatment was decided with 20 hyperbaric sessions per three months. Constipation did not reoccur during the 6-months follow-up.

Discussion

Our results suggest that the effect on the transit of cyst destruction, as sole treatment, demonstrates a clear link between the presence of the cysts in the rectal and sigmoid mucosa and outlet obstruction. This raises the question of the relationship between cysts and constipation.

The hypothesis of a mechanical luminal obstruction related to the presence of cysts was ruled out on the basis of
endoscopic findings excluding any luminal narrowing. In contrast, manometric findings suggested that cysts promoted a functional outlet obstruction.

Initially, when cysts were present, the patient had no urge to defecate and was unable to perceive rectal distension. It is likely that cysts prevented any increase of intrarectal pressure, particularly which followed the arrival of stools into the rectal ampulla. This increased pressure is considered essential for the activation ofafferent sensory pathways which inform the brain of stool issue into the rectum and trigger an urge to defecate. Via a similar mechanism, cysts could have promoted a reversible impairment of the intrinsic control of rectoanal function. Indeed, the absence of RAIR during rectal distensions, even after a distension of 50 mL (Fig. 3a), was one of the most significant manometric characteristics. In normal subjects, the passage of a stool is associated with transient relaxation of the internal anal sphincter permitting the anal canal sensors to determine rectal content. This rectoanal inhibitory reflex is normally detected by anal manometry immediately after balloon inflation (which reproduces the arrival of stools into the rectal ampulla). The pressure sensors normally detect a decrease of the upper anal canal pressure (corresponding to the transient relaxation of the internal anal sphincter). Absence or impairment of the RAIR have been reported in Hirschprung’s disease, a condition in which intrinsic nervous system is impaired. The lack of RAIR has also been reported in patients with diabetes mellitus [7] and recently with scleroderma [8]. Further investigation is required to ascertain if this manometric finding is common in pneumatosis cystoides intestinalis, since anal manometric findings to our knowledge have not yet been documented in this pathological condition.

The third manometric finding was spontaneous RAIR (Fig. 2). It is doubtful if spontaneous RAIR were in fact abnormal findings and whether or not they play a role in outlet obstruction. In our experience, we considered these spontaneous RAIR as a consequence of the stool stasis in the rectal ampulla. Indeed, this is a usual finding in patients referred to our center for the management of fecal impaction (unpublished data). This occurrence has also been reported in some neurological lesions such as myelomeningocele [9].

In conclusion, this case report suggests that in patients with pneumatosis cystoides intestinalis, cysts can promote a functional outlet obstruction, primarily by inhibiting rectal sensation and that distal constipation is completely reversible after hyperbaric oxygen cyst destruction.

Conflict of interest statement

No conflict of interest.

Acknowledgments

The authors are grateful to Richard Medeiros, Rouen University Hospital Medical Editor, for reviewing and editing the manuscript.

References


Table 1 Results of the anal manometry before and after hyperbaric treatment.

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<th>Before hyperbaric treatment</th>
<th>After hyperbaric treatment</th>
<th>Normal values</th>
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<tr>
<td><strong>Resting pressures</strong></td>
<td></td>
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<tr>
<td>Upper anal canal pressure (cm H2O)</td>
<td>84</td>
<td>112</td>
<td>32–105</td>
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<tr>
<td>Lower anal canal pressure (cm H2O)</td>
<td>27</td>
<td>59</td>
<td>10–82</td>
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<td><strong>RAIR</strong></td>
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<td>Spontaneous</td>
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<td>Triggered by balloon inflation</td>
<td>Presence</td>
<td>Absence</td>
<td></td>
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<td>Volume of first appearance of rectal distension (mL)</td>
<td>&gt; 50</td>
<td>10</td>
<td>Yes</td>
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<td></td>
<td>Rectoanal inhibitory reflexes (RAIR).</td>
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