Colitis cystica profunda is a rare pathology with approximately 200 cases reported in the literature. This entity is characterized by a slow, asymptomatic evolution and when finally recognized, confusion with a malignant tumor with subsequent application of unnecessary therapies or surgical resections is frequent. We present the case of a 41-year-old male with 7-day evolution ultimately presenting to the emergency room with abdominal pain. At physical examination, he was dehydrated with severe abdominal pain, tachycardia and tachypnea, abdominal distention, without bowel sounds, “wood abdomen” with clearly acute abdomen signs, and anuria for 48 h. The patient required mechanical ventilation secondary to abdominal restriction with severe metabolic acidosis. The situation of hemoglobin (14.5 mg/dL), hematocrit (38), leukocytes (18,500/mm$^3$ with 89% neutrophils), platelets (257,000), creatinine (2.1 mg/dL), glucose (230 mg/dL), and normal clotting times were included in the laboratory reports. The diagnosis was intestinal obstruction, abdominal compartment syndrome, and septic and hypovolemic shock. At exploratory laparotomy, we found a sigmoidal tumor totally occluding the lumen with severe dilatation of the proximal colon and haustral tearing with ischemia. Totally, 2 m of ileum showed necrotic patches secondary to compartment syndrome and compression to the abdominal wall. A total colectomy and resection of 2 m of ileum with ileostomy and Hartmann’s procedure were completed. The pathology examination reported colitis cystica profunda without atypia. After 3 weeks and stoma remodeling, the patient was discharged. This benign lesion could be treated with less aggressive procedures. With the correct pathological diagnosis, a better treatment with excellent prognosis could have resulted.

**Key words:** Colon adenocarcinoma, compartmental syndrome, deep cystic colitis, intestinal obstruction
The patient had received 2 previous medical examinations and laxatives as treatment with no relief. The medical history included diabetes mellitus type 2 treated with metformin. At physical examination, he was dehydrated, referring severe abdominal pain, tachycardia and tachypnea, with abdominal distention without bowel sounds, “wood abdomen” with clearly acute abdomen signs, and anuria for last 48 h. Patient required mechanical ventilation secondary to abdominal restriction to breathing with metabolic acidosis with pH = 7.0, 9.5 mmol/L HCO$_3^-$ and 17 mmol/L BECF, 18 mg/m$^3$ CO$_2$, 80% SO$_2$. The situation of hemoglobin (14.5 mg/dL), hematocrit (38), leukocytes (18,500/mm$^3$ with 89% neutrophils), platelets (257,000), creatinine (2.1 mg/dL), glucose (230 mg/dL), and normal clotting times were included in the laboratory reports. The diagnosis was intestinal obstruction, abdominal compartment syndrome, and septic and hypovolemic shock. At exploratory laparotomy, we found a sigmoidal tumor totally occluding the lumen [Figure 1], with severe dilatation of proximal colon and haustral tearing with ischemia. Totally, 2 m of ileum showed necrotic patches secondary to compartment syndrome and compression to the abdominal wall [Figure 2]. A total colectomy and resection of 2 m of ileum with ileostomy and Hartmann’s procedure were completed.

The pathology examination reported colitis cystica profunda with adipose degeneration and degeneration of muscle fibers in the wall without atypia. Benign and cystic glandular structures penetrating the muscular layer were present, showing inflammatory elements and mucin [Figures 3 and 4]. The patient presented high ileostomy discharge for 2 weeks, and an ileostomy remodeling was needed, secondary to enterocutaneous fistula, and initial necrosis of the stoma secondary to the septic and hypovolemic shock. After 3 weeks, the patient was discharged.

**Discussion**

Colitis cystica profunda is a rare disease, and acute presentation is unusual. The pathogenesis is associated with bowel inflammatory diseases such as ulcerative colitis, Crohn’s disease, rectal prolapse, trauma, ischemia, or radiation. The lesions are usually located in rectum, and the average length range is 5–12 cm from the anterior aspect of the rectum.$^1$ The disease is classified into 3 different forms by Herman and Nabseth according to the distribution localized, segmentary, and diffuse. At the macroscopic level, it appears as ulcerative in 57%, polypoid in 25%, and flat type in 18%.$^2$ $^4$ Microscopically, it demonstrates obliteration of the lamina propria by fibroblasts with disorientation of muscle fibers, regenerative changes in the crypt epithelium, and displacement of mucosal glands into the submucosa, which can cause retention of mucus, resulting in cystic formation. Almost 25% of 3 patients, which do not contain fibromuscosulosis in the lamina propria at rectal biopsy, could be mistakenly diagnosed as adenocarcinoma.$^3$

The symptomatology is nonspecific such as mucorrhea, abdominal pain, rectal bleeding, tenesmus, and constipation. The differential diagnosis includes inflammatory bowel disease, benign or malignant tumors, and infectious or pharmacological colitis. Imaging studies can help to elucidate
a more specific diagnosis. Ultrasound shows multiple hypoechoic cysts without rectal submucosa interruption and confirms lymph nodes absence or muscular layer invasion. A computed tomography (CT) scan could show a noninfiltrative submucosal lesion without perirectal fat tissue and with anus elevator muscle thickening. An MRI could demonstrate nodules with strong signals on T2 due to the mucoprotein content of the cyst.2,5

The histopathological study demonstrates an increased thickness of the submucosa layer that can stretch from the muscularis mucosa to the serosa. It is characterized by the formation of submucosa intramural cysts containing a thick and gelatinous material with fibromuscular obliteration. The cells are usually well differentiated, with edematous rectal mucosa and hypertrophy or mild glandular atrophy. The Lieberkuhn crypts may be elongated, distorted, or irregular with a chronic inflammatory infiltrate that may be associated with hyperplasia, hypertrophy, and disorganization of the muscle fibers and nuclear atypia stockings.2,5,6

According to the macroscopic, microscopic, and unspecific symptomatology, the definitive diagnosis is difficult to achieve, and sometimes, this entity is misdiagnosed as adenocarcinoma or another malignant pathologies resulting in more aggressive interventions such as surgical resection, with resulting adverse effects on both the patient and the family.

Conservative treatment includes topical corticoids for small lesions with minimal symptoms and an increase in dietary fiber, as correction of constipation has a positive effect, with favorable results in 34%. In the cases of larger lesions that protrude, present bleeding or as in this case, produce intestinal obstruction, a surgical resection will be needed. Prognosis after surgical resection is excellent, and there exist only one report in the literature of recurrence 20 years after resection.5,7

The few reported cases of colitis cystica profunda in the literature could result in misdiagnosis and an extensive resection and oncologic approach to a benign lesion that could be treated with less aggressive procedures, especially if it presented acutely with profuse bleeding or intestinal obstruction like the presented case. This pathology could be suspected by image studies such as CT scan if the patients are stable, but if it is not the case, the only way is the pathologic examination. The correct pathologic diagnosis allows us to provide better treatment and give the patient and family an excellent prognosis.

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Conflicts of interest
There are no conflicts of interest.

References