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Case Report

Colitis cystica profunda of the rectum with adenomatous dysplastic features: Radiologic-pathologic correlation

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ABSTRACT

Colitis cystica profunda is a rare nonneoplastic condition characterized by the presence of mucus-containing cysts in the submucosa of the right colon and rectum. The etiology is unclear, with a few cases reported in the literature. The presenting symptoms and signs may mimic colorectal adenocarcinoma. We report a case of colitis cystica profunda localized in the rectum, investigated by colonoscopy, CT, MRI, and subsequently surgically treated.

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Introduction

Colitis cystica profunda (CCP) is an uncommon benign condition of the colon and rectum, with a few cases reported in

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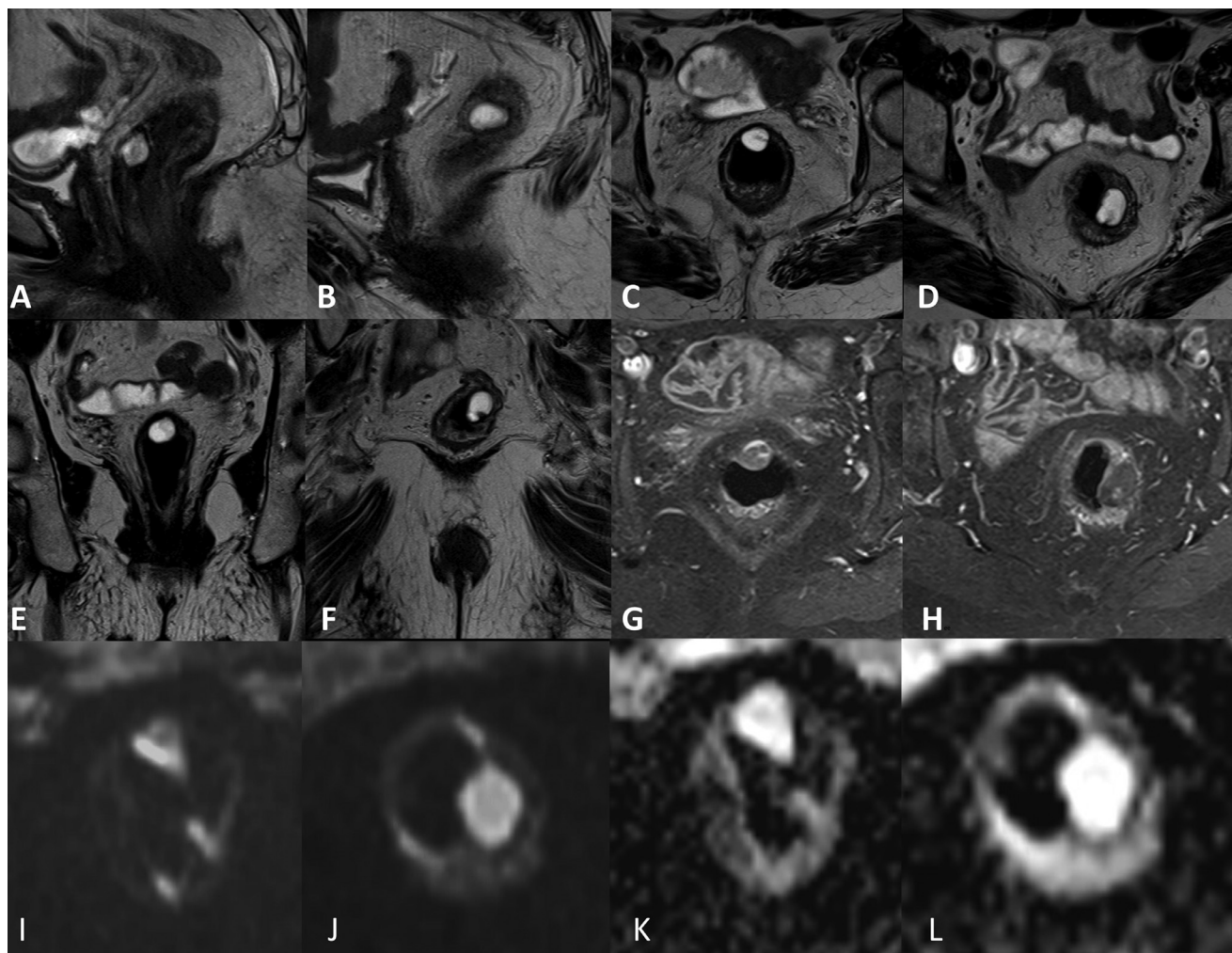


Fig. 1 – (A-F): T2-weighted MRI showing the presence of 2 cystic lesions of the rectal wall with an epicenter in the submucosa (A and B: sagittal plane; C and D: axial plane; E and F: coronal plane). (G and H): T1-weighted fat suppressed MRI in axial plane obtained after administration of contrast medium showing only peripheral enhancement of rectal cysts. (I and J): Diffusion-weighted imaging (DWI) MRI and (K and L): Apparent diffusion coefficient (ADC) in axial plane do not reveal diffusion restriction.

the medical literature [1]. CCP is often associated with solitary rectal ulcer and rectal mucosal prolapse syndromes [2]. CCP usually occurs in the third and fourth decades of life, but may occur also in pediatric age [3]. The main occurrence of CCP (70% of cases) is in the rectum while only 14.6% of cases are localized in the right colon, and diffusely large intestine involvement is also possible in rare cases [4].

Macroscopically, the lesion shows as an ulcerative aspect in 57%, polypoid in 25%, and flat type in 18% of cases [2], presenting with mucin filled cysts located deep in the submucosa [5]. Patients may be asymptomatic or they may present with rectal bleeding, mucorrea, tenesmus, mild anorectal pain, diarrhea, and constipation [5]. From a radiological point of view, this rare disease is relevant because its clinical and morphological features can mimic malignant lesions of the colorectum [6].

Case report

A 65-year-old woman, with no significant medical history and no family history of colorectal diseases, presented to our hospital for recurrent fresh rectal bleeding associated with mucus discharge and tenesmus. Endoscopic examination revealed 2 polypoid mass of the rectum; histology of the mass biopsies revealed low-grade dysplasia of the mucosa on the major cyst.

The patient underwent computed tomography (CT) and magnetic resonance (MRI) examinations that revealed 2 cystic lesions of the rectal wall (Figs. 1-3), suspected for colitis cystic profunda of the rectal wall.

On the basis of the biopsy histological findings, and considering the long period of the symptoms, the patient was treated surgically by anterior resection of the rectum. The resected

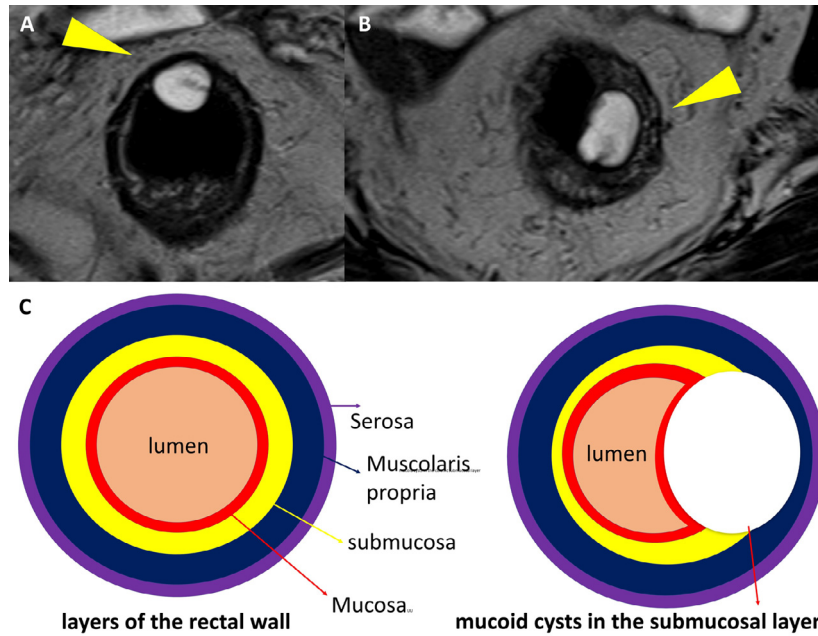


Fig. 2 – T2-weighted MRI in axial plane (A and B) showing 2 cystic lesions in the submucosa of the rectal wall. The schematic representation of the rectal wall layers demonstrates the exact location of the cysts containing mucin (C).



Fig. 3 – Axial computerized tomography in the venous phase revealing the fluid content of rectal lesions (yellow arrowhead), the peripheral calcifications (A) and the peripheral enhancement. (Color version of figure is available online.)

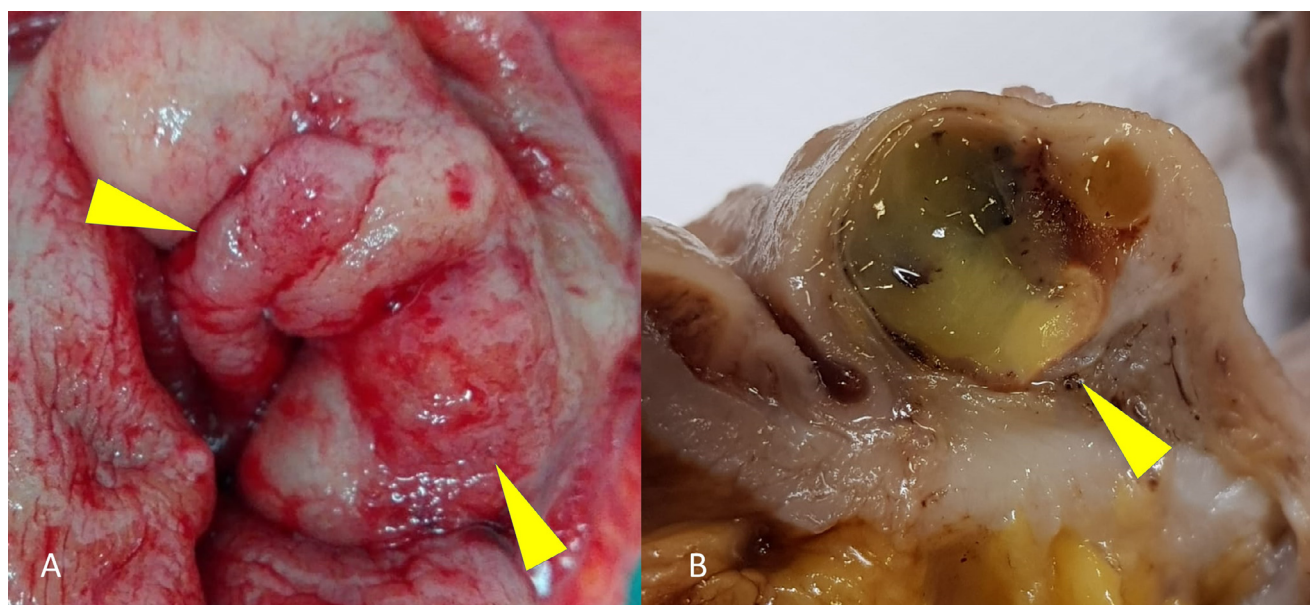


Fig. 4 – The resected bowel specimen revealing 2 polypoid submucosal lesions (yellow arrowheads in A). Gross appearance of the opened lesion shows the mucin-filled cyst (yellow arrowhead in B). (Color version of figure is available online.)

bowel specimen revealed 2 polypoid submucosal lesions and the gross appearance of the opened lesion showed the mucin-filled cyst (Fig. 4).

Histological examination confirmed the submucosal mucous cysts and the typical features of colitis cystica profunda (Fig. 5), also highlighting a focus of low-grade tubular adenoma in the mucosal layer of the major cyst. Cysts contained inspissated mucin and they were lined by a large bowel-type benign mucinous epithelium with surrounding fibrosis and presence of hemosiderin macrophages. In addition, calcifications were peripherally present to the cysts.

The postoperative course was regular with complete resolution of the symptomatology at the follow-up.

Discussion

CCP is a rare nonneoplastic condition well described in the literature since XVIII century [7,8]. It was classified in 3 forms based on its distribution: diffuse, segmental, and localized. In the diffuse form, frequently associated with inflammatory bowel disease, cystic lesions are observed throughout the colon and macroscopically appears as pedunculated or villous polypoid lesions, even ulcerated. The segmental form is characterized by the presence of lesions, predominantly polypoid, in one or more sections of the colon, typically in the rectosigmoid tract. In the localized form the lesion, polypoid with or without ulceration, is situated along the anterior wall of the rectum, usually 5-12 cm from the anal orifice [4,7].

The clinical presentation is variable, depending on the location, the number, and the macroscopic appearance of the lesions. The predominant symptoms are fresh rectal bleeding, mucus discharge, perineal or abdominal pain, diarrhea, tenesmus, and intestinal obstruction. At digital rectal examination,

this condition could appear as a rectal mass or an ulcerated lesion, associated in some cases with rectal prolapse [2,7]. As these signs and symptoms are all compatible with colorectal adenocarcinoma, the endoscopic examination with biopsy is mandatory. Endoscopic findings are not specific and revealed a single or multiple nodular or polypoid mucosal thickening or a mass lesion with or without ulceration [2]. The mucosa spread over the surface of the cysts may show irregularly distributed areas of edema, hyperemia, hypertrophy, and atrophy with occasional superficial ulceration or central umbilication [7].

In the early stage, barium enema studies may be negative or they may demonstrate narrowing and irregularity of the lumen due to the submucosal cystic lesion [2].

Transrectal ultrasonography can help to diagnosis. Hulsmans et al [9] defined typical features of CCP like the presence of multiple hypoechoic and anechoic cysts in the rectal submucosa, uninvolving the muscularis mucosae without associated lymphadenopathies. These features allow discriminating CCP from anorectal malignancy [10].

CT imaging shows noninfiltrating submucosal masses with loss of perirectal layers of fatty tissue and asymmetry and thickening of the levator ani muscle, without the presence of local or regional adenopathies [10]. On MRI the lesions appear as submucosal, homogenous, and variable hyperintense nodules on T2-weighted images, not involving the muscle layer. T1-weighted MRI sequence shows thickening of the levator ani muscle, asymmetry of the rectal lumen, without infiltrative signs, and confirms the absence of lymphadenopathies. On contrast-enhanced images, the lesion may have peripheral enhancement [10].

In the present paper, CT and MRI examinations highlighted only the cystic aspect of the CCP and its typical location in the submucosal layer. However, the thickening of the levator ani muscle was not present, unlike the literature suggestions.

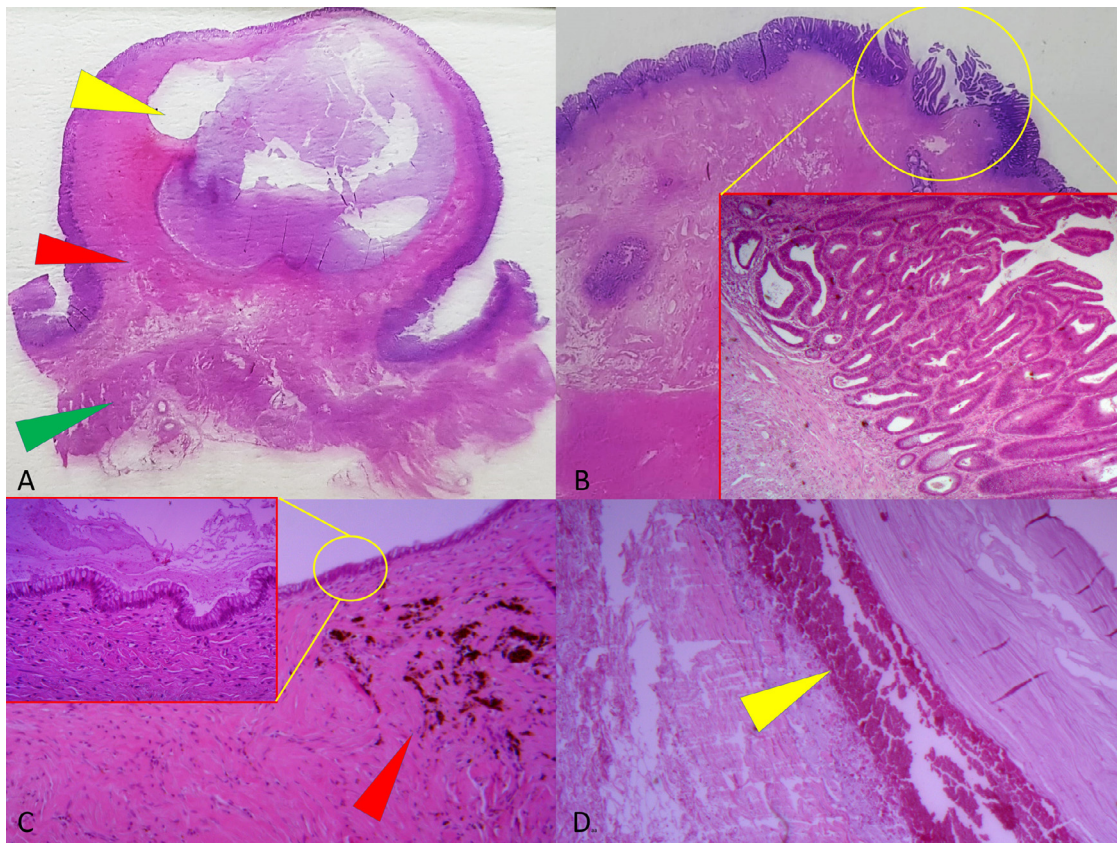


Fig. 5 – Cyst (A: hematoxylin and eosin X 2) containing inspissated mucin (yellow arrowhead) with an epicenter in the submucosa (red arrowhead); muscularis propria (green arrowhead). Detail of low-grade tubular adenoma in the mucosal layer (B: hematoxylin and eosin X2 and X10). Cyst (C: hematoxylin and eosin X2 and X10) lined by large bowel type benign mucinous epithelium with surrounding fibrosis and presence of hemosiderin macrophages (red arrowhead). Calcifications (D hematoxylin and eosin X10) are present peripherally to the cysts (yellow arrowhead). (Color version of figure is available online.)

Moreover, our case describes also the presence of parietal peripheral calcifications of cysts. This last observation is of great interest because it suggests a chronic benign condition, not associated with rectal cancer, in which calcifications are usually intratumoral calcification [11].

Although transrectal US, CT, and MRI can support a benign nature of the lesion, only adequate biopsy affords a definitive diagnosis. Typical histological findings of CCP are the presence of a submucosal cyst, delimited from mucosal, or epithelial cells without malignant atypia. The presence of lymphocytes, neutrophils, macrophages, and fibroblasts infiltrates together with smooth-muscle cells of the lamina propria is common [4]. Furthermore, the immunohistochemical negativity for p53 and the absence of cryptitis or crypt abscesses may confirm the diagnosis [2]. The association of CCP and adenocarcinoma presented in few cases [5].

The differential diagnosis of CCP includes any rectal or colonic polypoid or intramural mass (adenomatous polyps, lipoma, leiomyoma, sarcoma, and polypoid inflammatory granulomas), inflammatory bowel disease (ulcerative colitis, Crohn's disease, and ischemic proctitis or colitis), and endometriosis [2,7].

The etiology of CCP remains controversial. The association with Peutz-Jeghers syndrome and chronic inflammatory diseases, together to an iatrogenic origin, was reported [4]. The suggested pathogenetic mechanism is ischemia [4,5]. Treatment should be medical or surgical according to the severity of symptoms. In most cases, patient education with a diet high in fiber and pharmacological therapy such as glucocorticoids and sulfasalazine may reduce symptoms (70%-75%) [2].

Operative approach is necessary when symptoms are persistent, as in the present case, or severe such as intestinal obstruction or rectal prolapse [5]. The prognosis in patients with complete excisions is excellent, with a free recurrence rate of 80% [12].

This case described the typical features of the colitis cystic profunda on CT and MRI examinations. The CT and MRI findings allowed an accurate diagnosis, able to exclude a neoplastic pathology of the rectum. This case also described the association of the CCP with a low-grade tubular adenoma in the mucosal layer of the major cyst. This association constitutes a very rare event, and at the best of our knowledge it was reported only in an old case report [13].

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