Colitis Cystica Profunda with Gastrointestinal Stromal Tumor: A Case Report

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Case report

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Abstract

**Background:** Colitis Cystica Profunda (CCP) is a rare benign medical condition and its exact etiology is still unknown. The clinical symptoms could be atypical and even asymptomatic, and also associated with several diseases. Most CCP patients may be misdiagnosed due to its complexity, and can be recognized after long-term medical treatment or surgical resection, especially in the elder or patients with relevant family history.

**Case presentation:** In this case, a 58-year-old man presented with change in bowel habits for 3 years. Gastroscopy and colonoscopy were performed, and the lesions in stomach were resected. Colonoscopy revealed most lesions were significantly smaller after the resection of stomach neoplasm. Notably, these lesions recurred 3 years later.

**Conclusion:** This case is an important reference for primary physician and pathologist to make correct diagnosis through initial endoscopic and pathological examination.

**Background**

Colitis Cystica Profunda (CCP) is a rare benign medical condition and its exact etiology is still unknown. The clinical symptoms could be atypical and even asymptomatic, and also associated with several diseases, such as ulcerative colitis (UC), inflammatory bowel disease (IBD), radioactive enteritis, infectious colitis, colorectal polypoid, and even colonic adenocarcinoma or Gastrointestinal Stromal Tumor (GIST). Most CCP patients may be misdiagnosed due to its complexity, and can be recognized after long-term medical treatment or surgical resection, especially in the elder or patients with relevant family history. We firstly reported a patient with CCP and GIST concurrently. In this case, the lesions in the stomach and intestines simultaneously, but the natures of the lesions were completely different.

**Case Presentation**

A 58-year-old man presented with change in bowel habits for 3 years. He had history of asymptomatic gallstones. Family history of cancer, gallbladder cancer for his father and ascending colon cancer for his brother was noted. Colonoscopy revealed multiple broad-based hemisphere and lobular nodules (range from 1 cm to 3 cm) scattered in the ascending, the hepatic flexure and the transverse of colon (Fig. 1A). The endoscopic ultrasound (EUS) showed clear boundary nodules located in the intrinsic muscular layer (Fig. 1B). Additionally, the gastroscopy also showed a single smooth mucosa nodule (0.5 cm) existed in pylorus (Fig. 2A), and EUS displayed the lesion situated in intrinsic muscular with clear boundary (Fig. 2B). Then, endoscopic gastric submucosal mass resection and colonic submucosal mass biopsy was performed. The postoperative pathology analysis identified pylorus nodule as GIST (low-grade malignancy) (Fig. 2C).

However, the lesions of colon were polyp in the submucosa with fibrous tissue hyperplasia accompanied by inflammatory cells infiltration. Many inflammatory cells were enriched in the focal small vessels.
Partial region showed loose edema accompanied by cyst. Immunohistochemically, cells from colon polyp displayed partial positivity for CD34 and smooth muscle actin. CD117 S100, and Dog-1 were negative. The Ki67 proliferative index was about 2%. The diagnosis of inflammatory fibroid polyps was uncertain, so colonoscopy was performed again and revealed most lesions were significantly smaller than before (Fig. 1D). Taking all imaging and pathology analysis together, the nodules were proved to be CCP. Specially, mucin pool existed in submucosa and mucous membrane was polypoid and protruded into lumen. The dilated mucin pool was surrounded by chronic inflammatory cells and fibrous tissue. The inner membrane was filled with hyperplasia fibrous tissues and more thickened smooth muscle fibers (Fig. 1C). The patient underwent colonoscopy at 3 months after lesion resection. Most lesions disappeared with CCP scars left (Fig. 1E). Notably, these lesions recurred 3 years later (Fig. 1F).

**Discussion And Conclusions**

CCP is a rare benign medical condition and its exact etiology is still unknown. It may occur at either mucosa or occasionally submucosa containing mucous cysts focally or diffusely\[^1\]. CCP can be divided into diffuse, segmental and localized\[^2\]. The diffuse CCP often locates in the whole colorectal, segmental CCP states single or several segments of colorectal and localized CCP occurs in the rectum distant from anal verge 5-12cm\[^2\]. In this case, the lesions located at ascending colon, hepatic flexure of the colon and transverse colon, which should be classified as segmental type.

The CCP patients may present with abdominal pain, hematochezia, change in bowel habits, diarrhea, mucous fluid and tenesmus\[^3\]. The clinical symptoms could be atypical and even asymptomatic. What’s more, CCP was associated with several diseases, such as UC, IBD, radioactive enteritis, infectious colitis, colorectal ploypoid, and even colonic adenocarcinoma or GIST\[^4, 5\]. The etiology of CCP is still unknown and may be related to congenital, inflammatory reaction or trauma\[^6, 7\]. The isolated colonic ulcer in colonoscopy may be presented as polypoid mass slightly protruding from the enteric cavity, which is similar to intestinal cancer but rarely induces obstruction\[^1\]. Mucous adenocarcinoma and mucous cyst can be distinguished by pathological examination. In this case, pathological features are cystic dilatation of ectopic glands and mucin pool in submucosa, which makes mucosa present as a polypoid which irruptive into antrum. In addition, enlarged glands or mucus are often surrounded by chronic inflammatory reactions or fibrosis hyperplasia\[^8\].

Most CCP patients may be misdiagnosed due to its complexity, and can be recognized after long-term medical treatment or surgical resection, especially in the elder or patients with relevant family history. In this case, the process of diagnosis and treatment was complicated. There was no ulceration but only polyp lesion which protruded into antrum in the colonoscopy. The diagnosis was uncertain until biopsy. To a large extent, it relied on the fact that lesions were significantly shrink when reexamined. Notably, in this case, the etiology, treatment and recurrence are still not clear. It may work via following ways: Firstly, self-healing. Secondly, use of antibiotics. Thirdly, the remaining lesions were atrophied after a single resection. The hypothesis may improve the diagnosis of colon lesions. The mode of multi-disciplinary
team also plays an important role for the diagnosis and treatment. It is effective to integrate the advantages of various disciplines.

In summary, the endoscopic and clinical features of CCP and GIST are presented. This case is an important reference for primary physician and pathologist to make correct diagnosis through initial endoscopic and pathological examination.

**Abbreviations**

CCP: Colitis Cystica Profunda

GIST: Gastrointestinal Stromal Tumor

UC: Ulcerative Colitis

IBD: Inflammatory Bowel Disease

EUS: Endoscopic Ultrasound

**Declarations**

**Ethics approval and consent to participate**

All procedures performed in this case followed the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This case was approved by Shanxi Cancer Hospital (approval ID:201980). This study does not contain any animal experiments.

**Consent for publication**

Informed consent was obtained from the patient.

**Availability of data and materials**

All relevant data regarding the case report are displayed in the publication. Raw data used in this case report are not publicly available because this include some indirect identifying information (age, sex, case history, the expression of Immunohistochemistry and so on), but are available from the corresponding author on reasonable request.

**Competing interests**
The authors have no competing interests to declare.

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**Authors' contributions**

Dr. Ren had substantial contributions to the conception of the work, acquisition of data, analysis and interpretation of data and drafting the article. Dr. Zhang and Dr. Jiang contributed to conception of the work, analysis and interpretation of data and revised the draft critically for important intellectual content. All authors read and approved the final manuscript.

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Figure 1

Colonscopy and pathology. (A) First colonscopy: multiple broad-based hemisphere and lobular nodules (range from 1cm to 3cm) with smooth surfaces and pouch or solid contact. (B) EUS: The nodules were located in the intrinsic muscular layer, with well demarcation and hypoechoic. (C) Pathology: Immunohistochemistry displayed Dog1 was positive (×100) (D) second colonscopy: most of the lesions were significantly smaller than before. (E) Third colonscopy: most of the mass disappeared. (F) The latest colonscopy: the similar nodules recurred.
Figure 2

Gastroscopy and pathology. (A) Gastroscopy: A single smooth mucosa nodule (0.5cm) in pylorus. (B) EUS: The lesion was situated in intrinsic muscular with well demarcation and hypoechoic. (C) Pathology: There was no epithelium on the swollen gland. The dilated mucin pool was surrounded by chronic inflammatory reaction and fibrous tissue hyperplasia.