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AN INTIMATE RELATIONSHIP LIES BETWEEN THE APPENDIX AND THE COLON: A CASE REPORT OF COLITIS CYSTICA PROFUNDA POST-LAPAROSCOPIC APPENDECTOMY

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ABSTRACT

Cystica profunda is a rare benign finding of mucous-filled cysts in the submucosa of the gastrointestinal tract, more commonly found in the rectum and colon. Risk factors include rectal prolapse, inflammatory bowel diseases, pelvic radiation and being post-appendectomy. We present a case of a female patient presenting with rectorrhagia, found to have sigmoidal colitis cystica profunda (CCP) six months post-appendectomy. This case is one of the few in medical literature to highlight the direct association between laparoscopic appendectomy and CCP, previously discussed in the literature as a complication post-appendectomy in the American Journal of Gastroenterology.

KEYWORDS

Colitis cystica profunda, post-appendectomy, rectorrhagia

LEARNING POINTS

- Physicians should have a high index of suspicion to rule out colitis cystica profunda (CCP) when approaching a patient with rectorrhagia following laparoscopic appendectomy.
- It is pivotal to make a prompt diagnosis for CCP in the context of rectorrhagia and initiate timely management.
- It is important to differentiate CCP from other aetiologies of lower gastrointestinal tract bleed as it is coined 'the great imitator'.

INTRODUCTION

Cystica profunda is a rare benign finding of mucous-filled cysts in the submucosa of the gastrointestinal tract, more commonly found in the rectum and colon. When these cysts are found in the colon, they are termed colitis cystica profunda (CCP). Jiang et al. report that the term CCP was first coined in 1957 by Goodall and Sinclair to describe these findings[1]. Symptoms of CCP include abdominal pain, distention, haematochezia and rectorrhagia, mimicking an underlying malignancy[2]. Risk factors include rectal prolapse,

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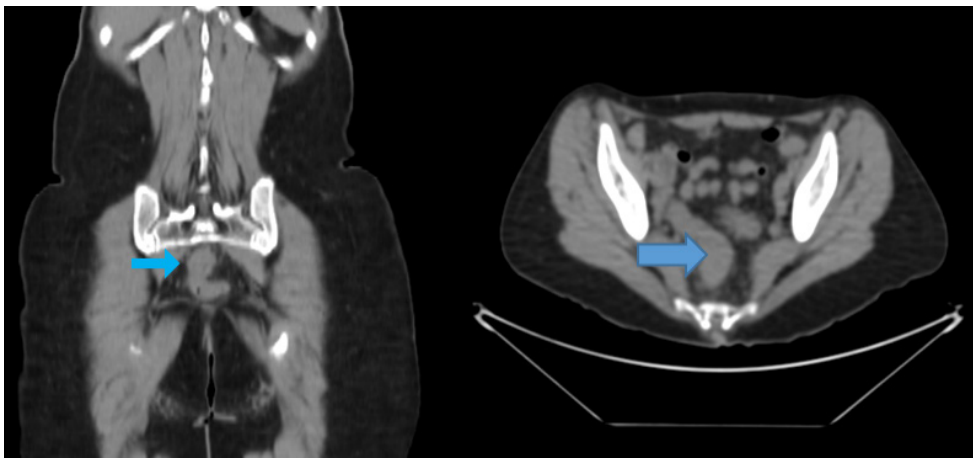

Figure 1. Coronal and axial reformatted images of an unenhanced CT scan of the abdomen and pelvis showing mild circumferential thickening of the wall of the sigmoid.

inflammatory bowel diseases, pelvic radiation and being post-appendectomy[3]. We present a case of a female patient presenting with rectorrhagia, found to have sigmoidal CCP six months post-appendectomy.

CASE DESCRIPTION

A 29-year-old previously healthy female presented to the emergency department with a three-day history of worsening lower left abdominal pain and rectal bleeding of one month's duration. A computed tomography (CT) scan with contrast revealed thickening in the sigmoid colon *(Fig. 1)*. She was admitted to the hospital and treated with intravenous antibiotics with no improvement. A colonoscopy was performed during her hospital stay, revealing a 1 cm polypoid mass with an ulcerated and inflamed surface in the sigmoid colon *(Fig. 2)*. This mass was removed using cold snare polypectomy with haemoclip application *(Fig. 3)*. On histology, the surgical specimen showed mucin-filled cysts indicating CCP located in the submucosa of the sigmoid colon, with no evidence of malignancy. Upon detailed anamnesis, the patient reported undergoing a laparoscopic appendectomy six months prior to presentation. Previous CT scan results at the time of appendectomy were unremarkable, with absence of sigmoid wall thickening.

DISCUSSION

CCP is an uncommon and benign condition characterised by multiple submucosal cysts filled with mucus, most commonly located in the rectum and sigmoid colon. The cysts are of variable morphologies and sizes, lined with a single layer of columnar epithelial cells filled with mucus. These findings can be described as a mucin-filled cyst surrounded by atrophic colonic mucosa and colonic mucinous epithelium[1,4].

This condition is often misdiagnosed due to the lack of specificity in endoscopic, radiological and histological examinations, resulting in an unnecessary surgical resection[1]. The presentation of CCP can mimic that of colonic malignancies, as stated in many articles, and should therefore be considered in the differential diagnosis of gastrointestinal masses. It is thus prudent to differentiate this 'rare mimicker of colorectal neoplasia', as termed by Zaki et al., from mucus-producing adenocarcinomas, although this is difficult sometimes[5-7]. This difficulty arises from the similar presentations between CCP and benign or malignant colorectal tumours, and inflammatory bowel disease. Common symptoms include mucorrhoea, rectal bleeding, diarrhoea and colonic obstruction. Histologically, the lesions in CCP are characterised by submucosal colonic glands, which may be mistaken for carcinomatous spread.

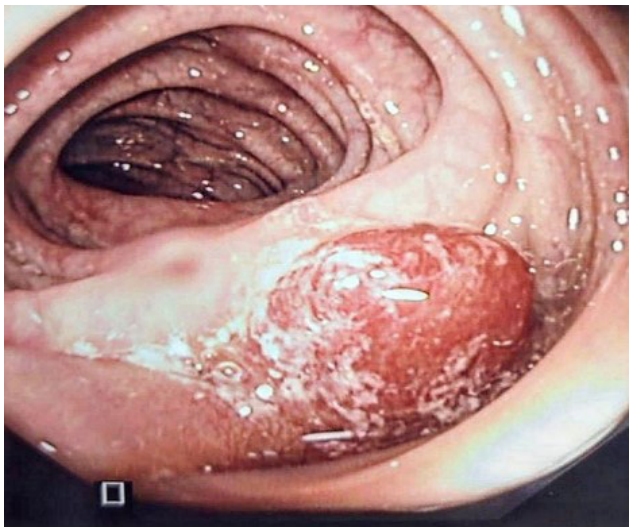

Figure 2. Snapshot from colonoscopy showing a 1 cm polypoid mass with an ulcerated and inflamed surface in the sigmoid colon.

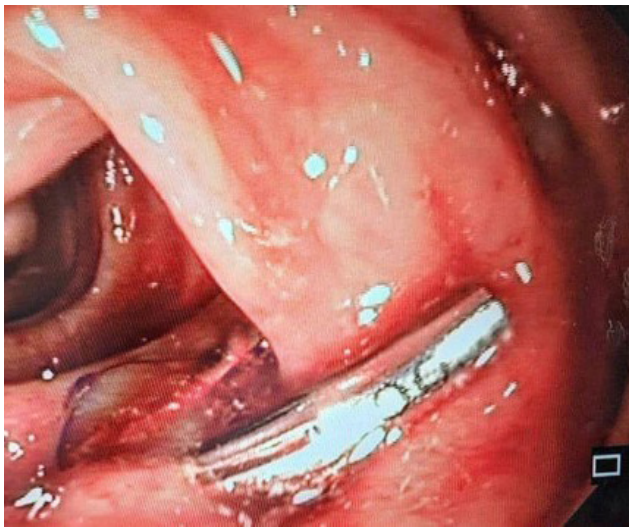

Figure 3. Removal of the mass using cold snare polypectomy with hemoclip application.

CCP may also occur in association with a gastrointestinal adenocarcinoma, as reported by Mitsunaga et al., where CCP lesions were found adjacent to colorectal adenocarcinoma as shown on histology. Mitsunaga et al. also reported that p53 mutations and high Ki-67 expression in adenocarcinomas helped differentiate them from CCP with benign epithelial cells lining the cysts[8]. The aetiology of CCP is not completely understood, but congenital or acquired weakness of the mucosal wall is thought to be involved in the pathogenesis of the condition. This weakness can be triggered by infection, inflammation, ischaemia or trauma, embedding the mucosal epithelium within the submucosa[1]. CCP is more common in middle-aged men in the third and fourth decades of life[1]. Clinically, patients present with non-specific signs and symptoms of diarrhoea or constipation, abdominal pain, blood or mucous in stool, and rectal pain[9]. The condition is classified as diffuse or local based on the degree of cystic invasion[9]. In the diffuse type, which accounts for less than 15% of cases, cysts span the entire colon due to diffuse intestinal inflammation and ulceration. It is associated with Crohn's disease, infectious colitis, ulcerative colitis and radiation enteritis[1]. The local type, which presents as nodules or polyps in the anterior wall of the rectum, is more common and is associated with rectal prolapse and isolated rectal ulcer syndrome[1].

CCP shows characteristic findings on imaging. X-rays can be normal in the early stages or can show irregularity and luminal narrowing caused by the multiple cysts in the submucosa[1]. On barium enema, single or multiple filling defects or thickened intestinal folds can be present[1]. On CT imaging, CCP presents as a non-infiltrating mass in the submucosa with borders that are well-defined and a cystic lumen that is variable in size[1]. Villous or tipped polypoid lesions appear on colonoscopy covered by normal, ulcerated or oedematous mucosa.

When nodular or polypoid mucosal thickening is present on colonoscopy, it can be misdiagnosed as colorectal cancer due to similar morphology[1]. Colonoscopy offers the advantage of biopsy, but misdiagnosis can still occur in cases of limited biopsy depth and not enough sampling[1]. When treating CCP, the aim is to relieve symptoms depending on how severe they are[1]. Conservative treatment with a high-fibre diet and lifestyle modifications is considered as a first-line treatment to avoid constipation and straining during defecation that precipitate mucosal defects. However, conservative treatment is often ineffective, and surgery is required in severe cases of bowel obstruction, rectal prolapse and bleeding[9]. Endoscopic submucosal dissection has been mostly used in recent years as it offers the advantages of speedy recovery, minimal trauma and preservation of colonic integrity[1].

This case is one of the few in medical literature to highlight the direct association between laparoscopic appendectomy and CCP, previously discussed in the literature as a complication post-appendectomy in the American Journal of Gastroenterology[3].

CONCLUSION

In conclusion, it is important to recognise CCP as a potential imitator of several gastrointestinal pathologies. Keeping it on the differential is of benefit to the patient, as this optimises treatment and limits unnecessary interventions. This is especially important in appendectomy patients, as this is a relatively common procedure where CCP might be under-reported due to a lack of clear margins of diagnosis. Further studies are needed to clearly identify and differentiate CCP from other, similar manifestations.

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