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CASE REPORT

Colitis cystica profunda of the rectum: An unexpected operative finding

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Abstract

Colitis cystic profunda is a rare entity benign condition of the colon and rectum that can mimic suspicious polyps or malignancy. The commonest sites of affectation are the rectum and the sigmoid colon but it can be unusually widely distributed in the colon. The aetiology of this condition is not fully elucidated and confident diagnosis can only be made on histological features. We hereby describe a patient who presented with significant rectal symptoms and an unexpected finding of a submucosal mucous cyst mimicking a suspicious rectal polyp and highlighted its significance and the review of the literature.

Key words: Colitis cystica profunda; Suspicious rectal polyp; Solitary rectal ulcer; Rectal prolapse; Adenocarcinoma

Core tip: Colitis cystic profunda is although rare but it is important that it is not mistaken for a mucinous adenocarcinoma, carcinoid tumour, pancreatic heterotopia, inverted or pseudo-invasion of adenomatous polyp of the anorectum for the purposes of treatment and prognostication.

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INTRODUCTION

Colitis cystic profunda (CCP) is a rare and uncommon benign lesion of the colon and rectum characterised by the presence of intramural or submucosal mucous containing cysts. Although it may diffusely involve the entire large bowel, the classical sites are usually in the rectum and sigmoid colon. Knowledge of this pathological entity is necessary and important as it can clinically and histological mimic a malignant lesion[1-3]. The aetiology of this rare condition is unclear and the diagnosis is usually by histology[1-3]. The first description of the lesions was by Stark[4] who found mucous cysts in the colon of two patients at autopsy who died of dysentery. Virchow later introduced the term "CCP" following a report of multiple polypoid cystic submucosal lesions[5]. We describe a case of a lady with rectal symptoms and a submucosal mucous cyst mimicking a suspicious rectal polyp and highlighted its significance and the review of the literature.

CASE REPORT

A 34-year-old lady presented with a history of recurrent fresh rectal bleeding for the past few years and associated more recently with mucus discharge and tenesmus. Apart from diarrhea-predominant irritable bowel syndrome she had no significant past medical problem and no family history of bowel pathology. Anorectal examination and rigid sigmoidoscopy revealed multiple rectal polyps with a particularly predominant one measuring about 3 cm on the right lateral wall of the rectum.

Flexible sigmoidoscopy confirmed a polypoid lesion in the rectum which turned out to be the previously noted 3 cm polyp on the lateral wall of the rectum (Figures 1 and 2). Histology of the multiple biopsies from this lesion and other multiple rectal polyps were reported as a "benign hyperplastic polyp with no dysplasia or malignancy". She subsequently underwent an examination under anesthesia showing a 3-cm submucosal lesion in the right lateral rectal wall with a central pit. There were findings of a small rectocele and rectal mucosal prolapse in addition to the submucosal lesion. A careful excision was achieved by raising the submucosal layer with 0.5% lignocaine and 1 in 200000 adrenaline. Two/zero vicryl stay sutures at the superior and inferior margins were used for retraction to aid excision. A complete excision was achieved through a vertical mucosal incision dissecting the lesion wholemeal from the submucosal layer (Figure 2). The excised lesion was found to be a cystic lesion containing mucus (Figure 3).

The H and E stain of the lesion confirmed cyst lined by atrophic colonic mucosa and colonic mucinous epithelium with surrounding fibrosis. The overlying mucosa shows some degree of benign hyperplasia. The cyst contains inspissated mucin with dystrophic calcification. The cyst extends into the submucosal tissue and up to the muscular layer but no evidence of dysplasia or malignancy is seen (Figure 4).

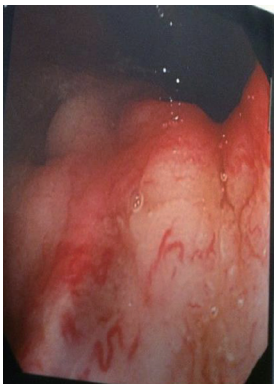

Figure 1 Endoscopic appearance of colitis cystic profunda - a polypoid lesion in the rectum covered by smooth mucosal.

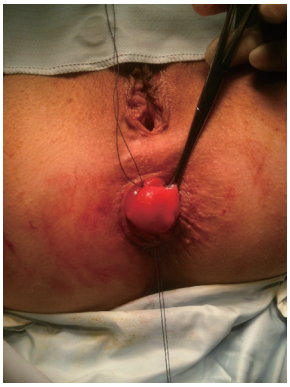

Figure 2 Intraoperative findings - excision through a vertical mucosal incision dissecting the lesion wholemeal from the submucosal layer.

DISCUSSION

CCP is a rare benign condition involving the rectum and colon often presenting as a polypoid lesion[1-3]. This entity is of clinical significance in that it can often mimics malignant lesions of the colorectum to which it must be distinguished[1]. The aetiology of this entity is unknown but there is a proposed theory of congenital origin. It has been found to be associated with several acquired conditions such as rectal prolapse, solitary rectal ulcer (SRU), inflammatory bowel disease, diverticulitis, local rectal trauma and adenocarcinoma[1-12].

Pathologically, CCP is a benign lesion characterized by mucin filled cysts beneath the muscularis mucosa. Its distribution can be in localized form with a polypoidal lesion or in a more diffuse pattern with variable length of the rectal mucosa or colon involved[9]. Both of these patterns of distributions have been well described in the setting of acquired conditions causing mucosal ulceration and inflammation[7]. The localized form has been reported in associated with solitary rectal ulcer syndrome, rectal prolapse[1,10,12] while the more diffuse types have been described in patients with Crohn's disease, ulcerative

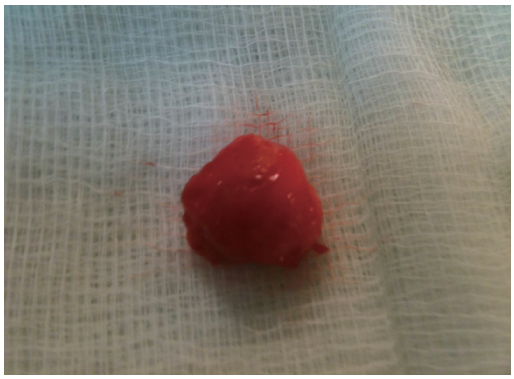

Figure 3 Operative specimen - the excised lesion was cystic containing mucous.

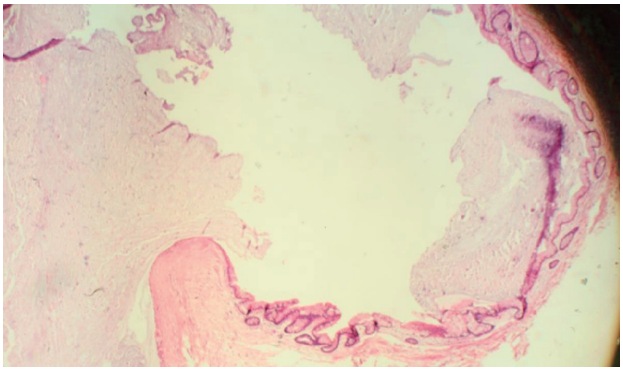

Figure 4 Hematoxylin and eosin stain of histopathology slide - this is a cyst containing inspissated mucin and lined by large bowel type benign mucinous epithelium with surrounding fibrosis.

colitis, radiation, and infectious colitis[7,11]. Mitsunaga *et al*[8] recently reported a case of a single polypoid CCP lesion in association with an adenocarcinoma. Therefore, a careful histopathological examination of all CCP specimens is advisable to exclude an associated malignancy.

The most common symptoms of this pathology include fresh rectal bleeding, mucus discharge, tenesmus, proctalgia fugax, altered bowel habits and a long existing history of obstructive defecation[2,8,12]. There are no peculiar characteristics of CCP on endoscopy. Endoscopic features are usually those of polypoid lesion covered by a normal, oedematous or ulcerated mucosa. Endoscopic anorectal ultrasound where available can be diagnostic as it shows a hypoechoic signal usually in the submucosal layer with no surrounding deeper layer infiltration. These important features distinguish it from malignancy of the anorectum[12]. Computerized tomography scan and/or magnetic resonance imaging typically reveal non-infiltrating submucosal cystic lesion with some loss of perirectal fatty tissue and thickening of levator ani muscles[1,13]. These imaging may show features of SRU and pelvic floor dysfunction. Anorectal physiology studies including evacuatory proctography have been reported to document rectal intussusceptions in up to 45%-80% of patients with localised CCP[14]. Features of localised CCP of the rectum are in most cases not distinguishable from those of SRU as they tend to have similar underlying factors including evacuatory dysfunction and rectal mucosal prolapse. However, this relationship between CCP and SRU is not always demonstrable in all patients. Association of rectal prolapse has been reported in up to 54% of localised CCP[1,15].

Treatment of CCP is mostly conservative and surgery is only indicated in patients with severe symptoms with associated significant rectal prolapse. First-line management option should commence with dietary and lifestyle changes. This is aim at avoiding constipation and straining during defecation. This treatment regime can be aided by the use of bulking laxatives, stool softeners and lubricants, hydrocortisone enemas and sucralfate[7,14]. Bowel retraining with biofeedback therapy may be successful in some group of patients[16]. Patients who fail to respond to conservative measures, with persistent, relapsing symptoms, obstructive defecation and full thickness rectal prolapse or where the initial diagnosis is not definite should be considered for surgery. Operative options include transanal excision of CCP as in this reported case, rectal mucosal excision (Delorme's operation), perineal rectosigmoidectomy (Altemeier's operation) with coloanal anastomosis, stapled transanal resection of rectum (STARR procedure) or abdominal approach laparoscopic ventral rectopexy[2,13,17].

Although CCP is rare, it is important to be aware of this clinical and pathologic entity so that it is not mistaken for a mucinous adenocarcinoma, carcinoid tumour, pancreatic heterotopia, inverted or pseudo-invasion of the adenomatous polyp of the anorectum for the reasons of therapy and prognosis.

COMMENTS

Case characteristics

A 34-year-old lady presented with significant rectal symptoms and an unexpected finding of a subucosal mucous cyst mimicking a suspicious rectal polyp.

Clinical diagnosis

A suspected 3 cm rectal mucosal polypoid lesion on the right lateral wall on sigmoidoscopy and at examination under anaesthesia.

Differential diagnosis

Mucinous adenocarcinoma, carcinoid tumour, pancreatic heterotopia, inverted or pseudo-invasion of the adenomatous polyp of the anorectum.

Pathological diagnosis

Histopathology with H and E of the lesion confirmed cyst lined by atrophic colonic mucosa and colonic mucinous epithelium with surrounding fibrosis. The overlying mucosa shows some degree of benign hyperplasia. The cyst contains inspissated mucin with dystrophic calcification. The cyst extends into the submucosal tissue and up to the muscular layer but no evidence of dysplasia or malignancy is seen. These features are in keeping with colitis cystica profunda.

Treatment

Complete surgical excision of lesion.

Related reports

Colitis cystic profunda (CCP) is a rare and uncommon benign lesion of the colon

and rectum characterised by the presence of intramural or submucosal mucous containing cysts. Knowledge of this pathological entity is necessary and important as it can clinically and histological mimic a malignant lesion.

Term explanation

CCP was first described as mucous cyst lesion by Stark. Virchow later introduced the term CCP.

Experiences and lessons

This pathological entity can be confused with mucinous adenocarcinoma, carcinoid tumour, pancreatic heterotopia, inverted or pseudo-invasion of the adenomatous polyp of the anorectum. Knowledge of this entity is therefore important to the clinician in guiding therapeutic intervention.

Peer-review

This is a well written case report, with adequate review of the literature. It is important to consider CCP in the differential of rectal submucosal lesions.

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