MRI Appearances of Colitis Cystica Profunda: A Rare Benign Mimicker of Colorectal Malignancy

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ABSTRACT

Colitis cystica profunda (CCP) is a rare benign disease characterised by mucin-filled cysts beneath the muscularis mucosae, which commonly involves the rectum and sigmoid colon. The disease process involving rectum is termed as proctitis cystica profunda. Trans-rectal ultrasound (TRUS) and magnetic resonance imaging (MRI) are an aid to diagnosis. However, histological analysis of deep biopsy is diagnostic. We report a case of 21-year male who presented with bleeding per rectum and was diagnosed as solitary ulcer of the rectum (SRUS) on superficial biopsies on sigmoidoscopy. Surgical intervention coupled with MRI findings and histopathological analysis formulated diagnosis of CCP. To our knowledge, we are second to report this entity with its MRI features.

Key Words: *Colitis cystica profunda. Solitary rectal ulcer syndrome. MRI appearance.*

INTRODUCTION

Colitis cystica profunda (CCP) is an unusual benign recto-colonic condition characterised by intramural mucin-filled cysts; which, because of its clinical expression and the way it appears on imaging studies, can mimick neoplasm of colon.[1,2] Stark first described it in the colon of two autopsied cases of chronic dysentery and Vinchow was the one to name it 'colitis cystica polyposa'.[2-6] Rectum and the sigmoid colon are the commonest sites of affliction, but wide colonic distribution is also seen.[2]

CCP has similar histological features as solitary rectal ulcer syndrome (SRUS) and the overlap between these conditions is not fully elucidated. CCP usually presents with nonspecific symptoms of mucorrhea, abdominal pain, rectal bleeding, tenesmus, and constipation; frequently misdiagnosed or confused with a malignancy, sometimes resulting in unnecessary interventions, be it surgical or pharmacological.[3,4] Our case illustrates the difficult diagnosis of this uncommon but problematic phenomenon, awareness of which is paramount for the concerning radiologists and clinicians.[5,6]

CASE REPORT

A 21-year male presented to our hospital with complaint of bleeding per rectum for two and an half years with associated history of weight loss. His laboratory investigations, which included tumor markers, were unremarkable. On digital rectal examination, a polypoidal lesion was found in rectum, 3 cm from anal verge at 9-10 o'clock position. Flexible sigmoidoscopy revealed significant mucosal edema and erythema in rectum with two superficial ulcers in mid rectum, measuring 4 and 2 mm, respectively, from where superficial biopsy was taken and it reported SRUS. However, patient's symptoms worsened and possibility of carcinoma of rectum was raised. Since MRI is the modality of choice to assess colon, his MRI was advised which showed extensive diffuse wall thickening of rectum with multiple multiloculated cystic areas in submucosal and intramural distribution within the rectum, possibly representing prominent glands/retained secretions with significant mucosal enhancement and few small perirectal nodes (Figures 1-3). Since colonic malignancy remained a question, he underwent laproscopic anterior resection with defunctioning diversion ileostomy. Histopathological analysis of deep biospsy revealed large bowel mucosa showing ulceration, splaying of muscle fibers, extravasated mucin with multiple cystic spaces filled with mucin; and lymph nodes showing reactive changes without evidence of granulomatosis or malignancy. Overall findings were characterised as CCP.

DISCUSSION

CCP is a benign condition with reactive changes in mucosa, characterised by submucosal cysts filled with mucin and lined by benign epithelium which might imitate carcinoma of colon from a radiologic and pathologic viewpoint.[2,7] CCP can present in a localised form with a polypoid lesion, or as a diffuse process involving a variable length of the rectal mucosa or colon, having predilection for male gender.[3,8,9] The etiology of this entity is not fully elucidated, but it has been described to have inflammatory/post traumatic origin with associations with a number of ulcerating diseases including inflammatory bowel disease, SRUS, diverticulosis, and infectious colitis. Features of CCP can overlap with carcinoid tumour, mucinous adenocarcinoma, pancreatic heterotopia or adenomatous polyps of the anorectal

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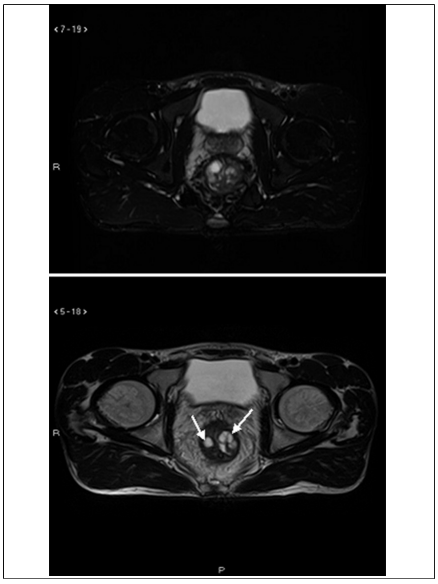

Figure 1: MRI axial T2 and fat sat images showing multiple multiloculated cystic areas in submucosal and intramural distribution within the rectum (arrows).

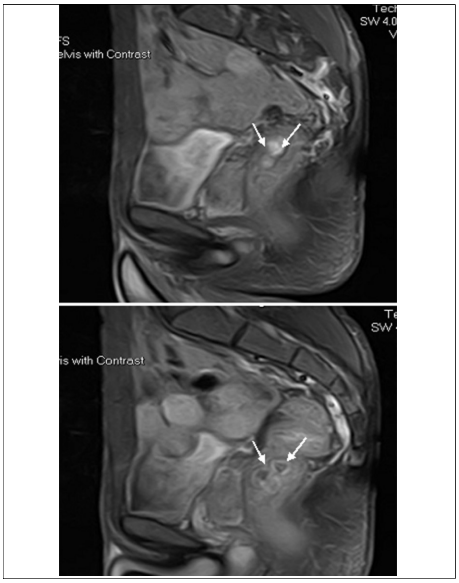

Figure 3: MRI saggital post contrast T1 weighted images showing extensive mucosal enhancement (arrows).

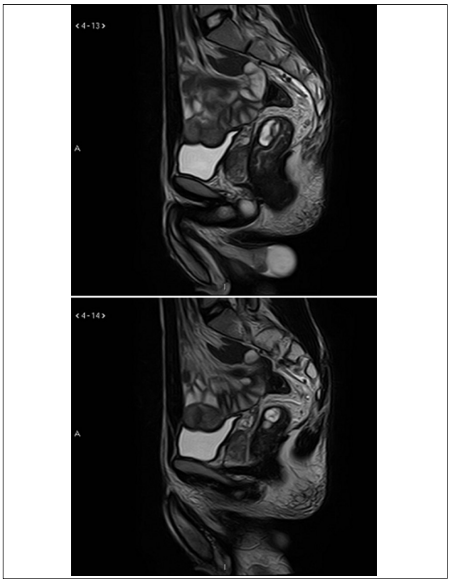

Figure 2: MRI sagittal T2 weighted images showing multiple submucosal cysts without any deeper layer infiltration (arrows).

region (inverted/pseudo-invasion).[3,4,9] Patients may present with a variety of symptoms, the most common being constipation, bleeding per rectum, mucus discharge, and abdomen pain.[5]

Colonoscopy shows polypoidal/sessile lesions enclosed within normal, edematous or inflamed mucosa and/or ulcerations.[4]

Barium enema exhibits nodularity of the mucosa, polypoidal lesions, rectal stricture/ulceration, with thickened rectal folds; all of which are inconclusive findings.[6] Endoscopic anorectal ultrasound can be diagnostic by showing hypoechoic signal in the submucosal layer without adjacent deeper or muscular layer involvement, which clearly differentiates benign from malignant disease process.[2,6] MRI typically reveals cystic, non-infiltrative, submucosal lesions with thickening of levator ani muscles, some loss of perirectal fat tissue or nodules with high signals on T2 sequences due to mucoprotein content of the cysts.[3,8,9]

Removal of lesions by colonoscopy, followed by histopathology, is the gold-standard for clinching the final diagnosis. Key histological features are obliteration of the lamina propria, muscle fibers extending upwards between the crypts with hypertrophied muscularis mucosae and glandular cryptic abnormalities.[6] Typically, there is a fibrous stroma (distinguished from desmoplastic stroma usually identified in adenocarcinoma) with mucin extravasation/mucin cysts without significant accompanying inflammation.[8]

Treatment depends on the severity of symptoms but generally targets on amelioration of constipation.[5] Laxatives and dietary modification may improve symptoms of some patients, while others may require a surgical intervention.[4,5]

Establishing the correct diagnosis for CCP is indispensable to avoid misdiagnosis accompanying extensive resection and oncologic attitude for a benign lesion that could be treated with appropriate measures, essentially maintaining the patient's quality of life.[9]

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