

CASE REPORT

특발성 정맥경화성 대장염: 만성 허혈성 장염의 드문 예

최종민, 이강녕, 김해수, 이상기, 이정규, 이성원, 이오영, 최호순
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Idiopathic Phlebosclerotic Colitis: A Rare Entity of Chronic Ischemic Colitis

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Colonic wall thickening is frequently encountered in various conditions, from acute or chronic inflammatory disease to colorectal carcinoma. Colonic wall thickening may be accompanied by calcifications in mucinous adenocarcinoma of the colon, leiomyosarcoma of the colon, schistosomiasis japonica, and phlebosclerotic colitis. Phlebosclerotic colitis is a rare entity of chronic ischemic colitis associated with sclerosis and fibrosis of mesenteric veins. Although its development is usually insidious, and, thus its diagnosis can be delayed, characteristic findings in phlebosclerotic colitis are calcifications of mesenteric veins as well as colonic wall thickening with calcifications. We report on a 71-year-old woman who presented with chronic diarrhea and intermittent hematochezia, who was first misdiagnosed as mucinous adenocarcinoma of the colon, but finally diagnosed as a rare entity of chronic ischemic colitis, phlebosclerotic colitis. Differential points of phlebosclerotic colitis from other diseases, including leiomyosarcoma and schistosomiasis japonica, are also described. (**Korean J Gastroenterol 2014;63:183-186**)

Key Words: Ischemic colitis; Mesenteric veins; Sclerosis; Vascular calcification

INTRODUCTION

Colonic wall thickening is frequently encountered in clinical practice. It is related to a variety of entities, including normal variants, inflammatory conditions, and colonic neoplasms.¹ Development of colonic wall thickenings may be not only primary, but also secondary in many clinical settings associated with body fluid retention due to heart failure, chronic liver disease, and renal insufficiency, and in other infiltrative diseases such as amyloidosis. Colonic wall thickenings can be accompanied by calcifications in mucinous adenocarcinoma of the colorectum,^{1,2} leiomyosarcoma of the colon,³ schistosomiasis japonica,⁴ and phlebosclerotic colitis (PC), a rare entity of ischemic colitis.

Ischemic colitis may develop by occlusion or stenosis in mesenteric arteries or veins due to atherosclerotic, thrombotic, or embolic causes, or by insufficient tissue perfusion without occlusive lesions of mesenteric vessels. Ischemic colitis by non-thrombotic occlusion of mesenteric veins is rarely reported; however, it may be associated with systemic disease, such as amyloidosis, lupus, or rheumatoid arthritis. Ischemic colitis due to disturbances in mesenteric blood flow due to sclerotic changes and calcifications of the venous wall is defined as PC. However, the etiology and pathogenesis of this rare entity of chronic ischemic colitis has not yet been clearly defined. Herein, we report on a case of colonic wall thickening with calcifications in PC, which was first misdiagnosed as a mucinous adenocarcinoma of the colon.

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CASE REPORT

A 71-year-old woman was admitted because of chronic diarrhea and hematochezia for the past seven months. One year ago, she had found that her stool caliber became narrow. However, no other alarming symptoms or signs suggestive of malignancy, including weight loss and fever, were noted. She had been living in America since immigrating 10 years ago. Concern for medical costs urged her to return to her hometown city, Seoul, Korea for further evaluation. Just before admittance to our hospital, she had undergone abdominal CT at a local clinic and was informed that colonic cancer was suspected. She does not drink alcohol and had not taken any herbal medications, and she had no past medical history of chronic liver or kidney disease.

At admission, vital signs were body temperature of 36.8°C, pulse rate of 82/min, respiratory rate of 18/min, and blood pressure of 120/70 mmHg. Abdomen was soft and non-tender, but bowel sounds were slightly increased. Digital rectal examination found no rectal bleeding. Laboratory findings were white blood cells of 3,800/mm³, hemoglobin of 11.8 g/dL, platelets of 251,000/mm³, total protein of 6.8 g/dL, albumin of 3.9 g/dL, BUN 10.6 mg/dL, creatinine 0.72 mg/dL, total bilirubin 0.43 mg/dL, AST 15 U/L, ALT 6 U/L, and CEA 2.9 ng/mL. Chest X-ray showed no enlargement of the heart. Electrocardiogram showed non-specific ST-T wave change. No ova or cyst suggesting infection of parasite was detected in stool examination.

We found multiple linear radiopacities in the right abdominal area on simple X-ray (Fig. 1). Abdominal CT showed diffuse thickening of bowel walls with mural calcifications from the cecum to splenic flexure; surrounding mesenteric veins were also calcified (Fig. 2). Wall thickening with calcifications of the colonic wall and mesenteric vein was not observed on the distal colonic wall. On the other hand, abdominal CT showed no findings suggestive of portal hypertension, such as contrast enhancement of the paraumbilical vein, dilated portal or mesenteric veins, splenomegaly, or ascites. In addition, no varices or portal hypertensive gastropathy suggesting portal hypertension were found on upper gastrointestinal endoscopy. Colonoscopy showed dark purple edematous mucosa and areas of luminal narrowing with multiple erosions or ulcerations (Fig. 3). The colonic mucosal lesions were distributed from the cecum to the sigmoid colon, but became less severe toward the distal colon with only mild dark edematous mucosa in the sigmoid colon. Microscopic examinations of colonic tissues obtained from the ascending and transverse colon showed marked thickening and calcifications of the colonic mucosal and submucosal layers with ulcerations and a large focus of dystrophic calcification. Mesenteric veins also showed an abnormally thickened wall with hyalinization (Fig. 4). After conservative management, she was discharged and returned to the United States.

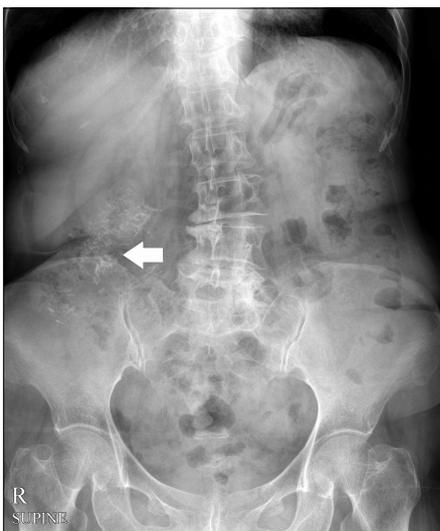


Fig. 1. Simple radiograph shows linear calcifications in the right abdominal area (arrow).



Fig. 2. Abdominal CT shows colonic wall thickenings accompanied by mural and mesenteric venous calcifications in the ascending colon (arrows).

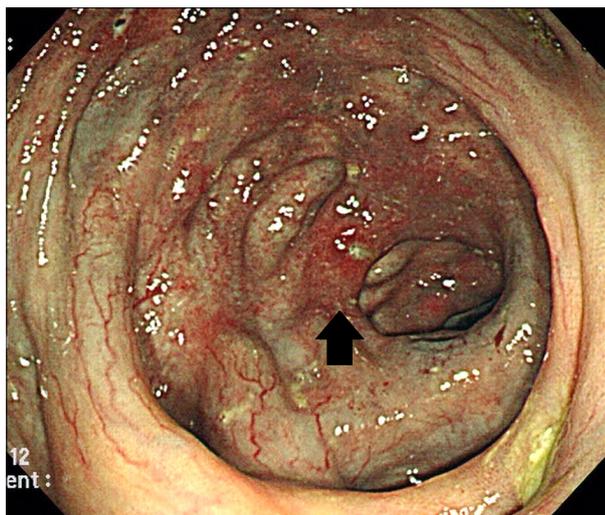


Fig. 3. Colonoscopy shows dark purple edematous mucosa with areas of luminal narrowing in the ascending colon. Multiple erosions or ulcerations with surrounding erythema are also noted (arrow).

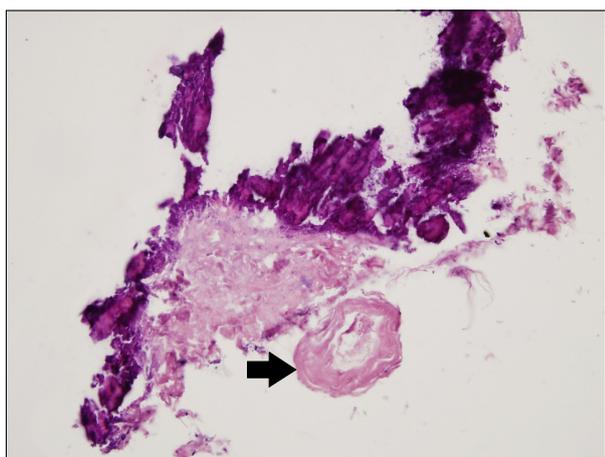


Fig. 4. Microscopic examinations. Marked thickening and calcifications of the colonic mucosal and submucosal layers are noted. Mesenteric veins show an abnormally thickened wall with hyalinization (arrow) (H&E, $\times 200$).

DISCUSSION

Colonic wall thickenings are frequently found because colonic responses are nonspecific to various stimuli, including infection, inflammation, or ischemia. This case of colonic wall thickening represents a rare entity of chronic ischemic colitis, PC. Although the suggested pathogenesis was that mesenteric veins gradually become sclerotic and fibrotic resulting in colonic wall thickening with calcifications, currently, its etiology and pathogenesis are not clearly defined.

Whatever the etiology and pathogenesis of PC are, its char-

acteristic finding of colonic wall thickening with calcifications on CT should be distinguished from mucinous adenocarcinoma, leiomyosarcoma, and schistosomiasis japonica. In mucinous adenocarcinoma of the colon, colonic wall thickening with mural calcifications is irregular and asymmetric with short-segment involvement, compared with regular and symmetric thickening with long segment involvement in PC.¹ Wall thickening in mucinous adenocarcinoma also tends to spread abruptly to the unaffected colonic wall and to cause obstruction of the bowel due to severe luminal narrowing. However, in our case, asymmetric thickening and severe narrowing in certain areas of the colonic wall might have caused confusion in differentiation of PC from mucinous adenocarcinoma, although the involvement of the long segment of the colon was strongly suggestive of PC. In leiomyosarcoma, a mesenchymal cancer of the colon, colonic wall thickenings are bulky mural masses with calcifications, heterogeneous hyper-enhancement, exophytic growth, and cystic degeneration.³ In schistosomiasis japonica, in which schistosoma japonicum, a kind of parasite, causes granulomatous disease in the intestine, colonic wall thickening with calcifications indicates calcified eggs originating from larvae matured in the portal vein, and are frequently shown in the distal colon compared with proximal involvement in PC.⁴ Taken together, the presence of mesenteric venous calcifications around the affected colon on abdominal CT is a differential point of PC from these diseases.

Colonoscopy of our patient showed dark purple discolorations of the mucosa with mucosal edema and erythema, erosions or ulcerations, and luminal narrowing, which are characteristic findings in PC. The dark purple discoloration of the mucosa seems to be caused by chronic congestion with ischemia associated with sclerotic and fibrotic changes of the mesenteric veins. However, certain toxic agents that can show staining in the colonic mucosa after being absorbed through the colonic wall were also suggested as a cause. In addition, the reason that PC usually involves the right-sided colon was also suggested to be due to the fact that certain toxic agents metabolized by colonic bacteria in the proximal colon were absorbed in the region.⁵ However, in our patient, mucosal lesions of varying severity were distributed throughout the colon, ranging from the cecum to the sigmoid colon with no skipped lesions. One report suggested that it may gradually progress to the entire colon.⁶ In addition, our pa-

tient denied use of any herbal medication and had no predisposing conditions that might increase mesenteric venous pressure. Therefore, the cause of PC in our patient seems to be idiopathic.

Microscopic findings of PC are variable. Colonic wall thickenings consist mainly of marked submucosal fibrosis; however, they may contain a variable degree of vascular congestion, hemosiderin-laden macrophages, erythrocyte extravasation, and brown to black-pigmented macrophages. Venous wall thickenings around the involved colon and mesentery are also shown, and are accompanied by fibrosis, sclerosis, and hyalinization as well as calcifications, which are suggested to result from coagulative necrosis in the muscle layer and subsequent myointimal hyperplasia of mesenteric veins.⁷ Then, the hyperplastic myointima are damaged repeatedly, leading to gradual occlusion of the mesenteric veins. These phlebosclerotic changes may be caused by long-term elevated pressure to the veins in conditions such as right-sided heart failure, liver cirrhosis, and portal hypertension.^{2,8} In addition, the suggested predisposing conditions include diabetes mellitus, dyslipidemia, and hemodialysis.^{5,9} On the other hand, all of these changes of the mesenteric veins are supposed to be attributable to certain toxic agents that are absorbed into venous return of the proximal colonic wall. Similarly, association of chronic use of herbal medicine and predilection of East Asians of female gender with PC has been reported.⁵

As in our patient, symptoms of PC include abdominal discomfort, diarrhea, constipation, hematochezia, and weight loss with chronic and insidious development.² The chronic and insidious development of symptoms may be attributable to the chronic congestion of mesenteric veins, thus resulting in delayed diagnosis of PC. For diagnosis of PC, other modalities except abdominal CT or colonoscopy are complementary. In simple abdominal radiography, multiple threadlike calcifications may be noted along the right hemicolon. Barium study of the colon shows disappearance of semilunar folds, luminal irregularities, and overall stenosis with rigidity. Angiography in PC may show narrowing of the marginal arteries with dilatation and tortuosity of veins along the vasa recta. In terms of treatment, reports have suggested that conservative management with bowel rest and hydration are sufficient in PC.² However, when conservative management fails to result in improvement of persistent abdominal pain or

ileus, surgery may be required.² PC may be complicated by dehydration, hemorrhage, perforation, and death.¹⁰

This report indicates that the combination of chronic abdominal discomfort with diarrhea, right-sided multiple calcifications on a simple abdominal radiography, dark purple and edematous mucosa with erosions and luminal narrowing on colonoscopy, and wall thickening with calcifications both in colon and in mesenteric veins is suggestive of PC, a very rare entity of chronic ischemic colitis. In addition, other diseases of colonic wall thickening with calcifications should be differentiated, particularly mucinous adenocarcinoma of the colon, although the differentiation might not be difficult because mucinous adenocarcinoma shows focal thickening of the colonic wall instead of long segment involvement. Therefore, awareness of this disease entity is needed because despite insidious and rare presentation of PC, its endoscopic and radiologic findings are characteristic.

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