DOI：10.6314/JIMT.202104_32(2).07 內科學誌 2021：32：136-141

A Palpable Abdominal Mass Caused by Phlebosclerotic Colitis: A Case Report

Jow-Jyh Hwang

Division of Hepatogastroenterology, Department of Internal Medicine, St. Martin De Porres Hospital

Abstract

Phlebosclerotic Colitis is a rare intestinal ischemic disease of unclear etio-pathogenesis seen almost exclusively in Asians and people of Asian descent. Most cases of Phlebosclerotic Colitis are initially indolent in clinical course, but it may also become lethal in severe cases. The condition predominantly affects the right-sided colon and the imaging specific pictures play a crucial role in its diagnosis. Here we report a Taiwanese female case of phlebosclerotic colitis with a palpable abdominal mass and a history of consuming herbal medicine for years. Examination of abdominal radiograph and colonoscopy could help to make clinical diagnosis. (J Intern Med Taiwan 2021; 32: 136-141)

Key Words: Phlebosclerotic colitis, Right-sided colon, Herbal medicine

Introduction

Ischemic colitis is primarily caused by arterial obstruction secondary to arteriosclerosis, thrombosis or embolism in the left-sided colon, especially in patients with risk factors including smoking, hypertension, arteriosclerosis, and diabetes mellitus[1]. Mesenteric venous abnormalities may also result in colon ischemic damage. Phlebosclerotic colitis (PC) is a rare chronic ischemia disease of the large intestinal bowel characterized by thread-like calcification of the mesenteric venous system[2] and its clinical presentation almost exclusively affects the right-sided colon[3]. Most case reports of PC are seen in the Asian population or those with Asian ancestry[2-5]. The etiology and pathogenesis of PC are not yet fully understood[6], but herb drug consumption may have contributed to the formation of PC[7-13]. Because colon mucosa biopsy results can be only ischemic pathology and non-diagnostic, knowledge of its characteristic radiographic imaging patterns and endoscopic appearances can help make a diagnosis[3]. However, characteristic computed tomography (CT) and endoscopic features may be missed or misdiagnosed due to the lack of knowledge of this rare entity. Here, we report a Taiwanese female patient with a right-sided PC with a palpable abdominal mass presentation and a history of herbal medicine intake.

Case report

A 53-year-old female was sent to the hospital because of exacerbated right abdominal pain for one day. She had previously been healthy. She had no

Reprint requests and correspondence：Dr. Jow-Jyh Hwang
Address：Division of Hepatogastroenterology, Department of Internal Medicine, St. Martin De Porres Hospital, No. 565, Sec. 2, Daya RD., Chiayi, 60069, Taiwan

history of diabetes, hypertension or other underlying systemic disease. She gave a history of gradual onset of right abdominal pain with constipation for 2 years, but she didn't pay attention to it. She is a non-smoker and teetotaler. There was no remarkable family history. The patient had used herbal medication "Xiao yao san" (containing sanshishi) for more than 10 years.

On physical examination, she was well developed and nourished, but had an ill-looking appearance. Her blood pressure was 130/80 mmHg, pulse rate 90 per min, respiratory rate 18 per min and body temperature 37.5°C. The head was normal and the neck was supple. The heart was normal and the lungs were clear. The abdomen was soft, but a palpable mass 8*5cm with local tenderness over right abdomen was noted. No rebounding pain, no muscle guarding and decreased bowel sounds were noted. Laboratory tests were as follows: white blood cell 6,040/mm^3, hemoglobin 10.9 g/dL, MCV 70.5 fL, Platelet count 252,000/L. The biochemistry and electrolyte levels were all within normal limits. A plain radiograph (Figure 1) showed linear threadlike calcifications oriented along the ascending colon extending toward the transverse colon. No features of bowel obstruction were noted.

Abdominal CT (Figure 2) revealed colonic wall thickening of the ascending colon and transverse colon, along with characteristic threadlike or serpentine calcified subserosal and mesenteric veins in the distribution of the cecum and ascending and transverse colon. Besides, long segmental small bowel loops dilatation was also noted. PC was impressed according to specific radiographic pictures.

Because of the patient's request, she was transferred to medical center for further therapy. Herbal medicine was discontinued. Her symptom improved after conservative treatment with pain killer and stool softener. After one year, she visited our hospital where colonoscopy was arranged. Colonoscopy (Figure 3) revealed edematous dark-bluish colonic mucosa, lumimal stenosis, overlying ulcer, progressively worsening while traversing through the transverse colon into the ascending colon and cecum. The endoscopic biopsy specimens revealed markedly increased perivascular collagen (Figure 4) and chronic ischemic change. Compared to her clinical symptoms, specific radiographic and endoscopic pictures, the patient's diagnosis was compatible with PC.

Discussion

PC is a rare disease of intestinal ischemia, but is mostly neglected during the early stage of the disease. Koyama et al reported a case of chronic ischemic colitis causing stenosis in 1991[14]. In 2000, Iwashita et al. first reported PC as a new disease entity, describing it as an ischemic lesion resulting from phlebosclerosis[15]. The signs and symptoms of PC are non-specific, such as constipation, diarrhea, nausea, vomiting, abdominal distension, abdominal

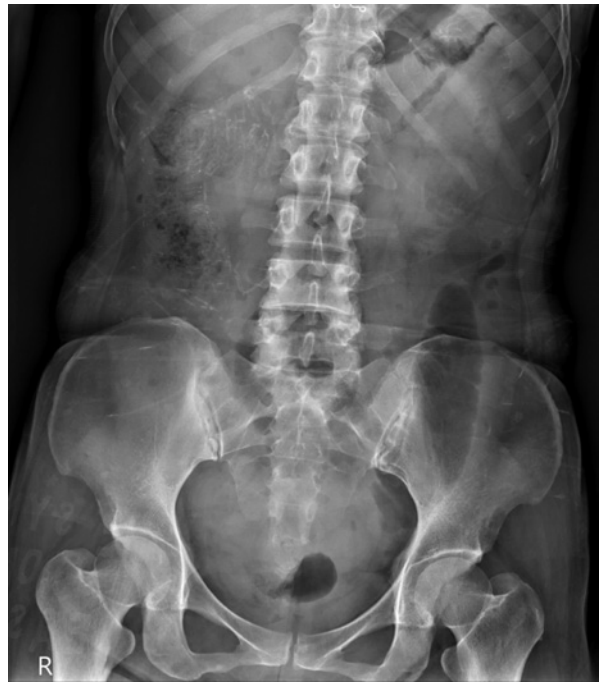

Figure 1. A plain abdominal radiograph showed linear threadlike calcifications oriented along the ascending colon extending toward the transverse colon.

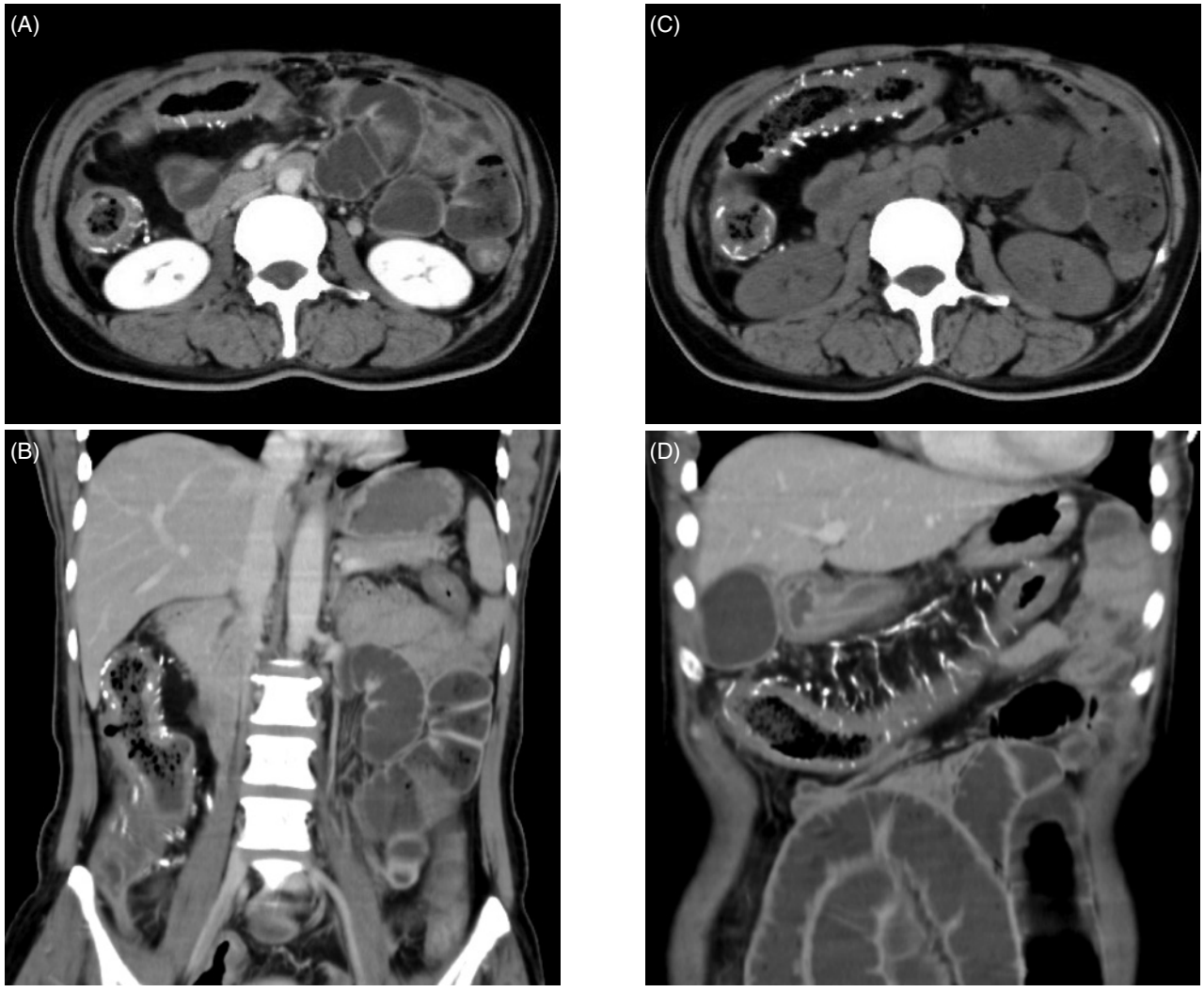

Figure 2. (A) (B) Abdominal CT revealed colonic wall thickening of the ascending colon, along with characteristic threadlike or serpentine calcified subserosal and mesenteric veins. (C) (D) Abdominal CT revealed transvers colonic wall thickening with threadlike or serpentine calcified subserosal and mesenteric veins.

pain, and gastrointestinal bleeding[3,16]. In general, the clinical symptoms depended on the severity of the disease which was attributable to the ischemic change in the colon secondary to the sclerosis of the draining veins and the onset of process was generally gradual. Some patients in the early stage of the disease can even be asymptomatic, while those with advanced disease can present with intestinal obstruction[17] and even perforation[18]. In addition to local abdominal tenderness, our patient also presented with a palpable abdominal mass due to colon congestion, calcification and wall thickening.

The pathogenesis of PC had not been well understood, but dialysis, portal hypertension, diabetes, and vasculitis have been suggested as possible causes[19,20]. Most case reports of PC are seen in the Asian population or those with Asian ancestry[2-5]. It is believed that ingested biochemicals and toxins, especially herbal medicine, invigorant water of unknown contents, and alcohol have been associated with the disease [7-9,21]. Our patient also had a history of long term herbal medicine usage, including sanshishi (gardenia fruit), an ingredient has been proven to be strongly associated with PC[10].

PC has a characteristic radiological and endoscopic appearance that can help diagnose the

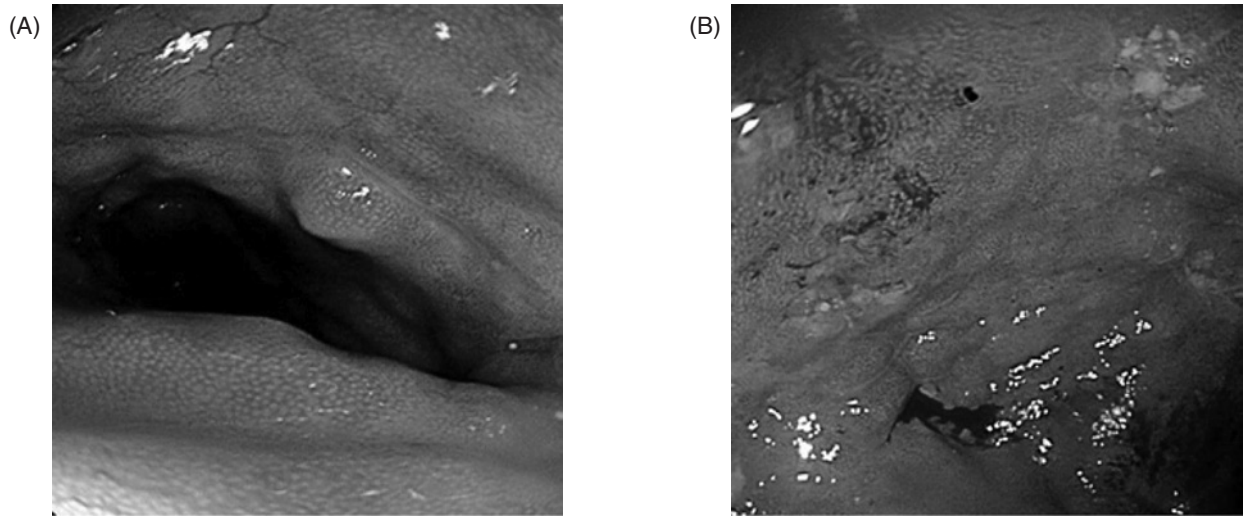

Figure 3. Colonoscopy revealed typical PC edematous dark-bluish colonic mucosa, lumimal stenosis and overlying ulcer through the transverse colon into the ascending colon.

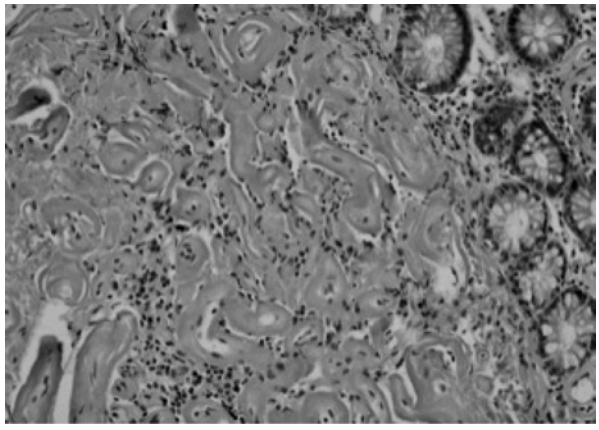

Figure 4. The endoscopic biopsy specimens revealed markedly increased perivascular collagen (H&E stain, x 200).

disease. The most striking and characteristic findings on plain ragiography[3] and abdominal CT[3,21] are multiple fine, tortuous, thread-like, or serpentine mesenteric venous calcifications that are perpendicularly oriented to the long axis of the affected colonic wall. Our patient also showed these specific pictures in ascending colon extending toward transverse colon, a typical location in this disease[21].

As is well known, water and some water-soluble materials absorption predominantly occurred in the ascending colon and cecum. In PC, certain toxic biochemical agents or water-soluble irritants, probably existing in the frequently ingested contents and absorbed to the venous return, may result in chronic venous damage of the colon[9]. In the initial stage of the disease, calcifications first involve the peripheral mesenteric veins, followed by the intramural tributaries of mostly the ascending colon along with sclerosis of the colonic muscular wall. With chronic stasis within the lumen leads to distal migration and disease progression, calcification of the mesenteric venous system extend to involve the transverse colon and even the entire colon[21,22]. The affected colon wall becomes thickened, which interferes with motility and may lead to subserosal calcifications and luminal stenosis[10], and results in characteristic colonoscopic pictures, such as mucosa dark purple discoloration, erythema, mucosal edema, erosion, ulceration, luminal narrowing , loss of normal haustra, rigidity of the colonic wall, and focal nodular surface[3]. Colonic wall thickening accompanied by calcifications may occur in other diseases like mucinous adenocarcinoma, leiomyosarcoma, schistosomiasis japonica, etc., in the literature report[23]. Other causes of chronic mesenteric ischemia such as Churg-Strauss syndrome, Behcet's disease, lymphocytic phlebitis, Degos disease, Wegener's granulomatosis, and

polyarteritis nodosa do not cause fibrosis or calcifications in the veins[6,21].

Nowadays, although more and more patients had been described in Taiwan[12,13,16,22,24], there was no consensus on the treatment. Management of PC is mostly conservative with close follow-up. Patients with long-term use of Chinese herbs and medical liquor were asked to discontinue. Surgery is reserved for those patients with severe complications (e.g., intestinal obstruction, perforation, and hemorrhage) and for those with persistent symptoms following conservative management[6]. If colectomy was performed, the histopathologic characteristics included marked thickening of the venous wall with calcifications, fibrosis, collagen fiber perivascular deposits, narrowing of the lumen with no thrombosis, which mainly involved the submucosa of colonic wall. In some patients, mucosal atrophy, ulceration, necrosis, and inflammation were also found. In our case, radiologic findings showed mesenteric vein calcifications, but endoscopic colon mucosa biopsy could not reveal mesenteric vein calcifications. Since a mucosal biopsy may not be sufficient for evaluation of submucosal vessels[4]. Definitely, tissue proof of PC needs colectomy. Knowledge of its distinctive endoscopic and radiologic appearance would help to make the diagnosis.

Acknowledgment

We would like to acknowledge the contribution and expertise of Dr. Chan Wai-Bin and Dr. Huang Yu-Hua for this case report.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Gandhi SK, Hanson MM, Vernava AM, et al. Ischemic colitis. Dis Colon Rectum 1996; 39(1): 88-100.
2. Iwashita A, Yao T, Schlemper RJ, et al. Mesenteric phlebosclerosis: a new disease entity causing ischemic colitis. Dis Colon Rectum 2003; 46: 209-20.
3. Chen W, Zhu H, Chen H, et al. Phlebosclerotic colitis: Our clinical experience of 25 patients in China. Medicine 2018; 97: e12824.
4. Lee SH, Kim JW, Park SJ, et al. Obstructive ileus caused by phlebosclerotic colitis. Intest Res 2016; 14(4): 369-74.
5. Fang YL, Hsu HC, Chou YH, et al. Phlebosclerotic colitis: A case report and review of the literature. Exp Ther Med 2014; 7(3): 583-6.
6. Park JK, Sung YH, Cho SY, et al. Phlebosclerotic colitis in a healthy young woman. Clin Endosc 2015; 48: 447-51.
7. Hiramatsu K, Sakata H, Horita Y, et al. Mesenteric phlebosclerosis associated with long-term oral intake of geniposide: an ingredient of herbal medicine. Aliment Pharmacol Ther 2012; 36: 575-86.
8. Nomura K, Kikuchi D, Iizuka T, et al. Idiopathic mesenteric phlebosclerosis associated with long-term use of Chinese herbs: a case report [in Japanese]. Nihon Shokakibyo Gakkai Zasshi 2012; 109: 1567-74.
9. Miyazaki M, Nakamura S, Matsumoto T. Idiopathic mesenteric phlebosclerosis occurring in a wife and her husband. Clin Gastroenterol Hepatol 2009; 7: e32-3.
10. Shimizu S, Kobayashi T, Tomioka H, et al. Involvement of herbal medicine as a cause of mesenteric phlebosclerosis: Results from a large-scale nationwide survey. J Gastroenterol 2017; 52: 308-14.
11. Chang KM. New histologic findings in idiopathic mesenteric phlebosclerosis: clues to its pathogenesis and etiology—probably ingested toxic agent-related. J Chin Med Assoc 2007; 70: 227-35.
12. Wang CH, Chen TY, Chin J et al. Phlebosclerotic colitis: A case with a History of Herbal Ingestion. J Soc Colon Rectal Surgeon(Taiwan) 2012;23:129-134.
13. Kao CN, Lin PY, Chang LC, et al. A rare cause of ischemic colitis: A case series of idiopathic mesenteric phlebosclerotic colitis from two medical centers in Taiwan. Adv Dig Med 2020;1-6.
14. Koyama N, Koyama H, Hanagima T, et al. Chronic ischemic colitis causing stenosis: report of a case [in Japanese]. Stomach Intestine 1991; 26: 455-60.
15. Yao T, Iwashita A, Hoashi T, et al. Phlebosclerotic colitis: value of radiography in diagnosis-report of three cases. Radiology 2000; 214: 188-92.

16. Lin PY, Chai CY, Chang CC. Recurrent abdominal pain in a 55-year-old woman. Diagnosis: idiopathic mesenteric phlebosclerosis. Gastroenterology 2011; 141: 36, 404.
17. Chang YY, Lin HH, Lin CC. Phlebosclerotic colitis presenting as intestinal obstruction. Clin Gastroenterol Hepatol 2014; 12: 2014-12.
18. Klein S, Buchner D, Chang DH, et al. Exclusive phlebosclerosis of submucosal veins leading to ischemic necrosis and perforation of the large bowel: First European case. Case Rep Gastroenterol 2018; 12: 137-42.
19. Song JH, Kim JI, Jung JH, et al. A case of phlebosclerotic colitis in a hemodialysis patient. Korean J Gastroenterol 2012; 59: 40-3.
20. Kang HY, Noh R, Kim SM, et al. Phlebosclerotic colitis in a cirrhotic patient with portal hypertension: the first case in Korea. J Korean Med Sci 2009; 24: 1195-9.
21. Lee SM, Seo JW. Phlebosclerotic colitis: Case report and literature review focused on the radiologic findings in relation to the intake period of toxic material. Jpn J Radiol 2015; 33: 2015-33.
22. Chen MT, Yu SL, Yang TH. A case of phlebosclerotic colitis with involvement of the entire colon. Chang Gung Med J 2010; 33: 581-5.
23. Choi JM, Lee KN, Kim HS, et al. Idiopathic phlebosclerotic colitis: a rare entity of chronic ischemic colitis. Korean J Gastroenterol 2014; 63: 183-6.
24. Tsai CC, Chou JW, Chiang LP, et al. Mesenteric phlebosclerosis. Intern Med 2008; 47: 183-4.

靜脈硬化性結腸炎造成的腹部腫塊：一個病例報告

黃國智

天主教中華聖母修女會醫療財團法人天主教聖馬爾定醫院　內科部腸胃肝膽科

摘　要

靜脈硬化性結腸炎是一種罕見的缺血性腸疾病，目前病理機轉還不確定，它一開始的臨床症狀不明顯，但在一些嚴重的病例有可能致命，這種病例大部分都發生在亞洲人或亞裔人士，主要影響右側結腸，影像學的特殊表徵對這個疾病的診斷佔有重要角色，這裡我們報告一個靜脈硬化性結腸炎的台灣女性患者以腹部腫塊來表現，同時她有服用中藥多年的習慣，腹部放射性影像及大腸鏡的檢查可以幫助臨床上的診斷。