

Case Report

## A Case of Primary Colonic Signet Ring Cell Carcinoma in a Young Man which Preoperatively Mimicked Phlebosclerotic Colitis

Ayako Watanabe<sup>a\*</sup>, Yoshihiko Kadowaki<sup>a</sup>, Kenji Hattori<sup>a</sup>, Mika Ohmori<sup>b</sup>,  
Hiroyuki Tsukayama<sup>a</sup>, Nobuhito Kubota<sup>a</sup>, Tatsuo Okumoto<sup>a</sup>, Nobuhiro Ishido<sup>a</sup>,  
and Takeshi Okino<sup>c</sup>

Departments of <sup>a</sup>Surgery, <sup>b</sup>Radiology, and <sup>c</sup>Pathology, Japanese Red Cross Kobe Hospital, Kobe 651-0073, Japan

A 35-year-old man was referred to our hospital for chronic abdominal pain and diarrhea. Computed tomography showed wall thickening, poor contrast enhancement and calcification of the ascending colon, which were consistent with phlebosclerotic colitis. Malignant character was not detected from a biopsy specimen. Operatively, we observed a scirrhous mass of the ascending colon invading surrounding tissue, which was diagnosed as signet ring cell carcinoma based on analysis of an intraoperative frozen section. Right hemicolectomy with regional lymph node dissection was performed. This case was extremely similar to phlebosclerotic colitis in clinical findings; surgical resection was required for correct diagnosis.

**Key words:** phlebosclerotic colitis, colorectal cancer, signet ring cell carcinoma, young colorectal cancer.

Colorectal signet ring cell carcinoma (SRCC) accounts for approximately 1% of cases of colorectal cancers [1-7]. Compared with common adenocarcinoma (AC), the ratio of younger patients in SRCC is relatively high; however, younger patients still make up, less than 10% of patients with SRCC [7,8]. Phlebosclerotic colitis (PC), which is characterized by sclerosis and calcification of the colonic wall and mesenteric veins, is a rare disease causing chronic ischemic colitis [9,10]. Clinical features in the present case of SRCC, showed findings highly similar to those of PC, such as thickening and calcification of the proximal colon in computed tomography (CT) images. Although both diseases are rare, and have similar clinical features and clinical symptoms, there are no available examinations for differentiating PC from SRCC aside from pathological findings. Thus, strategies should be more considered to rule out SRCC when a colon disease is

diagnosed as PC.

### Case Presentation

A 35-year-old man visited his local hospital after experiencing chronic abdominal pain and diarrhea for more than 6 months. Blood examination showed anemia (Hb 7.1 g/dl), and colonoscopy revealed edematous and ulcerative obstruction of the ascending colon which an endoscope could not pass through. The lesion was suspected of being colon cancer, but no malignancy was detected by a biopsy specimen. Therefore, he was referred to our hospital for further advanced examinations and treatments. He was taking a medication for hypertension, and there were no remarkable findings in his physical examination or family history of cancer. His blood examination including tumor markers showed normal results without anemia.

We first performed contrast enhanced CT imaging.

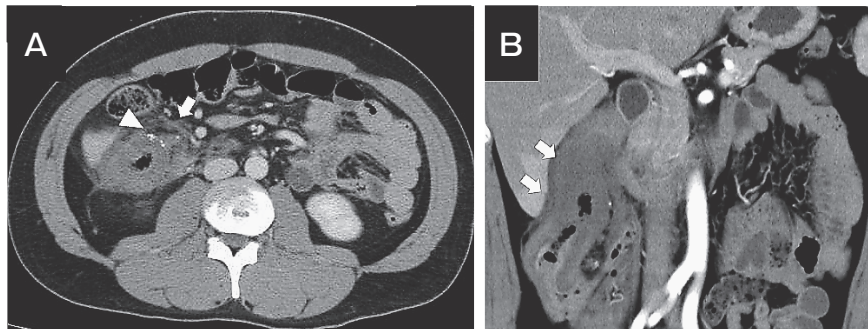
The results showed wall thickening, poor contrast enhancement, and calcification of the ascending colon (Fig. 1A, 1B). The peripheral veins of the ascending colon were not enhanced (Fig. 1A). The thickness of the wall and poor enhancement of the mucous membrane were consistent with characteristics of ischemic colitis. In addition, we observed calcification of the colon and occlusion of the surrounding veins, and we thus suspected PC as a differential diagnosis.

Next, we performed a repeat colonoscopy at our hospital. It showed a dark-purple membrane with loss of visible vascular pattern, but also a submucous elevated lesion with edematous and ulcerative obstruction of the ascending colon which an endoscope was unable to pass through (Fig. 2). These findings were not consistent with characteristics of PC, leading us to further consider suspected type 4 malignant tumor or non-epi-

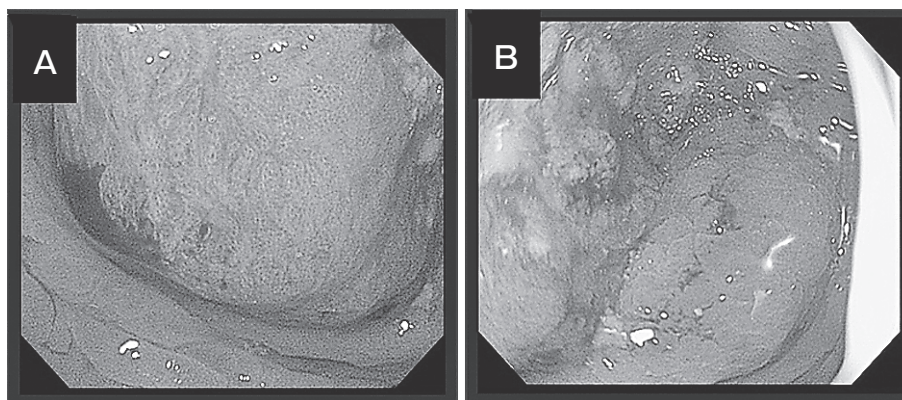
thelial malignant tumor as a differential diagnosis. However, no malignancy was detected on pathological examination of a biopsy specimen at our hospital.

Therefore, we presumed the lesion was PC, type 4 malignant tumor or non-epithelial malignant tumor. Considering his age, the appearance of the lesion, CT images, colonoscopic findings and the biopsy results, we could not reach definitive diagnosis clinically. To relieve his chronic symptoms and ensure a correct diagnosis, a surgical resection was performed.

A scirrhous mass of the ascending colon invading the liver, duodenum, and retroperitoneum was found operatively (Fig. 3), and was strongly suspected of being a malignant tumor. SRCC was detected by an intraoperative biopsy from a chorionic membrane of the ascending colon. The patient underwent right hemicolectomy with regional lymph node dissection with com-



**Fig. 1** Computed tomography images. Contrast enhanced computed tomography showed thickening, poor contrast enhancement (B: arrow) and calcification (A: arrowhead) of the ascending colon. The peripheral veins of the ascending colon were not enhanced (A: arrow).



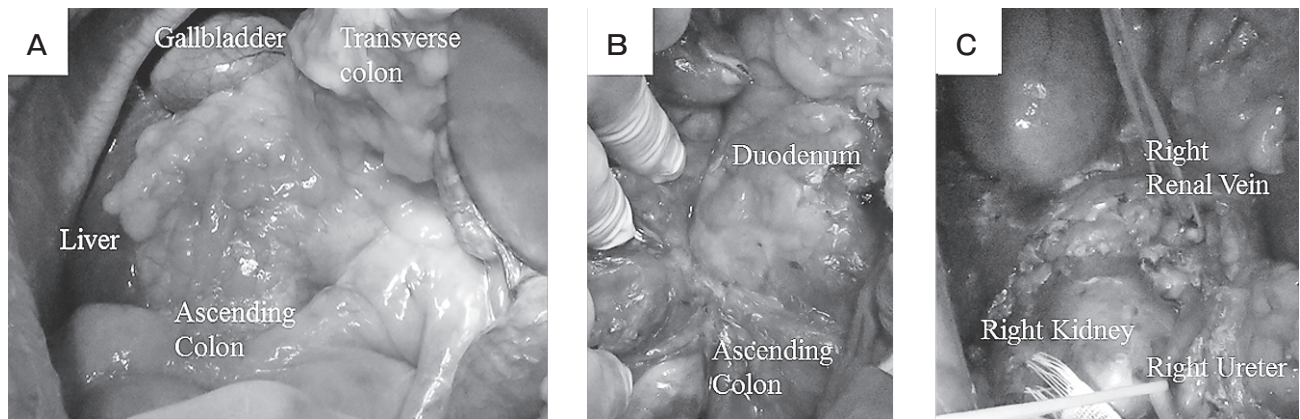
**Fig. 2** Colonoscopic images. Colonoscopy showed a dark-purple membrane and loss of visible vascular pattern, but also presented a submucous elevated lesion with edematous and ulcerative obstruction of the ascending colon (A, B).

bined resection of the invaded liver, duodenum, and retroperitoneum. The operation time was 339 min, and blood loss was 120 ml.

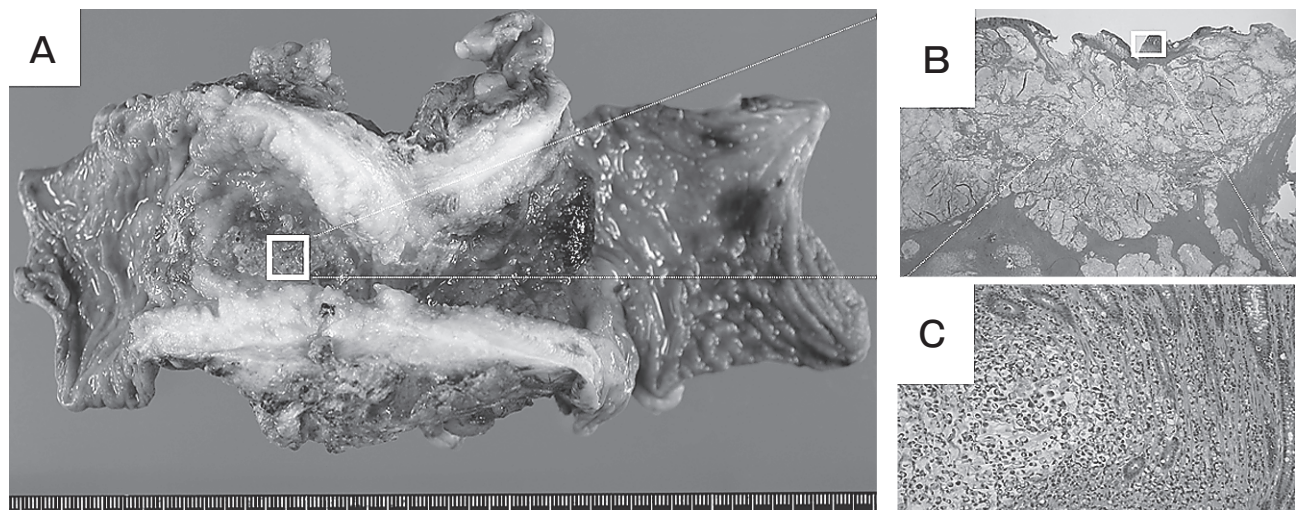
The final pathological examination identified type 4 tumor of the ascending colon and invasive hyperplasty of signet ring cell carcinoma with mucinous components (Fig. 4). The tumor invaded surrounding tissues under the colon serosa, such as the mesentery, retroperitoneum and duodenum, especially pronounced infiltration into the muscularis propria of the duodenum. The lateral surgical margin of the retroperitoneum was slightly positive. The exact distal margin was 6 mm from the extramural invasion of the signet ring

cell carcinoma. There was no evidence of PC pathologically. Eight out of 27 dissected lymph nodes showed metastasis of signet ring cell carcinoma. The tumor was graded as pStage IIIb (pT4bN2M0, Cur B) according to the Japanese Classification of Colorectal Carcinoma, eighth edition [11].

Though the patient needed a nasogastric tube for 5 days for postoperative paralytic ileus, he was discharged on the 11th postoperative day without severe perioperative complications. He started adjuvant chemotherapy using mFOLFOX6 at 6 weeks after surgery. He is doing well without any evidence of recurrence 24 months after surgery.



**Fig. 3** Surgical findings. A scirrhous mass of the ascending colon invaded the liver (A), duodenum (B) and retroperitoneum (C: after resection).



**Fig. 4** Pathological findings. There was a Type 4 tumor of ascending colon (A) and invasive hyperplasty of signet ring cell carcinoma in the mucous membrane and floating in the submucosal mucinous components (B, C).



## Discussion

The first description of PC was reported in Japan in 1991 as a case of ischemic colitis with stenosis of the right colon and sclerosis of mesenteric veins [12]. PC is characterized by calcification of the colonic wall and mesenteric veins and differentiated from other forms of ischemic colitis [9,10,12]. The lesion starts from the right colon and spreads out to the anal side during chronic course [10,13-15]. The etiology and pathogenesis of this rare chronic ischemic colitis have not yet been clearly defined.

The clinical diagnosis of PC is typically based on a combination of clinical symptoms, endoscopic findings, and radiological images; the clinical symptoms are abdominal pain, diarrhea, constipation, hematochezia, and ileus due to intestinal stenosis caused by chronic venous congestion [16]. Colonoscopy findings are rigidity of the colonic wall, luminal narrowing, mucosal edema and ulceration with a dark purple edema [9]. Abdominal CT scanning shows the most typical images of PC, such as thickening of the colonic wall with calcifications along the colonic wall and of veins near the superior mesenteric venous trunk [9]. In the previous reports, CT study has been the most useful diagnostic tool for PC and for evaluating the severity of mesenteric venous calcification [9,17]. The management of PC ranges from conservative care to surgery, based on disease severity. If there are complications such as hemorrhage, bowel obstruction, and perforation, surgical resection should be considered [9,18].

This case was preoperatively suspected as PC since it coincided with the criteria of PC; all clinical features of this disease were presented, especially CT images, such as the thickness, ischemia and calcification of the colorectal wall. However, Li *et al.* [19] reported that SRCC should be considered in cases with CT signs such as long segmental bowel-wall thickening, target enhancement, and peritoneal seeding as atypical features of colorectal carcinoma. They also reported that intratumoral calcification is more frequently seen in mucinous adenocarcinoma. Because our case included mucinous components, we considered that intratumoral calcification may have occurred. Furthermore, no malignancy was detected from a biopsy specimen because the extramural invasion of the signet ring cell carcinoma existed on the anal side of the pathology specimen. These features were so similar as to make the differential diagno-

sis between PC and SRCC and the choice of an optimal treatment strategy impossible without a surgical operation.

The most common subtype of colorectal carcinoma is the differentiated adenocarcinoma; on the other hand, SRCC is found in only about 1% of all colorectal carcinoma [1-7]. As described above, clinical characteristics of SRCC include localization in the proximal colon [1,7,20-22], higher incidence of scirrhous carcinoma [23], younger age at diagnosis [7,8,24] and more advanced stage at presentation [7].

SRCC is diagnosed in more advanced tumor stages, but its dismal prognosis seems to more related to its intrinsic tumor biology [4]. 5-year survival rates for SRCC of the colon and rectum are a relatively poor 30.8% and 19.5%, respectively, compared with 56.8% and 58.5% for AC in these locations [7]. The survival difference between AC and PC was found in stage II, but was most prominent in stage III [7]. In general, for most patients with colorectal cancer, younger age is associated with better survival [8,25-27]. However, younger age is associated with poor survival outcome in SRCC patients [7,24]. This is especially true for stage III SRCC patients younger than 35 years of age, who were shown to have a significantly lower cancer-specific-survival (CSS) than those older than 35 years ( $p=0.008$ ): the 5 year CSS rates were 23.4% and 42.5%, respectively [24].

The patient in the present case had a stage III SRCC and was 35 years of age. Previous reports would tell us to expect a poor prognosis, but he has not had any recurrences 24 months after radical surgical resection followed by adjuvant chemotherapy.

In conclusion, we herein report the case of a young patient with colorectal SRCC which was preoperatively suspected of being PC. Because these diseases have very similar clinical findings, surgical resection should be considered when the differential diagnosis includes both entities.

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