Case Report
pISSN 1738-2637 / eISSN 2288-2928
J Korean Soc Radiol 2016;75(1):12-16
http://dx.doi.org/10.3348/jksr.2016.75.1.12

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CT Findings of Solitary Fibromatosis in the Colon: A Case Report
대장에 생긴 섬유종증의 전산화단층촬영 소견: 증례 보고

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Fibromatosis is a rare benign neoplasm that appears as a sporadic lesion or is found in patients with familial adenomatous polyposis. Fewer than 7 cases of intraabdominal solitary fibromatosis arising from the colon have been reported in the English literature. This small number of reported cases may be not only because of the low incidence of the disease but also because of the difficulty in making proper diagnosis. We present here a case of histologically confirmed intraabdominal solitary fibromatosis arising from the colon, with an emphasis on computed tomography findings.

Index terms
Fibromatosis
Fibromatosis, Abdominal
Colonic Neoplasm
Computed Tomography

Received November 13, 2015
Revised January 5, 2016
Accepted March 11, 2016
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INTRODUCTION

Fibromatosis is a benign tumor that is composed of fibrous tissue without apparent histologic features of malignancy. However, it has significant potential for local invasiveness and recurrence (1). Solitary intestinal fibromatosis is very rare: fewer than 7 cases of intraabdominal solitary fibromatosis arising from the colon have been reported in the English literature.

We report here a case of histologically confirmed intraabdominal solitary fibromatosis arising from the colon with an emphasis on computed tomography (CT) findings. To the best of our knowledge, this is the first report of such a case in Korea.

CASE REPORT

A 43-year-old female presented with a 2-day history of lower abdominal pain. The patient had undergone laparoscopic myomectomy 2 years ago. On physical examination, she was found to have diffuse abdominal tenderness, mainly in the epigastrium, with increased bowel sounds. Laboratory studies revealed a white blood cell count of 15.4 × 10^3/uL and a C-reactive protein level of 1.2 mg/dL. A supine abdominal radiograph showed no gross abnormality. CT with contrast enhancement showed a well-circumscribed homogeneous ovoid-shaped mass in the splenic flexure of the colon, measuring 7 × 5 × 7 cm. The mass showed mild delayed enhancement [pre-contrast: 27 Hounsfield unit (HU), arterial phase: 40 HU, portal phase: 47 HU] (Fig. 1A, B). There was fat infiltration and edematous change in the greater omentum, mesentery, and splenic hilum, suggesting the likelihood of malignancy (Fig. 1C). Its imaging appearance was thought to be like that of a gastrointestinal stromal tumor. Esophagogastroduodenoscopy showed extrinsic compression along the greater curvature of the gastric body. Colonoscopy showed a 5 cm sized elevated lesion with normal mucosa at the splenic flexure of the colon, suggesting a submucosal tumor (Fig. 1D). Biopsy during colonoscopy only revealed colonic mucosa with lymphoid follicle but there was no evidence of malignancy.

On laparoscopic exam, there was a 7 cm sized submucosal mass arising from the splenic flexure of the colon with pericolic adhe-

sion to the stomach, proximal jejunum, splenic hilum, and diaphragm. Laparoscopy-assisted left hemicolectomy, splenectomy, and stomach wedge resection were performed.

Histopathological evaluation of the excised colonic tumor showed a well-circumscribed, yellowish-white, firm and myxoid submucosal mass of the colon, measuring 7.5 × 7 × 4.8 cm in size. The tumor directly extended from the submucosa of the colon (Fig. 1E) to the splenic capsule and gastric proper muscle. The tumor was composed of spindle cell proliferation and finely collagenous stroma (Fig. 1F). Immunochemical stains showed immunopositivity for β-catenin (Fig. 1G) and smooth muscle actin (Fig. 1H), and negativity for the other markers (C-kit, CD34, S-100). The histopathological findings were consistent with deep fibromatosis.

DISCUSSION

Fibromatosis is a benign proliferation of fibrous tissue, characterized by infiltrative growth and frequent tendency to recur locally without metastatic potential. Fibromatoses are broadly categorized as superficial and deep on the basis of their anatomic location. Deep fibromatoses in the abdomen are classified as

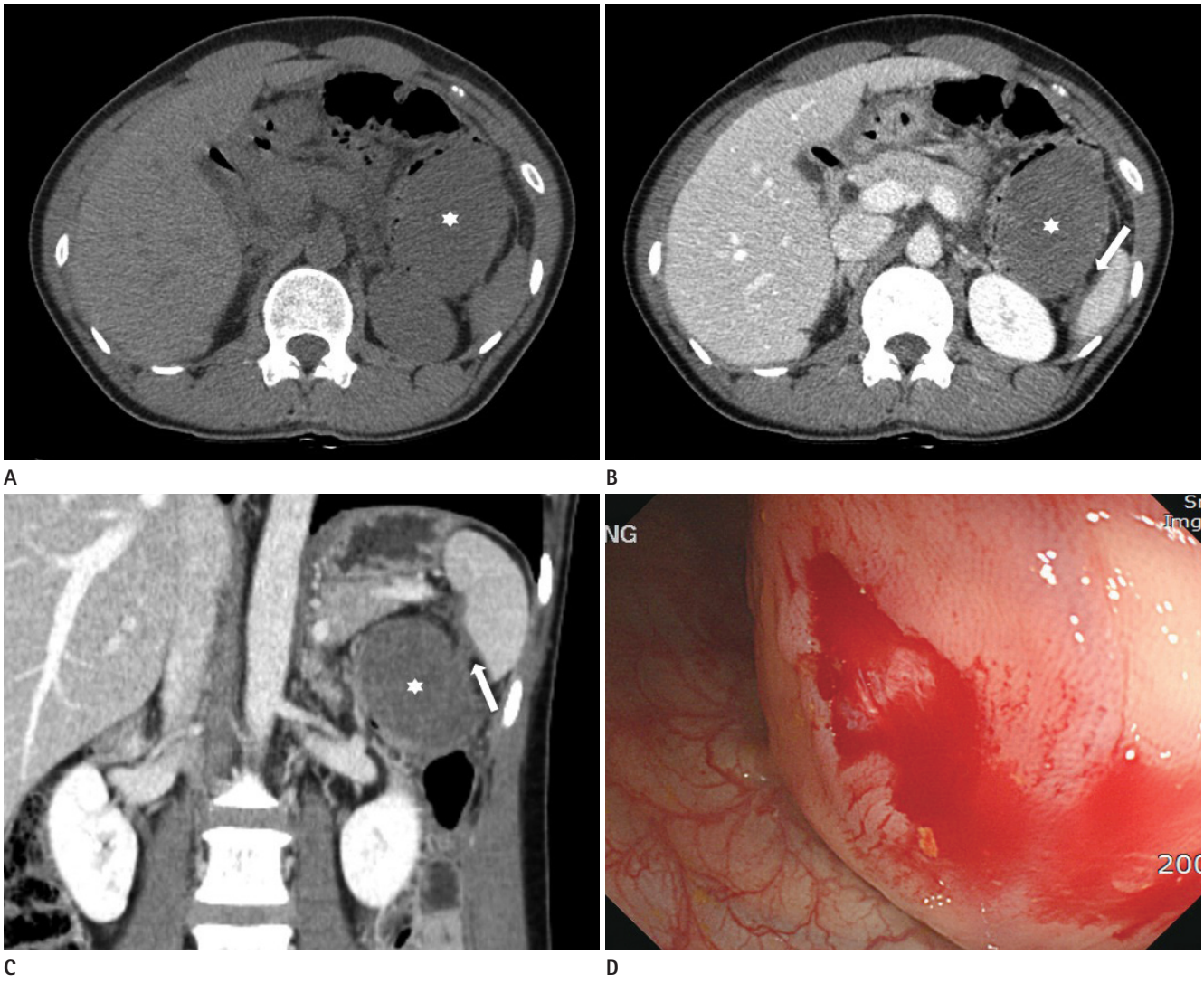

Fig. 1. A 43-year-old female with intraabdominal solitary fibromatosis of the colon.
A. Axial image in the pre-contrast phase shows a 7 × 5 × 7 cm well-circumscribed homogeneous ovoid-shaped mass (asterisk) in the splenic flexure of the colon.
B, C. Axial **(B)** and coronal **(C)** reformatted contrast-enhanced CT images in the portal venous phase show the mildly delayed enhancement of the mass (asterisks). There is fat infiltration and edematous change in the greater omentum, mesentery, and splenic hilum (arrows).
D. Colonoscopy shows a 5 cm sized mass protruding into the lumen, covered with normal mucosa at the splenic flexure of the colon.

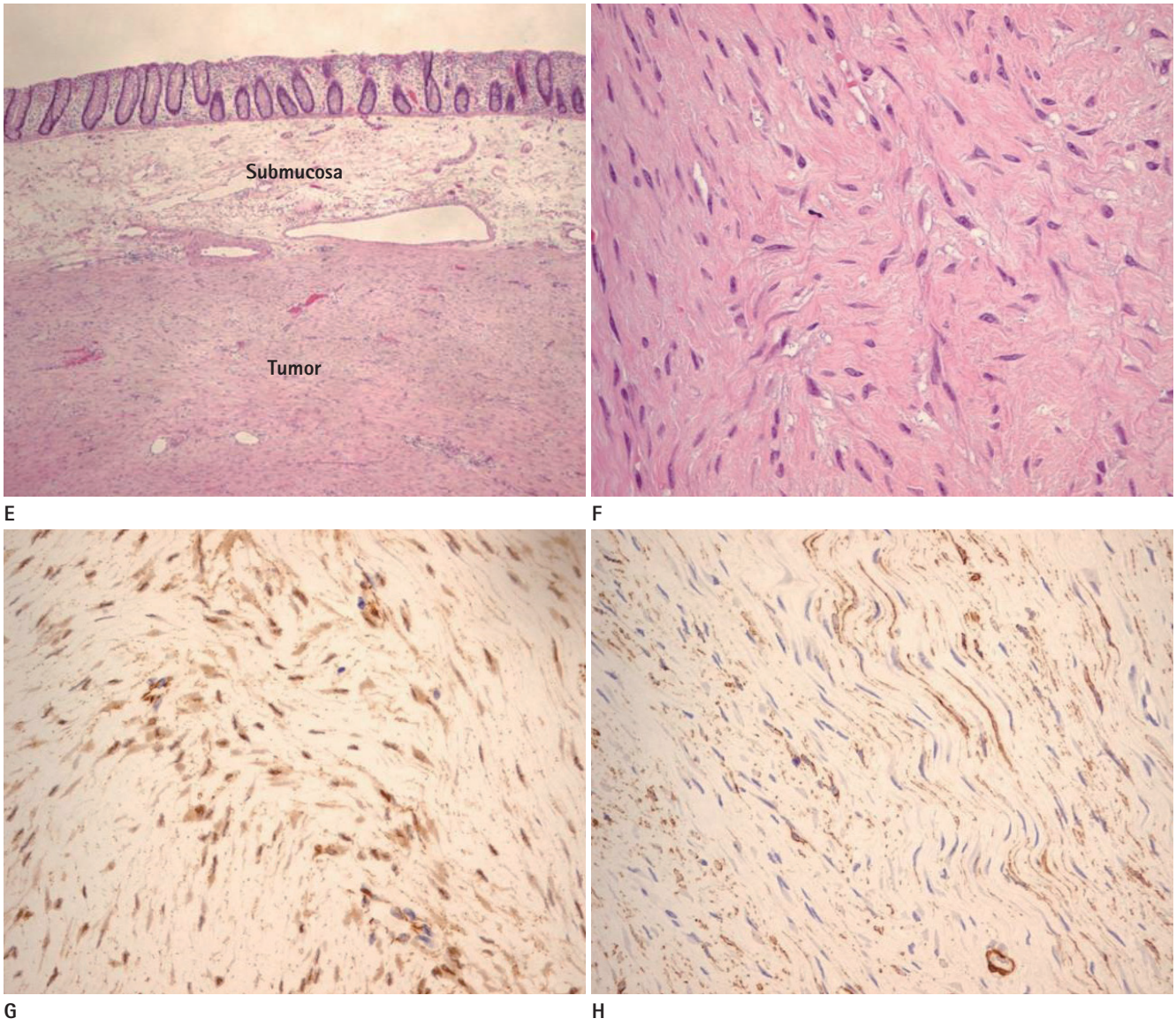

Fig. 1. A 43-year-old female with intraabdominal solitary fibromatosis of the colon.
E, F. Microscopic photomicrograph demonstrates that the tumor involves the submucosa of the colon (**E**, hematoxylin and eosin stain, × 40), and it is composed of fascicles of spindle cells with bland nuclear features and finely collagenous stroma (**F**, hematoxylin and eosin stain, × 400).
G, H. Immunohistochemical stains (× 400) show immunopositivity for β-catenin **(G)** with nuclear accumulation and smooth muscle actin **(H)**.

abdominal wall fibromatosis and intraabdominal fibromatosis (1). Intraabdominal solitary fibromatosis is extremely rare, and only 7 cases have occurred in the colon (Table 1). To the best of our knowledge, this is the first case report in Korea describing the CT findings of intraabdominal solitary fibromatosis arising from the colon. Familiarity with the imaging findings of fibromatosis is useful because complete resection may result in an apparent cure.

The majority of abdominal fibromatoses are sporadic. Among the 8 cases of solitary fibromatosis in the colon, this tumor was more commonly found in females (male:female ratio, 3:5) and the mean age at diagnosis showed a bimodal age distribution (40.5 days and 48.5 years) (2-8).

However, 9–18% of cases of mesenteric fibromatoses have a notable genetic association with familial adenomatous polyposis, specifically Gardner syndrome. Fibromatoses associated with Gardner syndrome are small and multiple and have a female predilection (ratio 3:1), whereas sporadic fibromatoses are generally larger and singular. Fibromatoses associated with Gardner syndrome tend to involve the mesentery and abdominal

Table 1. Reported Cases of Solitary Fibromatosis in the Colon

Authors	Sex/Age	Clinical Presentation	Location	Imaging Findings
Srigley and Mancer (5)*	M/2 days	Abdominal distension	Transverse colon	Abdominal radiography: dilated bowel loops and focal calcifications Barium enema: perforation of the transverse colon
Al-Salem et al. (6)	F/4 days	Intestinal perforation	Descending colon	Abdominal radiography: pneumoperitoneum
Eriguchi et al. (2)	F/38 years	Palpable mass	Transverse colon	CT: a large mass lesion which revealed a slightly lower attenuation than muscle
Lacson et al. (7)	M/5 months	Intestinal obstruction	Splenic flexure	Barium enema: apple core deformity
Numanoglu et al. (8)	M/6 days	Intestinal obstruction	Transverse colon	Abdominal radiography: multiple dilated bowel loops with air fluid levels
Zhu et al. (3)	F/55 years	Abdominal pain	Cecum	CT: about 4 x 3.4 x 3 cm sized ovoid-shape mass with soft tissue density PET-CT: increased FDG activity (SUVmax: 5.4)
Makis et al. (4)	F/58 years	Abdominal pain, hematochezia	Ascending colon	CT: rapidly growing submucosal mass PET-CT: increased FDG activity (SUVmax: from 4.2 to 7.4)

*The number in parenthesis refers to reference number.
CT = computed tomography, FDG = fluorodeoxyglucose, PET = positron emission tomography, SUVmax = maximum standardized uptake value

wall, whereas sporadic fibromatoses occur in the retroperitoneum and pelvis. In addition, Gardner syndrome-associated fibromatoses have a higher rate of recurrence after resection than do sporadic fibromatoses (9).

Imaging findings are variable and reflect the underlying histologic characteristics. On ultrasonogram, a lesion is visualized as a well-circumscribed or ill-defined solid mass with relatively homogenous hypoechogenicity with internal echogenic lines (10). CT is the best modality for establishing the diagnosis, determining the complications, and monitoring the treatment response. On CT, lesions with a highly collagenous stroma may be homogeneous and isoattenuating to muscle. Some lesions may appear striated or whorled when alternating collagenous and myxoid stroma is present. Contrast enhancement is variable; mild to moderate enhancement is typically noted, and delayed enhancement may be seen. Predominantly myxoid lesions do not enhance and remain hypoattenuating (2-4). On magnetic resonance (MR) imaging, most lesions show low to intermediate signal intensity on T1-weighted images; on T2-weighted images, the lesions can have heterogeneous intermediate or high signal intensity, and myxoid stromal elements contribute to high signal intensity. The enhancement pattern on MR imaging is variable, similar to the pattern observed on CT (10). The role of positron emission tomography-CT in evaluating fibromatoses remains undefined. 18F-fluorodeoxyglucose (FDG) uptake has been described in a segment of the colon affected by intussusception caused by solitary intestinal fibromatosis (maximum standardized uptake value = 5.4), although the inflammation associated with the intussusception precluded the assessment of FDG avidity of fibromatosis itself (3).

Differential diagnoses are gastrointestinal stromal tumor, inflammatory pseudotumor, and extrapleural solitary fibrous tumor. Other differential diagnoses include adenocarcinoma, leiomyosarcoma, plasmacytoma, and lymphoma.

The optimal therapeutic regimen for intraabdominal fibromatoses has not yet been determined. Some authors suggest that surgery is the first choice for treating fibromatoses and that the surgical aim is complete resection (10). Other authors recommend surgery only in cases that are complicated by bowel or ureteric obstruction. Some authors have suggested that chemotherapy is effective in aggressive cases of intraabdominal fibromatoses. When wide surgical margins cannot be achieved or when the tumor is not resectable, radiation therapy is generally recommended.

In conclusion, when confronted with a homogeneous submucosal mass arising from the colon that shows delayed mild enhancement, intraabdominal solitary fibromatosis should be included in the differential diagnosis.

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대장에 생긴 섬유종증의 전산화단층촬영 소견: 증례 보고

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섬유종증은 드문 양성 종양으로, 특발성 혹은 가족성 선종성 용종증 환자에서 발생한다. 이 중 대장에서 단일병변으로 기원한 섬유종증은 매우 드물며 7개의 영문 증례만이 보고되었는데 이는 섬유종증의 낮은 발생률뿐 아니라, 정확한 진단의 어려움 때문으로 생각된다. 저자들은 조직학적으로 확진된, 단일병변으로 대장에서 생긴 섬유종증의 증례를 전산화단층촬영 소견을 중점으로 보고하는 바이다.

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