

Isolated Colonic Schwannoma in the Ascending Colon: A Case Report and Literature Review of Schwannomas in the Large Intestine¹

상행결장에 생긴 고립 신경초종: 증례 보고 및 보고된 증례에 대한 검토¹

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Schwannomas are benign mesenchymal spindle cell tumors arising from the Schwann cells that form the peripheral neural sheath. Several recent studies indicate that although reports of gastrointestinal schwannomas have increased with advanced technological developments in immunohistochemical staining, isolated colonic schwannomas are extremely rare. Moreover, it is known to be somewhat difficult to diagnose colonic schwannoma before surgical operation. In this paper, we report a case of isolated schwannoma that was incidentally discovered in the ascending colon, along with a review of few recent literatures.

Index terms

Schwannoma
Colon
Computed Tomography

INTRODUCTION

Schwannomas are mostly benign neoplasms arising from the Schwann cells that form the neural sheath. Schwannomas of the gastrointestinal tract are relatively rare and are known to be separate entities from conventional schwannomas which originate in soft tissue or the central nervous system (1). Of all submucosal tumors in the gastrointestinal tract, only 2% to 6% have been reported as schwannomas (2). Most gastrointestinal schwannomas occur in the stomach. Schwannomas in the colon are quite rare. Here, we introduce a case of isolated colonic schwannoma and review the recent literature.

CASE REPORT

A 79-year-old woman was referred to our colorectal outpatient department for further evaluation and management of an incidentally discovered mass in the ascending colon. She had a medi-

cal history of hypertension treated with medication for twenty years. On physical examination, there were no specific findings in her abdomen. Laboratory tests including routine complete blood count, liver function test, and urine analysis were within normal ranges. A colonoscopy was performed and a 4 cm sized intraluminal protruding submucosal tumor, about 70 cm near the anal verge, was found in her ascending colon (Fig. 1). Subsequent biopsy was performed and the pathology showed granulation tissue formation with suppurative inflammation. Multidetector computed tomography (CT) of the abdomen was performed. The pre-contrast image showed a well-defined isodense mass in the ascending colon (Fig. 2A). There was no calcification. On the contrast-enhanced image, the mass showed homogenous enhancement and thin enhancement of intact colonic mucosa was noted (Fig. 2B, C). There were several small perilesional lymph nodes along the ileocolic vessel. The preoperative impression was a submucosal tumor such as neurogenic tumor, leiomyoma, or lymphoma. Her serum carcinoembryonic antigen was 1.4 ng/mL.

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Our surgeon performed laparoscopic assisted right hemicolectomy with side to side ileocolic anastomosis. Gross examination of the ascending colon revealed a yellowish gray fungating mass measuring 4.1×3.4 cm (Fig. 3). On microscopic evaluation, the tumor was composed of plump spindle cells with an occasional fascicular pattern. The tumor cells had normochromatic elongated nuclei without significant nuclear atypia. Mitoses were present in 2/50 high power fields but no atypical forms were identified. Immunohistochemical staining was strongly positive for S-100 protein, but negative for CD34 and CD117 (Figs. 4, 5). The patient was diagnosed with a colonic schwannoma. Thirty-one regional lymph nodes were resected and the pathology revealed reactive hyperplasia. The patient's postoperative course was uneventful and there was no recurrence or metastasis after one year of follow-up.



Fig. 1. A colonoscopy reveals an intraluminal round submucosal tumor, which is measured approximately 4 cm in diameter, in the ascending colon.

DISCUSSION

Although the number of reports of gastrointestinal schwannomas has increased with advances in immunohistochemical staining, primary gastrointestinal schwannomas are still rare tumors (2). In the gastrointestinal tract, this tumor mostly occurs in the stomach. Colonic schwannomas are quite rare, comprising 6.1% of gastrointestinal schwannomas reported in previous studies (3). There is no gender predilection in the incidence of colonic schwannomas and they most commonly appear after the sixth decade of life (4). Although usually asymptomatic, colonic schwannomas may cause clinical symptoms such as bleeding, constipation, and anal pain depending on the size and location (2, 5).

Imaging diagnosis of schwannoma before surgery is not easy. According to a previous review of colonic schwannomas in Japan, only 4 of 41 cases (10%) were correctly diagnosed as schwannoma preoperatively (6). Preoperative imaging can be helpful to define the exact tumor location and obtain information about the tumor. However, imaging cannot provide detailed information about benign or malignant features of the tumor and there is overlap in imaging features between schwannomas and other submucosal tumors. On CT, schwannomas usually appear as well defined, round, mural masses with homogeneous enhancement and no evidence of cystic changes, necrosis, hemorrhage or calcification (1). However, small gastrointestinal stromal tumors (GISTs) with low malignant risk may present CT findings similar to schwannomas (1). It has been reported that perilesional lymphadenopathy are more often observed in gastric schwannomas than in gastric GIST (7). Although it is not certain, these lymph nodes are presumed to be reactive hyperplasia due to the fact that

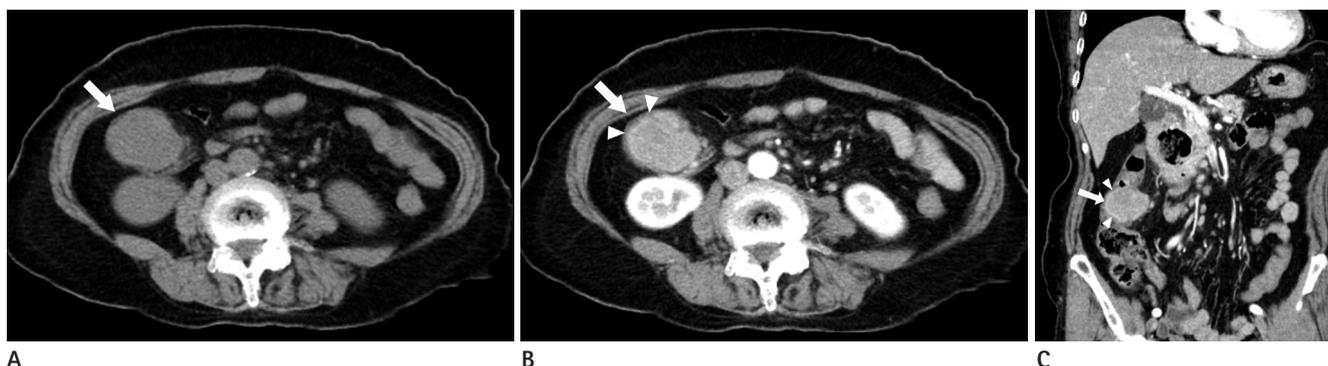


Fig. 2. Pre-contrast (A) and contrast enhanced axial (B) and coronal (C) CT scan demonstrates a lobulated contoured mass (arrow) in the ascending colon. On pre-contrast axial CT scan (A), there is no calcification or cystic component. Enhanced axial (B) and coronal reformatted (C) CT images show homogeneous enhancement of the mass and intact overlying mucosa (arrowheads).

gastric schwannomas include many inflammatory cells (7). According to the report, perilesional lymphadenopathy may be helpful for differentiation between gastrointestinal schwannoma

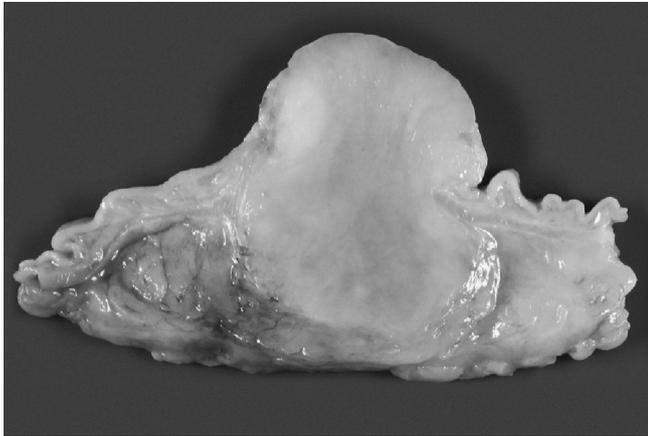


Fig. 3. Gross examination of the ascending colon revealed a yellowish gray fungating mass measuring 4.1 × 3.4 cm.

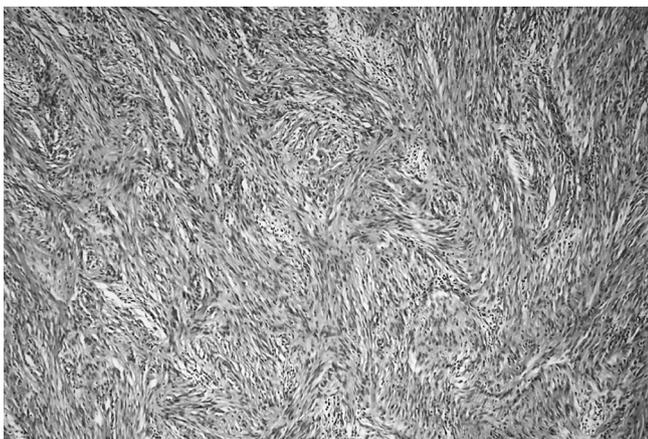


Fig. 4. Photomicrography demonstrates proliferation of plump spindle cells with occasional fascicular pattern, lacking significant nuclear atypia (hematoxylin and eosin, × 100).

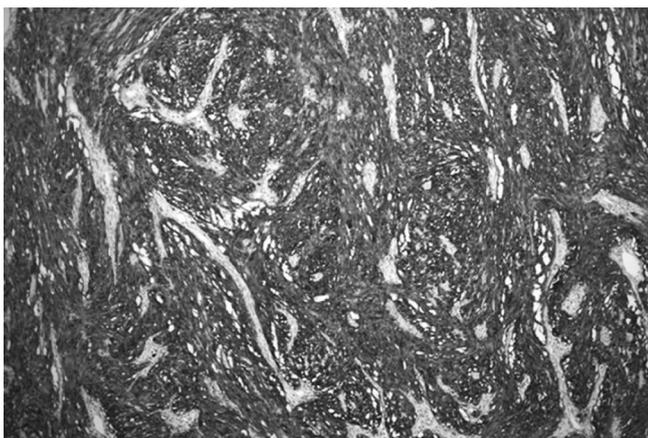


Fig. 5. Immunohistochemical staining for S-100 protein is diffuse and strong (× 100).

and GIST. Primary or secondary lymphomas also have CT findings similar to schwannomas because they show homogenous enhancement before treatment. Accurate preoperative diagnosis of schwannoma is not easy, although the degree of lymph node enlargement can be helpful for differentiating schwannomas from lymphomas because lymphomas are commonly associated with significant lymph node enlargement in the mesentery and retroperitoneum (1). Therefore, preoperative histological examination is usually performed.

Gastrointestinal schwannomas may share gross morphological features with other submucosal tumors. Endoscopic biopsy does not always provide an accurate diagnosis because it is difficult to obtain sufficient biopsy tissue volume in some cases due to tumor hardness (2). In our patient, the result of endoscopic biopsy was granulation tissue formation with suppurative inflammation, which was not helpful for diagnosis. Large forceps biopsy or endoscopic ultrasound-guided biopsy may improve the diagnostic yield (6). For definite diagnosis, immunohistochemistry is usually necessary. Characteristically, schwannomas are composed of spindle cells showing strong positivity for S-100 protein and vimentin, and variable positivity for glial fibrillary acidic protein, while showing negativity for CD34 and CD117. These patterns allow for differentiation of schwannomas from GISTs, gastrointestinal autonomic nerve tumors, and leiomyomas (2, 3).

There are fourteen case reports of colonic schwannomas written in English or Korean and these reports provide accessible information about imaging findings (6, 8-19). We reviewed these cases as well as ours (Table 1). The mean age of patients was 61.7 years (range, 49–79 years) and the male to female ratio was 1:2. The mean size of tumors was 3.6 ± 2.1 cm. Most of the patients had no symptoms and the tumor was detected on screening colonoscopy. In 6 patients (40%, 6/15), the tumor was located in the ascending colon. Most of the cases showed homogenous enhancement on the preoperative CT. A definite preoperative imaging diagnosis was not made in most cases, but instead, nonspecific submucosal tumor was the most common preoperative diagnosis. Endoscopic biopsy was performed in three patients, but only one patient (16) was accurately diagnosed with schwannoma after multiple colonic biopsies. All the patients underwent surgical resection for treatment.

In general, gastrointestinal schwannomas follow a benign clinical course. Once a schwannoma is diagnosed preoperatively,

Table 1. Summary of Cases with Colonic Schwannoma

No.	Author	Year	Age/ Sex	Symptoms	Location	Size (cm)	Preoperative CT Finding	Lymph Nodes Enlargement	Endoscopic Biopsy	Treatment
1	Tomozawa et al.	1998	66/M	None	Cecum	3.5	High density mass	No	Neurogenic tumor	Ileocecal resection
2	Lee et al.	2006	61/F	None	Rectum	4	Homogenous enhancing mass	N/A	-	Transanal excision
3	Kang et al.	2007	58/M	None	Transverse colon	3.2	Homogenous enhancing mass	No	-	Wedge resection
4	Min et al.	2007	49/F	RLQ pain	Ascending colon	4	Homogenous enhancing mass	Several	Inflammation	Hemicolectomy
5	Tsunoda et al.	2009	67/F	None	Transverse colon	3	Irregular mass	Several	Granulated tissue indicating blood vessel proliferation, inflammatory cell infiltration and fibrin deposition	Transverse colectomy
6	Hsu et al.	2009	65/M	Diarrhea	Ascending colon	3.1	Homogenous enhancing mass	No	Necrotizing inflammation	Hemicolectomy
7-1	Park et al.	2010	52/F	None	Ascending colon	1.8	Homogenous enhancing mass	No	-	Laparoscopic hemicolectomy
7-2	Park et al.	2010	59/F	None	Descending colon	1.4	Homogenous enhancing nodule	No	-	Laparoscopic hemicolectomy
8	Matsumoto et al.	2011	59/F	None	Cecum	2	Homogenous enhancing mass	N/A	No abnormality	Laparoscopic wedge resection
9	Vasilakaki et al.	2012	68/M	Hematochezia	Ascending colon	4	High density enhancing lesion	No	Adenocarcinoma	Ileo-hemicolectomy
10	Kim et al.	2012	61/M	None	Ascending colon	1.7	Homogenous enhancing mass	No	Schwannoma	Laparoscopic hemicolectomy
11	Baek et al.	2013	70/F	None	Ascending colon	1.3	Homogenous low attenuation lesion	No	-	Laparoscopic wedge resection
12	de Mesquita Neto et al.	2013	78/F	RLQ pain	Cecum	10	Homogenous and minimal enhancing mass	N/A	-	Laparotomy
13	Jung et al.	2014	59/F	Hematochezia	Descending colon	4.8	Intussusception	Several	-	Hemicolectomy
14	Current case	2014	79/F	None	Ascending colon	4.1	Homogenous enhancing mass	Several	Granulation tissue with inflammation	Laparoscopic hemicolectomy

N/A = not available, RLQ = right lower quadrant, - = not performed

minimal invasive treatment such as endoscopic resection (2) may be considered instead of aggressive surgery. Therefore, efforts to diagnose schwannomas preoperatively are very meaningful. On CT, homogenous enhancement patterns may be helpful for differentiating schwannomas from other submucosal tumors such as GISTs. Multiple endoscopic biopsies or endoscopic ultrasound-guided biopsy can be very helpful in improving diagnostic yield.

In conclusion, although schwannoma in the colon is a rare type of submucosal tumor, preoperative diagnosis is important for a proper surgical plan. When a colonic submucosal tumor with homogenous enhancement is observed, efforts for preoperative diagnosis such as multiple endoscopic biopsy or ultrasound-guided biopsy should be considered under the radiologic impression of schwannoma.

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상행결장에 생긴 고립 신경초종: 증례 보고 및 보고된 증례에 대한 검토¹

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신경초종은 말초신경 다발에서 신경집을 만드는 슈반세포에서 기원하며 대부분 양성 증배염성 종양이다. 신경초종은 위장관을 포함하여 인체의 어디서든 생길 수 있다. 최근의 문헌에 따르면 면역형광조직염색의 발달에 힘입어 위장관에 생기는 신경초종의 발견 보고가 증가하고 있지만, 결장에 생기는 원발성 신경초종은 매우 드물다. 게다가 이러한 신경초종은 수술 이전에 진단하는 것이 어려운 것으로 알려져 있다. 상행결장에서 우연히 발견되어 수술로 확진된 고립 신경초종 1예를 최근 문헌에 대한 고찰과 함께 보고한다.

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