

CASE REPORT

Isolated primary schwannoma arising on the colon: report of two cases and review of the literature

Ki Jae Park, Ki Han Kim, Young Hoon Roh, Sung Heun Kim, Jong-Hoon Lee¹, Seo-Hee Rha², Hong Jo Choi

Departments of Surgery, ¹Internal Medicine, and ²Pathology, Dong-A University College of Medicine, Busan, Korea

Primary schwannoma of the large intestine is an extremely rare neoplasm. Here, we report two cases of colonic schwannoma confirmed pathologically after laparoscopic resection. A 52-year-old female and a 59-year-old female were referred by their general practitioners to our coloproctologic clinic for further evaluation and management of colonic submucosal masses. Colonoscopies performed in our institution revealed round submucosal tumors with a smooth and intact mucosa in the mid-ascending and descending colon, respectively. Computed tomography (CT) scans showed an enhancing soft tissue mass measuring 2 × 2 cm in the right colon and well-defined soft tissue nodule measuring 1.5 × 1.7 cm in the proximal descending colon, respectively. We performed laparoscopic right hemicolectomy and segmental left colectomy under the preoperative impression of gastrointestinal stromal tumors. Two cases were both diagnosed to be benign schwannoma of the colon after immunohistochemical stains (S-100 (+), smooth muscle actin (-), CD117 (-), and CD34 (-)).

Key Words: Schwannoma, Colon, Immunohistochemical stain

INTRODUCTION

Schwannomas (cellular neurilemmomas) are predominantly benign tumors derived from the Schwann cells that form the neural sheath. The incidence of submucosal schwannoma has been estimated at 2 to 6 percent of all submucosal tumors of the intestine [1]. The largest published series documents that the most frequent site of primary gastrointestinal schwannoma is the stomach [1]. Isolated schwannomas of the colon without associated neurofibromatosis are extremely rare [1,2]. Here, we present two cases of laparoscopically resected schwannoma of the colon together with a review of the literature.

CASE REPORTS

Case 1

A 52-year-old woman came to our coloproctologic clinic for further evaluation and management of a right colonic mass. Routine colonoscopy for a health examination led to the discovery of a 1.3 cm-sized submucosal mass in the mid-ascending colon. At the follow-up colonoscopy five months later, the size of the mass was increased to 2 cm (Fig. 1A). The lesion was thought to represent a gastrointestinal stromal tumor (GIST) or lipoma. The patient's previous medical history stated hypertension under medical treatment. There was no family history of neurofibro-

Received December 17, 2009, Accepted March 2, 2010

Correspondence to: Hong Jo Choi
Department of Surgery, Dong-A University College of Medicine, 1 Dongdaesin-dong 3-ga, Seo-gu, Busan 602-714, Korea
Tel: +82-51-240-5146, Fax: +82-51-247-9316, E-mail: colonch@donga.ac.kr

© Journal of the Korean Surgical Society is an Open Access Journal. All articles are distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/3.0/>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

matosis.

Upon physical examination, neither the patient's abdomen nor other systems showed any positive findings, nor did subcutaneous nodules. Laboratory findings were nonspecific. Contrast-enhanced computed tomography

(CT) scan showed a well-defined homogeneously enhancing submucosal mass in the posterior wall of the cecum (Fig. 2A, B). The preoperative impression was a submucosal tumor such as a GIST.

We performed a laparoscopic right hemicolectomy. The

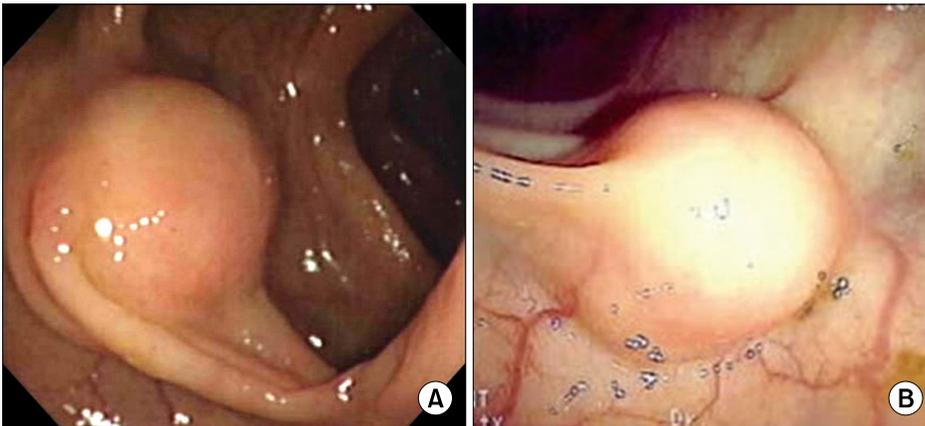


Fig. 1. Colonoscopic findings of both cases showing submucosal tumors measuring approximately 2 cm (A) and 1.5 cm (B) in diameter.

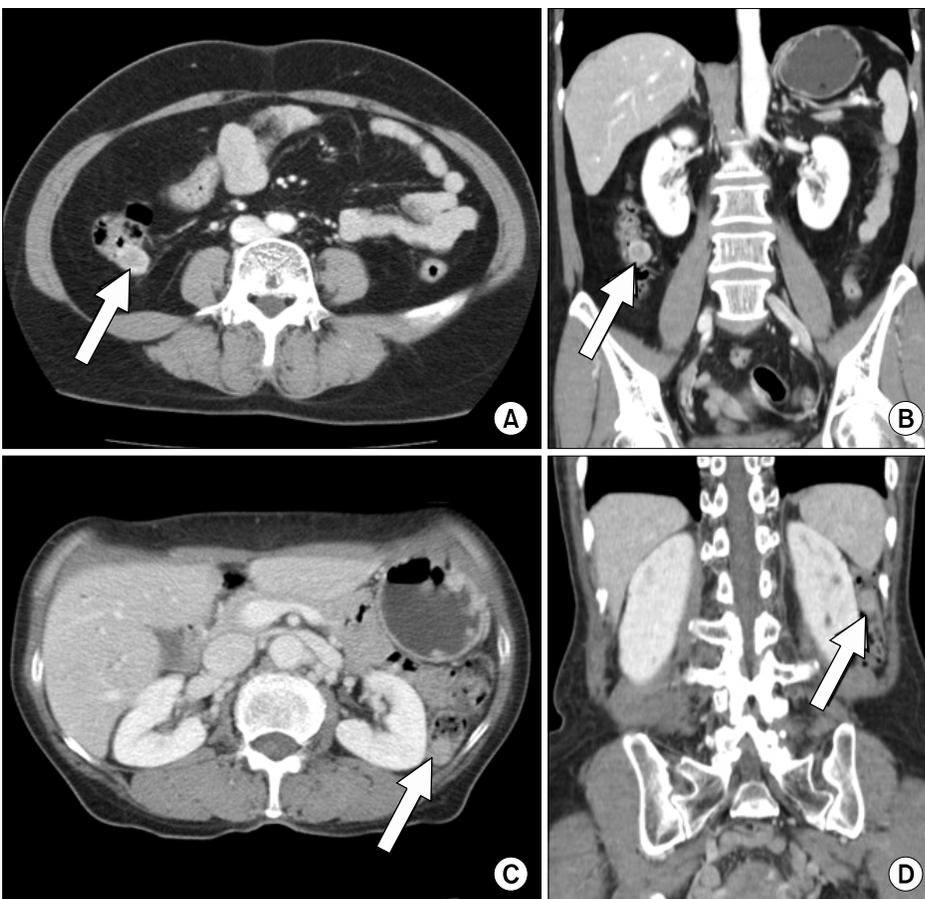


Fig. 2. Computed tomography scans. (A, B) Images of the first case show a well-defined homogeneously enhancing submucosal mass in the posterior wall of the cecum (arrow). (C, D) A well-defined soft tissue nodule can be seen in the proximal descending colon (arrow).

end-to-side ileocolic anastomosis was performed by using a biofragmentable anastomosis ring (Valtrac, United States Surgical Co., Norwalk, CT, USA), extracorporeally. Macroscopically, the ascending colon bore a well circumscribed mass measuring 1.8×1.6 cm. The proximal and distal margin were 5.7 cm and 15 cm, respectively. The mass was intramurally located, with a whitish fibrotic and firm cut surface, and the overlying mucosa was intact (Fig. 3A). Histologically, the mass was composed of spindle cells. The tumor cells were uniform and showed few mitotic figures (Fig. 3B, C). Immunohistochemical assays demonstrated lack of reactivity to CD117 (KIT), CD34, and

smooth muscle actin (SMA), with a diffusely positive cytoplasmic reactivity to S-100 protein (Fig. 3D), definitively confirming the tumor as a schwannoma. The transient ileus was developed at postoperative 7 days but completely resolved after conservative management. At 18 months follow-up, the patient is doing well without any evidence of recurrence.

Case 2

A 59-year-old woman referred to our clinic with a descending colonic submucosal mass (about 1.5 cm in size) which had been detected on the screening colonoscopy for

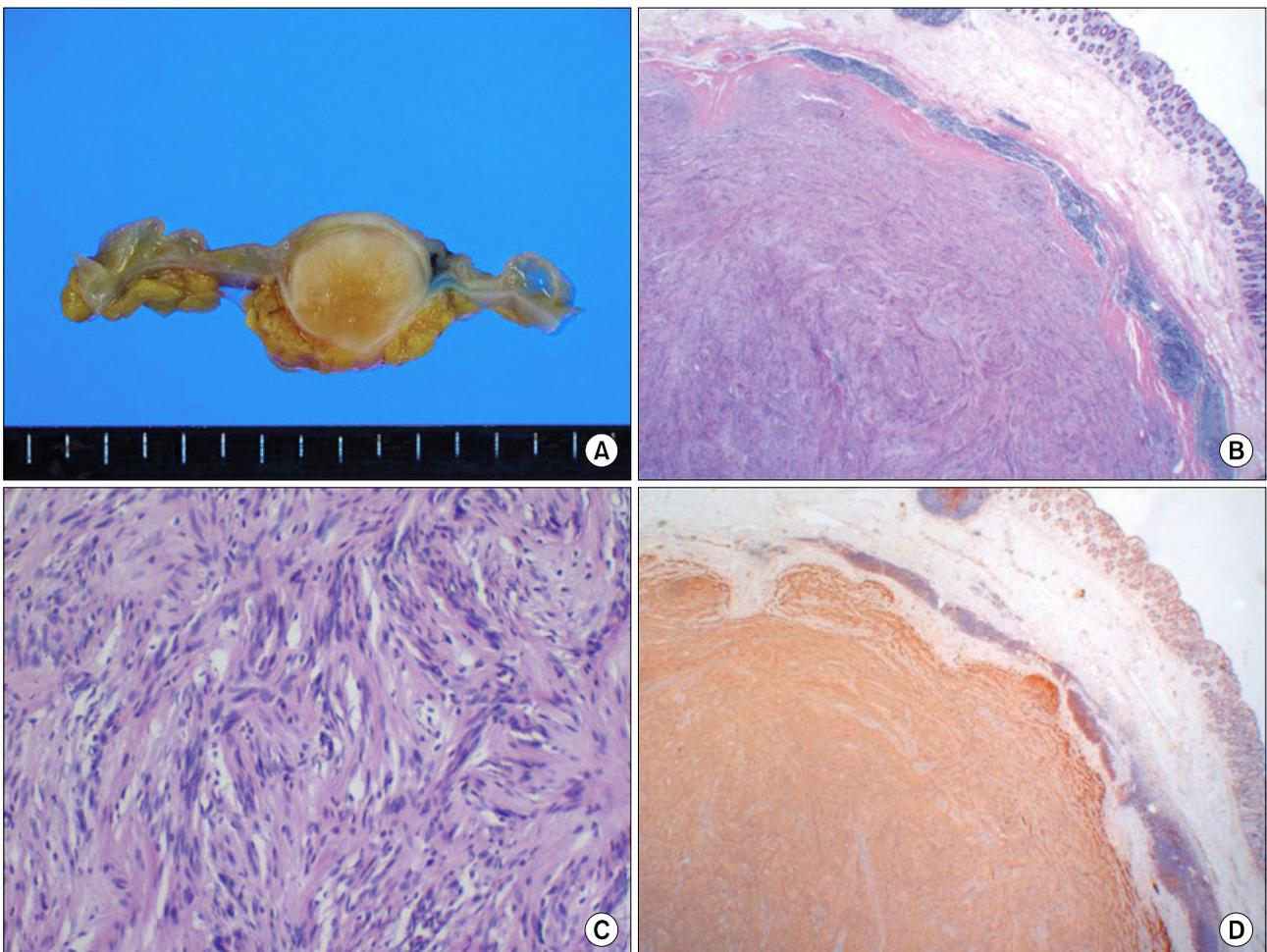


Fig. 3. Pathological examinations. (A) Ascending colon shows a well-circumscribed mass, measuring 1.8×1.6 cm. The mass is intramurally located and shows whitish fibrotic and firm cut surface without definitive hemorrhage or necrosis. The covering mucosa is intact. (B) The well-circumscribed intramural mass is surrounded by lymphocytic rim (H&E, $\times 10$). (C) The mass is composed of whorled arranged spindle cells and the tumor cells are uniform and show few mitotic figures (H&E, $\times 100$). (D) The tumor shows a diffuse cytoplasmic immunoreactivity for S-100 protein ($\times 10$).

a health examination (Fig. 1B). Her medical history and all findings on physical examination were unremarkable. Contrast-enhanced CT scan showed a well defined soft tissue nodule measuring 1.5 × 1.7 cm in size in the proximal descending colon (Fig. 2C, D).

We performed a laparoscopic segmental left colectomy under the diagnosis of a submucosal tumor such as a GIST. The end-to-end colocolic anastomosis was extracorporeally performed by using a biofragmentable anastomosis ring. Macroscopically, the mucosal surface of the mass is unremarkable. On section, the muscle layer shows a well demarcated oval mass, measuring 1.4 × 1.2 cm in dimension. The proximal and distal margin were 6.5 cm and 3 cm, respectively. The mass showed yellowish gray granular cut surface. Immunohistochemical assays demonstrated that the tumor cells were strongly positive for S-100 protein, whereas CD117, CD34, and SMA were consistently negative. As a consequence of these pathological findings, the tumor was diagnosed to be a schwannoma. The postoperative course was uneventful. At 6 months after surgery, she was free of recurrence.

DISCUSSION

Schwannoma is a rare neoplasm that originates from Schwann cells and which commonly occurs in the peripheral nerve of the limbs or body, spinal cord, and central nervous system. When it occurs in the gastrointestinal tract (GIT), the colon is the least likely site [2]. The stomach has been reported to be the most common site of GIT involvement [1,3].

Although the exact incidence of colonic schwannoma is unknown, some studies have estimated the incidence indirectly. Schwannomas are vastly less common than GISTs, being outnumbered by approximately 50-100:1 [4]. Hou et al. [3] determined that benign schwannomas represented 2.9% of gastrointestinal mesenchymal tumors in their investigations. Another study similarly identified twenty (3.3%) colorectal schwannomas among 600 mesenchymal tumors of the colon and rectum [5]. Inagawa et al. [6] reviewed reports of benign schwannomas arising from the colon and rectum in a Japanese literature search and

found rectum (45.7%) to be the most frequent site of occurrence. They also calculated the incidence of schwannoma arising from the right colon, excluding the appendix, at 19.6% (9 cases among forty-six) of cases of colon and rectal schwannomas.

The extremely rare incidence of colonic schwannoma may be explained at least in part in light of the result of Hou et al. In their review of 25 cases of schwannoma, only twelve (48%) were primarily diagnosed as schwannomas; the other thirteen cases were originally diagnosed as leiomyoma, cellular leiomyoma, leiomyosarcoma or neurofibroma [3]. Studies in 1988 and 2001 by two different groups defined and fully documented the morphological and phenotypic characteristics of digestive schwannomas, which included a prominent lymphoid cuff with or without germinal center formation [1,5]. Schwannomas of the GIT evaluated on a purely morphologic basis can be misdiagnosed as GISTs with neural differentiation [2,3]. Owing to advances in immunohistochemical staining, however, the number of reports of intestinal schwannoma has gradually increased, and its differential diagnosis from other spindle cell tumors such as leiomyoma can now be clearly made [3,6].

The most important immunohistochemical markers for diagnosis of schwannoma are S-100 protein, CD117 (KIT) and CD34. Cells from neurofibromas are 30 to 40% immunoreactive with S-100 protein, in contrast to 100% immunoreactivity in schwannomas [4]. In addition, schwannomas are consistently negative for CD117 and are usually negative for CD34, while in contrast GISTs exhibit high expression of CD117 and CD34 (70%) but are negative for S-100 protein [4,5]. A combinatorial assay of S-100, CD34 and CD117 proteins, therefore, is definitive in differentiating schwannoma from GISTs. Glial fibrillary acidic protein (GFAP) serves as another useful marker in immunohistochemical studies, allowing differentiation of schwannomas from gastrointestinal autonomic nerve tumors (GANTs). Schwannomas of the GIT express GFAP (63.6%), whereas GANTs do not express GFAP [3].

We made the preoperative diagnoses of both cases as submucosal tumor such as GISTs. Inagawa et al. [6] demonstrated that the rate of accurate preoperative diagnosis for benign schwannomas was only 15.2% (seven of for-

ty-six cases) and that the most common preoperative diagnosis was in fact submucosal tumor (39.1%). The reasons for inaccurate preoperative diagnosis can include an insufficient volume of biopsy specimen for pathologic confirmation and a limited knowledge of such tumors. Levy et al. [7] described schwannomas evaluated by CT as homogeneously attenuating, well-defined, mural masses. These CT findings were also observed in our case. Contrary to these CT findings of gastrointestinal schwannomas, most of GISTs show a much more heterogeneous appearance resulted from the presence of hemorrhage, necrosis, and cystic change [7].

We did not have a preoperative diagnosis of schwannoma and could not confirm as being benign tumor. So, we performed right hemicolectomy and segmental left colectomy on oncologic basis, laparoscopically. Although surgical resection with adequate margins is the treatment of choice, the question still persists as to whether or not radical resection according to oncologic principles should be performed for all GIT schwannomas. Controversy over the optimal extent of resection for these cases arises partly from the rarity of the disease itself and partly from confusion between schwannomas and GANTs. Some authors have proposed that radical resection is necessary for the management of schwannoma, but they seem to have considered schwannomas as merely a subset of GANTs [8]. Although both schwannomas and GANTs originate from nerve tissue, however, their clinical courses are quite different. While GANTs are invariably accompanied by aggressive clinical behavior [9], digestive schwannomas generally pursue a benign course regardless of the highly variable extent of resection employed for their treatment, ranging from simple enucleation to radical operations [1,3,6]. Braumann et al. [10] suggested that in the absence of adequate preoperative diagnosis, the extent of excision should be determined by the characteristics of the tumor such as size, location, and consistency. Needless to say, radical excision according to oncologic principles is warranted for an optimal outcome when the tumor shows a higher aggressiveness on pathologic result. However, the complete local excision with a sufficient surgical margin is generally considered to be a correct treatment of colonic schwannomas because the risk of malignant trans-

formation is extremely low [3,6].

In summary, because of the uncertainties inherent in traditional methods of diagnosis, we strongly recommend the use of immunohistochemical assays in order to differentiate schwannomas from other types of mesenchymal tumors such as GISTs and GANTs. Correct preoperative diagnosis, in addition of course to the operative findings or characteristics of the tumor itself, can be important in governing the extent of resection in digestive schwannoma.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Daimaru Y, Kido H, Hashimoto H, Enjoji M. Benign schwannoma of the gastrointestinal tract: a clinicopathologic and immunohistochemical study. *Hum Pathol* 1988; 19:257-64.
2. Skopelitou AS, Mylonakis EP, Charchanti AV, Kappas AM. Cellular neurilemoma (schwannoma) of the descending colon mimicking carcinoma: report of a case. *Dis Colon Rectum* 1998;41:1193-6.
3. Hou YY, Tan YS, Xu JF, Wang XN, Lu SH, Ji Y, et al. Schwannoma of the gastrointestinal tract: a clinicopathological, immunohistochemical and ultrastructural study of 33 cases. *Histopathology* 2006;48:536-45.
4. Miettinen M, Lasota J. Gastrointestinal stromal tumors--definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. *Virchows Arch* 2001;438:1-12.
5. Miettinen M, Shekitka KM, Sobin LH. Schwannomas in the colon and rectum: a clinicopathologic and immunohistochemical study of 20 cases. *Am J Surg Pathol* 2001;25: 846-55.
6. Inagawa S, Hori M, Shimazaki J, Matsumoto S, Ishii H, Itabashi M, et al. Solitary schwannoma of the colon: report of two cases. *Surg Today* 2001;31:833-8.
7. Levy AD, Quiles AM, Miettinen M, Sobin LH. Gastrointestinal schwannomas: CT features with clinicopathologic correlation. *AJR Am J Roentgenol* 2005;184:797-802.
8. Mulchandani MH, Chattopadhyay D, Obafunwa JO, Joypaul VB. Gastrointestinal autonomic nerve tumours--report of a case and review of literature. *World J Surg Oncol* 2005;3:46.

9. Lauwers GY, Erlandson RA, Casper ES, Brennan MF, Woodruff JM. Gastrointestinal autonomic nerve tumors. A clinicopathological, immunohistochemical, and ultrastructural study of 12 cases. *Am J Surg Pathol* 1993;17: 887-97.
10. Braumann C, Guenther N, Menenakos C, Junghans T. Schwannoma of the colon mimicking carcinoma: a case report and literature review. *Int J Colorectal Dis* 2007;22: 1547-8.