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Schwannoma of the Colon, a Rare Tumor Entity: A Case Report

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Abstract Schwannoma is a usually benign tumor derived from Schwann cells. It can be found throughout the body, particularly in the head, neck, and extremities.. However, colonic schwannoma is extremely rare. Since most colonic schwannomas are asymptomatic, it can be incidentally found during colonoscopy or imaging studies performed for other reasons. A preoperative diagnosis of schwannoma may be made with imaging studies, such as computed tomography (CT) and magnetic resonance imaging (MRI), but a preoperative diagnosis is very difficult, and most of the cases are diagnosed after pathologic examination. Immunohistochemical staining is the basis of a definite diagnosis, and it can differentiate schwannoma from other mesenchymal tumors. Here, we present a case of a schwannoma of the cecum, which was preoperatively misdiagnosed as a gastrointestinal stromal tumor (GIST). A 65-year-old woman was referred for the treatment of a submucosal mass in the cecum. Colonoscopy revealed a 2 cm round submucosal mass in the cecum and abdominopelvic CT showed an enhancing mass in the cecum without lymphadenopathy. With the presumed diagnosis of GIST, laparoscopic right hemicolectomy was performed. Pathologic reports showed that it was consistent with schwannoma, immunohistochemically positive for S100 protein and negative for CD117 (c-kit), CD34, and smooth muscle actin. This case represents an unusual case of a schwannoma in the cecum, which could not be preoperatively diagnosed. Although its preoperative diagnosis is difficult given its rarity, a schwannoma can be considered as one of the differential diagnoses of a submucosal colon mass.

Keywords: *schwannoma, colon, immunohistochemical staining*

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1. Introduction

Schwannomas are usually benign tumors that are derived from Schwann cells composing the nerve sheath. [1] These tumors are mainly found in the head, neck, and extremities. However, gastrointestinal schwannomas are very rare, and the colon is an extremely rare location of this type of tumor. [2] Even though a presumptive preoperative diagnosis may be made with an imaging study or more rarely an endoscopic biopsy, a definite diagnosis is difficult without immunohistochemical (IHC) staining using a surgical specimen. [3] Treatment of colonic schwannoma is surgical excision, but the surgery should be optimized according to the tumor's nature, whether benign or malignant. [4,5] Here, we present a case of a 65-year-old woman with schwannoma of the cecum, which was preoperatively diagnosed as a gastrointestinal stromal tumor (GIST).

2. Case Report

A 65-year-old woman was referred to the Department of Surgery of Inje University, College of Medicine, Ilsan Paik Hospital for the treatment of a submucosal mass in the cecum. She had undergone a colonoscopy 1 month ago at a local clinic to screen for colorectal cancer, and was referred to the Department of Gastroenterology for further evaluation and management. She had no specific past medical history and did not complain of any symptoms related to the mass. Upon admission, her vital signs were stable and her laboratory results were within normal limits, including those of tumor markers. The colonoscopy showed a 2 cm round submucosal mass in the cecum (Figure 1).

Biopsy findings were mild active colitis with glandular hyperplasia without evidence of neoplasm. Abdominopelvic computed tomography (CT) indicated an approximately 2.5 cm enhancing mass in the cecum and there was no evidence of enlarged lymph nodes (Figure 2).

With the presumed diagnosis of a submucosal tumor, such as a gastrointestinal stromal tumor (GIST), laparoscopic right hemicolectomy was performed. Subsequently, microscopic analysis showed that the tumor was composed of fascicles of spindle cells with nuclear palisading in hematoxylin and eosin (H&E) staining. Immunohistochemically, the tumor cells were positive for S100 protein and negative for CD117 (c-kit), CD34, and smooth muscle actin, which was consistent with schwannoma. Ki-67 labeling index was 1% and there were no lymph node (LN) metastases

out of the 12 LNs examined (Figure 3).

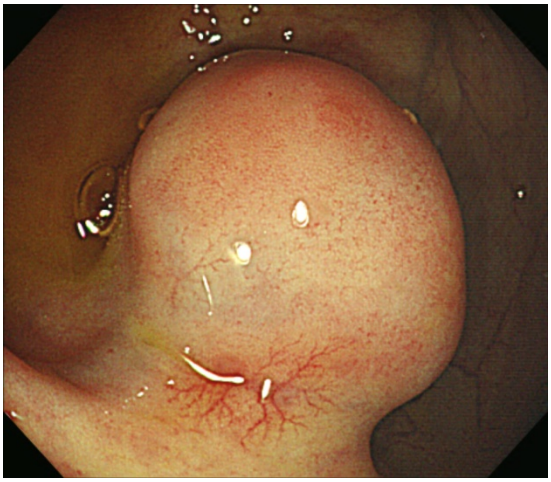

Figure 1. Colonoscopic findings. An approximately 2 cm, round, submucosal mass with a hard consistency is located in the cecum

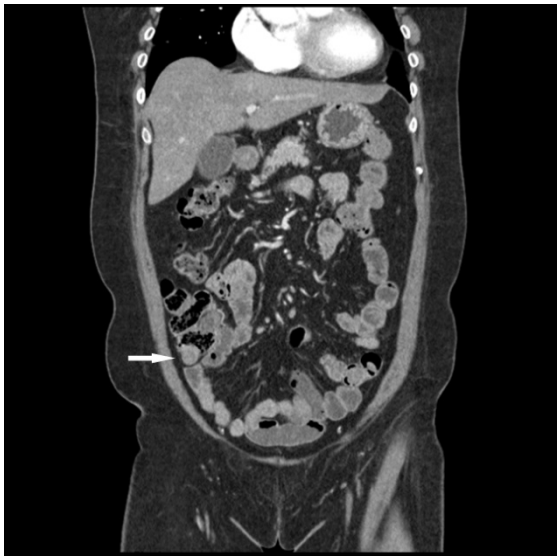

Figure 2. Abdominal CT (coronal image) showing an enhancing mass in the cecum without pericolic infiltration (white arrow)

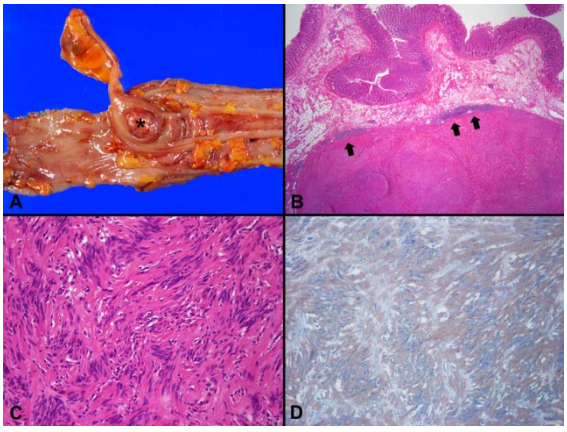

Figure 3. Pathologic findings. A) A subserosal protruding mass (asterisk) is noted in the cecum, measuring 2.4 × 1.8 × 1.5 cm. B) Low magnification view demonstrates a well-circumscribed solid mass with a peripheral lymphoid cuff (arrows) (H&E, × 10). C) The tumor is composed of fascicles of spindle cells showing nuclear palisading (H&E, × 200). **D)** Immunohistochemically, the tumor cells are positive for S100 protein (× 200), and negative for CD117 (c-kit), CD34, and smooth muscle actin

The postoperative course was uneventful, and the patient was discharged on the 7th postoperative day. Since then, she did not receive adjuvant chemotherapy and no evidence of distant metastasis or local recurrence was observed at 6 months' interval follow up or on a CT scan performed 2 years after the surgery.

3. Discussion

Schwannoma, as the name implies, is a tumor that originates from Schwann cells in the Auerbach's plexus composing the neural sheath. [1] Since gastrointestinal autonomic nerve tumors (GANTs) were described by Herrera et al., schwannoma, which is part of the family of GANTs, has been distinguished from other tumors, such as leiomyoma, leiomyosarcoma, leiomyoblastoma, and GIST on the basis of IHC staining. [3,6] Commonly involved organs are the head and neck, spinal cord, and extremities.[2] However, gastrointestinal schwannoma is very rare, comprising less than 5% of mesenchymal tumors of the gastrointestinal tract. The stomach and small intestine are the most common locations, but primary colonic schwannoma is extremely rare. [3] Among colorectal schwannomas, although the frequency of location is different in different reports owing to its rarity, cecum is the most common location followed by the sigmoid colon. [7,8]

As schwannoma is usually a slow-growing benign tumor, most of the cases are asymptomatic like the present case, but sometimes, depending on the size and location, they can cause symptoms such as abdominal pain, bleeding, and obstruction as a result of ulceration and mass effect. [7] Even though it may cause clinical symptoms and signs like typical colorectal cancer originating from the mucosa, the preoperative diagnosis of colonic schwannoma is very difficult. The first reason is that unlike colorectal cancer, the incidence of the tumor is extremely low, which makes unlikely to be considered in a differential diagnosis. Second, the tumors present as a submucosal tumor. Therefore, a routine superficial mucosal biopsy is more likely to indicate normal mucosa or inflammation as in our case. Third, the definite diagnosis of schwannoma requires IHC staining for differentiation from other mesenchymal tumors using mesenchymal tissue. That is why the rate of preoperative diagnosis has been reported to be only around 10% in the literature. [7]

Although most definite diagnoses are made after surgery, imaging studies such as CT and MRI may be helpful to evaluate the location, relationship with surrounding organs, LN metastases, and submucosal mass nature. While colonic schwannoma is depicted as a homogeneously attenuating, well-defined mass in the CT findings, GIST shows a more heterogeneous mass because of necrosis and hemorrhage. [9] In the present case, when we reviewed the CT findings retrospectively, they suggested schwannoma rather than GIST.

Therefore, schwannoma could be a differential diagnosis for a submucosal mass of the colon although its incidence is extremely low. In addition to CT findings, colonoscopic findings may assist in making a differential diagnosis. It has been reported that ulceration and

hardness of the mass may be specific findings of schwannoma because 34% of cases have exhibited ulceration. [7] In the present case, although hardness could be felt by the indirect tactile sense of the colonoscopy forceps, ulceration was not found.

For the definite diagnosis of schwannoma, pathologic examination is mandatory. Grossly, it appears as a spherical and well-circumscribed solid mass. H&E staining indicates a tumor mass composed of fascicles of spindle cells showing a nuclear palisading pattern. However, the diagnosis of schwannoma cannot be made with H&E staining alone because immunohistochemical staining is the most important diagnostic tool for differentiation of mesenchymal tumors. Immunohistochemically, smooth muscle tumors such as leiomyoma and leiomyosarcoma are positive for smooth muscle actin and desmin, while GIST is positive for CD117 (c-kit) and CD34 but not S-100 protein. In contrast to smooth muscle tumors and GIST, being positive for S100 protein and negative for CD117 (c-kit), CD34, and smooth muscle actin indicates schwannoma. [3,10]

Although the prognostic pathologic variables have not been well-established, a mitotic activity rate of more than 5 mitoses per field at high magnification and a tumor size larger than 5 cm might be correlated with a high risk of recurrence or distant metastasis, and tumors with a Ki-67 labeling index more than 10% can be malignant tumors. [2] In the present case, the tumor size was 2.4 × 1.8 × 1.5 cm and the Ki-67 labeling index was 1%, which indicated a benign tumor.

The treatment of choice of colonic schwannoma is surgical excision. Surgical options vary from local excision to oncological radical resection. Although distant metastasis has only rarely been reported, segmental resection or local excision with a clear margin could be a surgical option because the tumor is usually benign. [4,5] However, as schwannoma is very hard to differentiate from other submucosal tumors of the colon preoperatively, oncological radical resection could be reasonable. Moreover, laparoscopic oncologic colorectal surgery is feasible in most of cases of colorectal cancers. Therefore, we recommend laparoscopic oncologic resection in patients with a submucosal colon mass that has not had a confirmatory preoperative diagnosis.

4. Conclusions

This case represents an unusual case of schwannoma in the cecum, which could not be preoperatively diagnosed. Although its preoperative diagnosis is difficult given its rarity, schwannoma may be one of the differential diagnoses for a submucosal mass of the colon.

List of Abbreviations

computed tomography (CT)
magnetic resonance imaging (MRI)
gastrointestinal stromal tumor (GIST)
immunohistochemical (IHC)
hematoxylin and eosin (H&E)

Statement of Competing Interests

The authors have no competing interests.

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