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Single Case

Schwannoma of the Small Intestine

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Keywords
Schwannoma · Small intestine · Operation

Abstract
Schwannomas of the gastrointestinal tract are rare. Herein, we report a case of schwannoma originating from the small intestine. A 78-year-old woman underwent medical follow-up after surgery for bladder cancer, and a mass in the upper part of the pelvis was revealed by abdominal CT. With the diagnosis of a submucosal tumor of the small intestine, she underwent partial intestinal resection. The submucosal tumor was pathologically composed of S100-positive spindle cells and diagnosed as schwannoma. We report this case of rare schwannoma of the small intestine and review the literature.

Introduction

Schwannomas arise from Schwann cells of the peripheral nerve sheath that frequently develop in areas of the central nervous system, such as the spinal cord and the brain, the visceral peritoneum, the head and neck, and the surface of limbs. However, when schwannomas develop in the gastrointestinal tract, they are thought to originate from the Auerbach plexus or the Meissner plexus [1]. The frequency of schwannoma occurrence is 44.8% in the head and neck, 19.1% in the upper limbs, 13.5% in the lower limbs, and less than 10% in the gastrointestinal tract [2]. It is difficult to distinguish from mesenchymal tumors, it often requires

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diagnostic resection, and immunostaining is needed for accurate diagnosis. Here, we report a case of rare schwannoma of the small intestine.

Case Report

A 78-year-old woman underwent medical follow-up after surgery for bladder cancer, and a mass in the upper part of the pelvis was revealed by abdominal CT. She only had a history of hyperlipemia and had no history or family history of neurofibrosis and malignant diseases. She had no digestive symptoms, and her appetite had been normal. She had a soft abdomen and no pain, but the physical examination was positive for a 3-cm induration in the midline below the umbilicus. Laboratory findings included hemoglobin 11.8 g/dL, white blood cell count 6.2 × 103/μL, C-reactive protein 0.1 mg/dL, platelet count 252 × 103/μL, creatinine 0.67 mg/dL, alanine transaminase 21 U/L, and aspartate transaminase 23 U/L. They showed no abnormalities, and tumor markers (CEA: 0.1 ng/mL, CA19-9: 23 U/mL) were within normal limits. Enhanced abdominal CT demonstrated a 25 × 30 × 35 mm mass in the upper part of the pelvis, and the tumor was suspected to originate from the small intestine (Fig. 1). No enlarged lymph nodes or distant metastases were demonstrated. Colonoscopy showed only polyps. Therefore, with the diagnosis of a submucosal tumor of the small intestine, possibly gastrointestinal stromal tumor (GIST), she underwent laparotomy, at which partial intestinal resection was performed with a mechanical side-to-side anastomosis with a 1-cm margin from the tumor. Intraoperatively, the tumor was palpated in the wall of the small intestine.

Pathologically, a hard submucosal tumor of 4.5 × 3.4 × 2.4 cm was detected macroscopically (Fig. 2). Histologically, relatively uniform spindle-shaped cells were formed, in a palisading pattern, from the lamina propria to the subserous lamina of the small intestine. Immunohistochemical examination demonstrated that the tumor cells were positive for S100, and negative for αSMA, desmin, CD34, and c-Kit (Fig. 3). The MIB1 index was low, and no findings indicating malignancy were observed. Hence, the tumor was diagnosed as schwannoma, and the patient was discharged 10 days after operation. However, 12 days after discharge, she died of asphyxia.

Discussion/Conclusion

Schwannomas are tumors derived from Schwann cells, which develop preferentially in the head and neck, trunk and limb, and rarely develop in the gastrointestinal tract. It has been reported that of 246 cases of schwannomas and neurofibromas, 42 cases (17.1%) occurred in the gastrointestinal tract, among which 37 (88.1%) occurred in the stomach, 3 (7.1%) in the small intestine, and 2 (4.8%) in the colon [3]. It occurs most commonly in females between 30 and 60 years of age [4]. The main symptoms include abdominal pain, mass palpation, and bleeding, and less frequently, intestinal obstruction. Neurogenic tumors are submucous masses that are rich in blood vessels, and it is thought that bleeding associated with neurogenic tumors is due to the necrosis that accompanies tumor growth. It has also been reported that neurogenic tumors develop under the serosal membrane on the contralateral mesentery and exhibit exophytic growth. Therefore, intestinal obstruction by schwannoma of the intestine is uncommon. However, these symptoms appear as a result of tumor growth and are not disease specific [5, 6]. Therefore, as in the current case, such tumors are sometimes discovered incidentally by diagnostic imaging.

Histopathologically, schwannomas are well-defined tumors that show spindle-shaped cells on HE staining. Such tumors are classified into Antoni A type, in which spindle-shaped cells form palisading-like patterns, and Antoni B type in which the stroma is edematous and the tumor is hypocellular. Type B is reported as a secondary change that results from the growth of type A, and there are no differences in prognosis between type A and type B. In the current case, type A was mainly observed. As a differential diagnosis, mesenchymal tumors such as leiomyoma, leiomyosarcoma, and GIST can be considered; however, it is difficult to distinguish these tumors by HE staining alone, and immunohistological staining is usually required. Schwannomas are typically positive for S-100 and vimentin, and negative for desmin, keratin, c-kit, CD34, and αSMA [7–10].

If without symptoms, follow-up observation is also possible, but it is difficult to make a diagnosis before operation. Surgical treatment is often selected to treat symptoms or manage malignant diseases such as GIST [1]. Although the current case was asymptomatic, operation was performed because of the possibility of malignant diseases. Immunohistochemically, the tumor cells were positive for S-100 and negative for αSMA, desmin, CD34, and c-Kit, which was compatible with the diagnosis of schwannoma. The long-term follow-ups of patients with schwannomas of the gastrointestinal tract have not shown any propensity to recurrence following complete excision [4]. In the current case, the patient died of other factors soon after discharge, and therefore, tracking of the prognosis was impossible.

In summary, we experienced a case of incidental schwannoma of the small intestine. Although schwannomas rarely develop in the small intestine, it is important to keep them in mind as a differential diagnosis of neoplasms of the small intestine.

Statement of Ethics

This study was conducted in accordance with the Declaration of Helsinki.

Disclosure Statement

The authors declare that there are no conflicts of interest.

Author Contributions

N. Fukushima wrote this article. H. Aoki supervised the writing of the manuscript. K. Yoshida and K. Yanaga approved the final submission of the manuscript. M. Ogawa performed the preoperative management and operation. N. Fukazawa performed pathological analysis. All authors read and approved the final manuscript.

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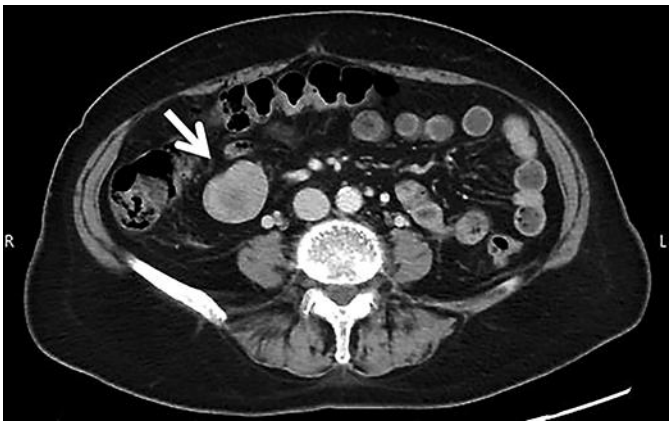

Fig. 1. Enhanced abdominal computed tomography showed a 30 × 30 × 35 mm mass in the upper part of the pelvis (arrow).

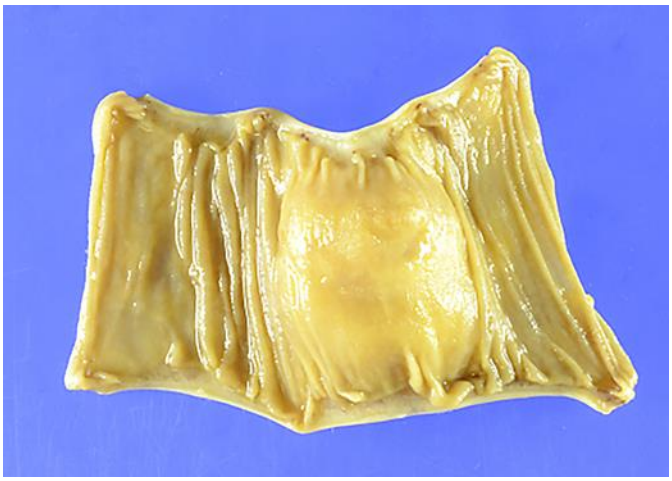

Fig. 2. Macroscopic findings showed a hard submucosal tumor in the small intestine.

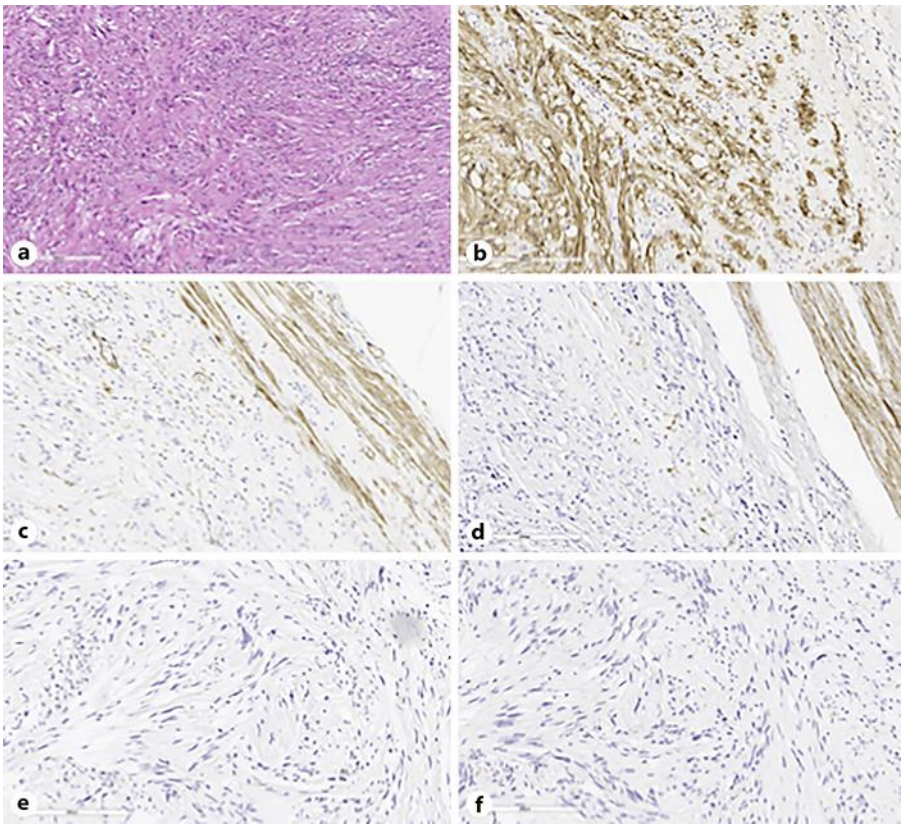

Fig. 3. Histopathology of the specimen. **a** Histopathological findings revealed relatively uniform spindle-shaped cells. HE. ×100. Immunohistochemically, the tumor cells were positive for S-100 (**b**) and negative for αSMA (**c**), desmin (**d**), CD34 (**e**), and c-Kit (**f**). ×100.