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Incidental Finding of an Asymptomatic Jejunal Schwannoma: A Rare Case Report and Review of Literature

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Statistical Analysis C
Data Interpretation D
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Patient: Female, 57-year-old
Final Diagnosis: Jejunal schwannoma
Symptoms: Asymptomatic
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease

Background: Schwannomas are tumors that arise from Schwann cells that surround and support nerve cells. Most common sites for presentations are head, neck, and extremities. Schwannomas of gastrointestinal tract are rare, slow-growing tumors, usually benign, arising from gastrointestinal tract's neural plexus. They are histologically distinguishable from conventional schwannomas that arise in soft tissue or the central nervous system. Preoperative diagnosis of gastrointestinal schwannoma is challenging, requiring immunohistological confirmation of the nature of the tumor. Here, we report a case of 57-year-old woman with an incidental finding of an asymptomatic submucosal jejunal schwannoma.

Case Report: A 57-year-old woman with a medical history of hematological disorder underwent a contrast abdominal computed tomography as part of medical follow-up. The imaging revealed the presence of a jejunal mass. The patient underwent laparoscopic surgical resection of the lesion, followed by side-to-side jejuno-jejunal anastomosis with 4-cm clear surgical margins. The final pathologic study revealed the presence of jejunal schwannoma, as tested positive for S-100 protein. The patient was discharged home on the fourth postoperative day, having an uneventful recovery.

Conclusions: Jejunal schwannoma are usually benign and asymptomatic, and they are often discovered incidentally during diagnostic tests for other conditions; therefore, it should be included in the differential diagnosis of gastrointestinal tumors. Surgical treatment appears to be necessary to achieve a definitive diagnosis through a biopsy of the tumor tissue. Benign jejunal schwannomas have a good prognosis.

Keywords: Schwann Cells • Jejunal Neoplasms • S100 Proteins • Jejunum • Digestive System Neoplasms

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Introduction

Schwannomas are tumors deriving from excessive proliferation of Schwann cells, which produce the myelin sheath for the peripheral nervous system [1]. They can be found in different parts of the body, including the central nervous system, the head and neck, the peripheral nerves in the extremities, and less frequently in the gastrointestinal system [1]. Gastrointestinal schwannomas have been described as slow-growing tumors, usually benign in nature, originating in the Auerbach or in the Meissner myenteric nerve plexus [2,3]. They are identified as mesenchymal tumors that infrequently occur. Generally, gastrointestinal schwannomas constitute about 2-6% of all mesenchymal tumors, most commonly occurring in the stomach (60-70%), followed by the colon and rectum (3%); they tend to occur even less frequently in the esophagus and small intestine [1,2,4,5].

The age of occurrence is about 60-65 years old, with a female preponderance [4]. Gastrointestinal schwannomas can present with various symptoms, depending on the site of development and size of the tumor [4,6-8]. Preoperative diagnosis of schwannoma is difficult, resulting in surgical resection and subsequent histological and immunohistochemical testing as the leading option for establishing diagnosis [2].

Here, we report a rare case of jejunal schwannoma, presented asymptomatically in a female patient, which was detected by abdominal computed tomography (CT).

Case Report

A 57-year-old woman was referred to the surgery department for a jejunal mass, having been revealed after an abdominal computed tomography (CT) scan requested for her hematological disorder follow-up.

According to her medical history, she had persistent monocytosis, rendered as possible chronic myelomonocytic leukemia, hypothyroidism, and adult xanthogranuloma (multiple cutaneous lesions at the neck region). She had only been treated with levothyroxine 64 μg. Previous surgeries included appendicectomy (at the age of 20) and hysterectomy due to uterine fibroids (at the age of 46). No specific family history was identified.

She had no clinical manifestations of gastrointestinal disorder, and upon physical examination, no abnormalities were detected except for a palpable spleen.

Laboratory findings showed hemoglobin 12.2 g/dl, white blood cell count 8.23×10^3/μL (monocytes 41%), platelet count 155×10^3/μL, and tumor markers within normal limits (carbohydrate antigen 19-9: 2,00 U/mL, carcinoembryonic antigen: 0,58 ng/mL).

As part of the work up examinations for her hematological disorder, an abdominal CT scan with contrast agents, including intravenous (IV) and oral, was requested. The enhanced abdominal CT scan showed a rounded, well-defined, homogeneous mass 3 cm in diameter, at the level of the jejunum (**Figure 1**). Other noticeable findings included multiple lesions at the liver, resembling hepatic hemangiomas, and spleen dimensions of 14.2×5×13 cm.

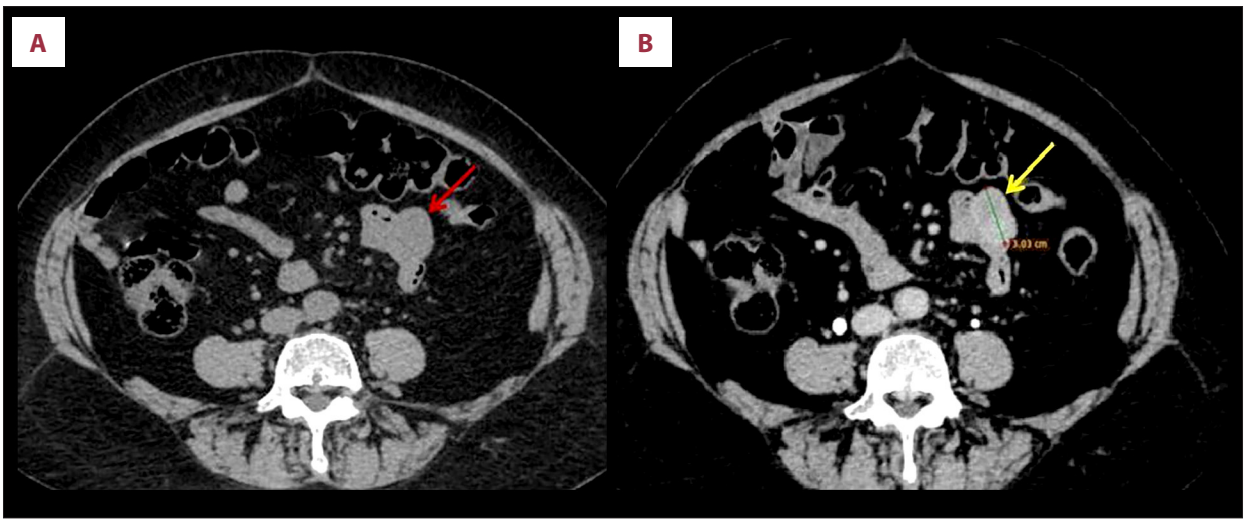

Figure 1. Abdominal computed tomography images of the benign jejunal schwannoma in a 57 -year-old woman. Axial view. (**A**) Isodense, well-defined, mural mass in the jejunum (red arrow). (**B**) Delayed phase. Homogeneous progressive attenuation of the lesion. Lesion measuring 3 cm (yellow arrow).

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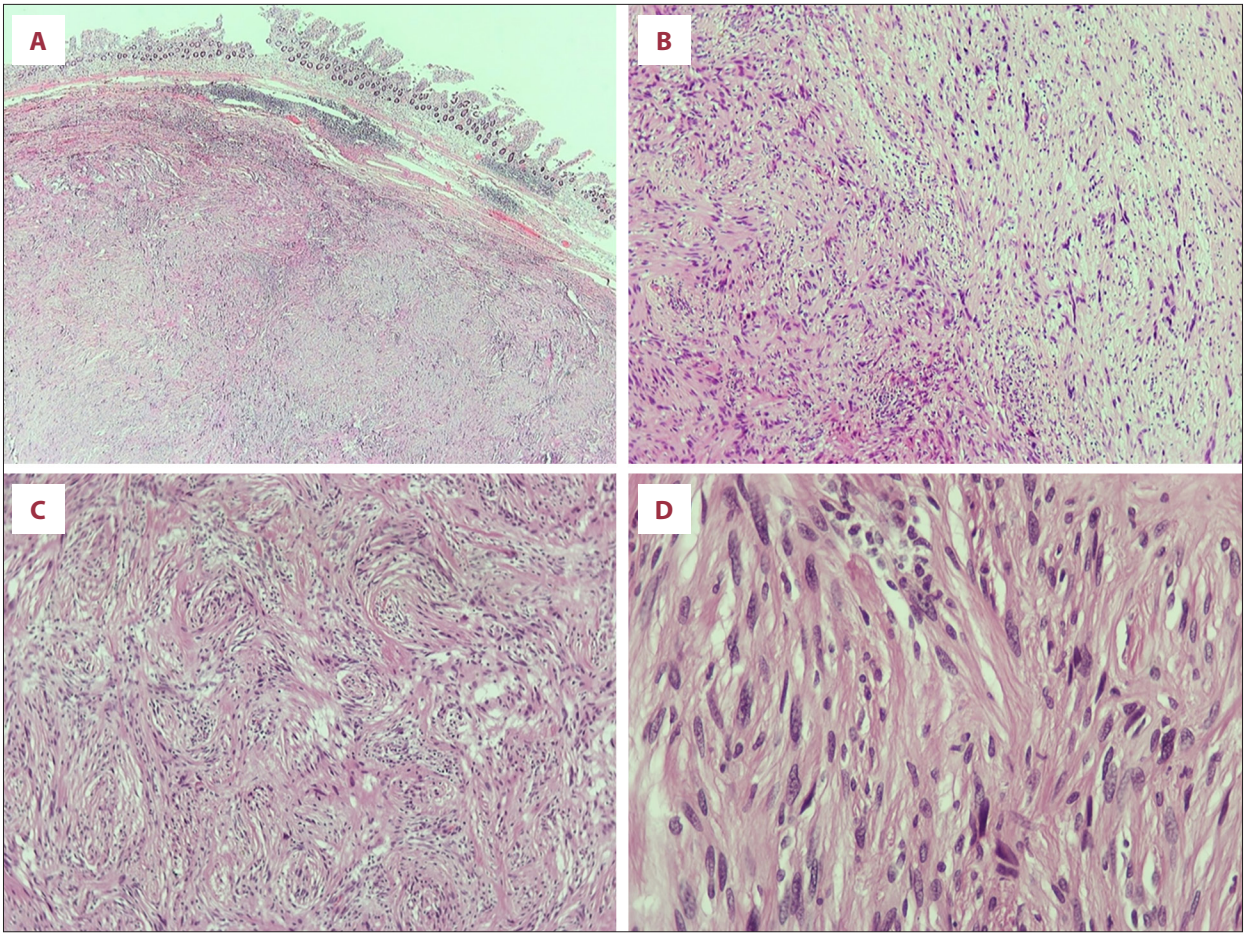

Figure 2. (**A-D**) Histologic analysis of the benign jejunal schwannoma in a 57-year-old woman. Hematoxylin and eosin stains, (20-400× magnifications), showing the submucosal neoplasm below the intestinal mucosa (**A**) showing areas of mild cellularity and moderate cellularity (**B**). The neoplastic cells were arranged in fascicles and whorls (**C**) and they show spindle to ovoid nuclei with focal mild pleomorphism (Antoni A type), without increased mitotic activity (**D**).

Taking into consideration the main differential diagnosis of mesenchymal tumors, we suggested the patient undergo surgical excision of the tumor. After patient consent, we proceeded to laparoscopic surgical resection of the jejunum containing the lesion, followed by side-to-side jejuno-jejunal anastomosis with 4-cm clear surgical margins.

According to the morphological and immunohistochemical findings, the tumor was diagnosed as schwannoma of the jejunum, excluding other, primarily spindle cell, neoplasms.

More specifically, the macroscopic examination revealed an exophytic lesion, with largest diameter of 3 cm, bulging on the external wall and protruding in the lumen, elevating the mucosal surface but without penetrating through it.

The histologic analysis showed the neoplastic cells, which were arranged in intermingled whorls and fascicles, with areas of mild and moderate cellularity, resembling features that are mainly observed in Antony A type arrangement of Schwann cells. The neoplastic cells were spindled or ovoid, medium-to-large size, with spindle or ovoid nuclei with fine chromatin, and eosinophilic cytoplasm (**Figure 2**). The nuclei showed focally mild pleomorphism and rare mitoses (1/10HPF), and necrosis was absent. The neoplastic cells were found in the submucosal (primarily) and muscular wall, without penetrating the serosa or the mucosa. Peritumoral lymphoid cuffs were present.

Immunohistological examination showed that the neoplastic cells were positive for protein S100, SOX10, and CD56, and focally for GFAP. They were negative for CD117, DOG1, CD34, smooth muscle actin, caldesmon, desmin, keratins 8/18, chromogranin, synaptophysin, and HMB45. The mitotic index Ki67/MIB1 was 1-2%, reaching 5% in rare hot spots (**Figure 3**).

The patient was discharged 4 days after the operation without exhibiting any complications. Because the pathology report described a benign lesion, no additional treatment measures

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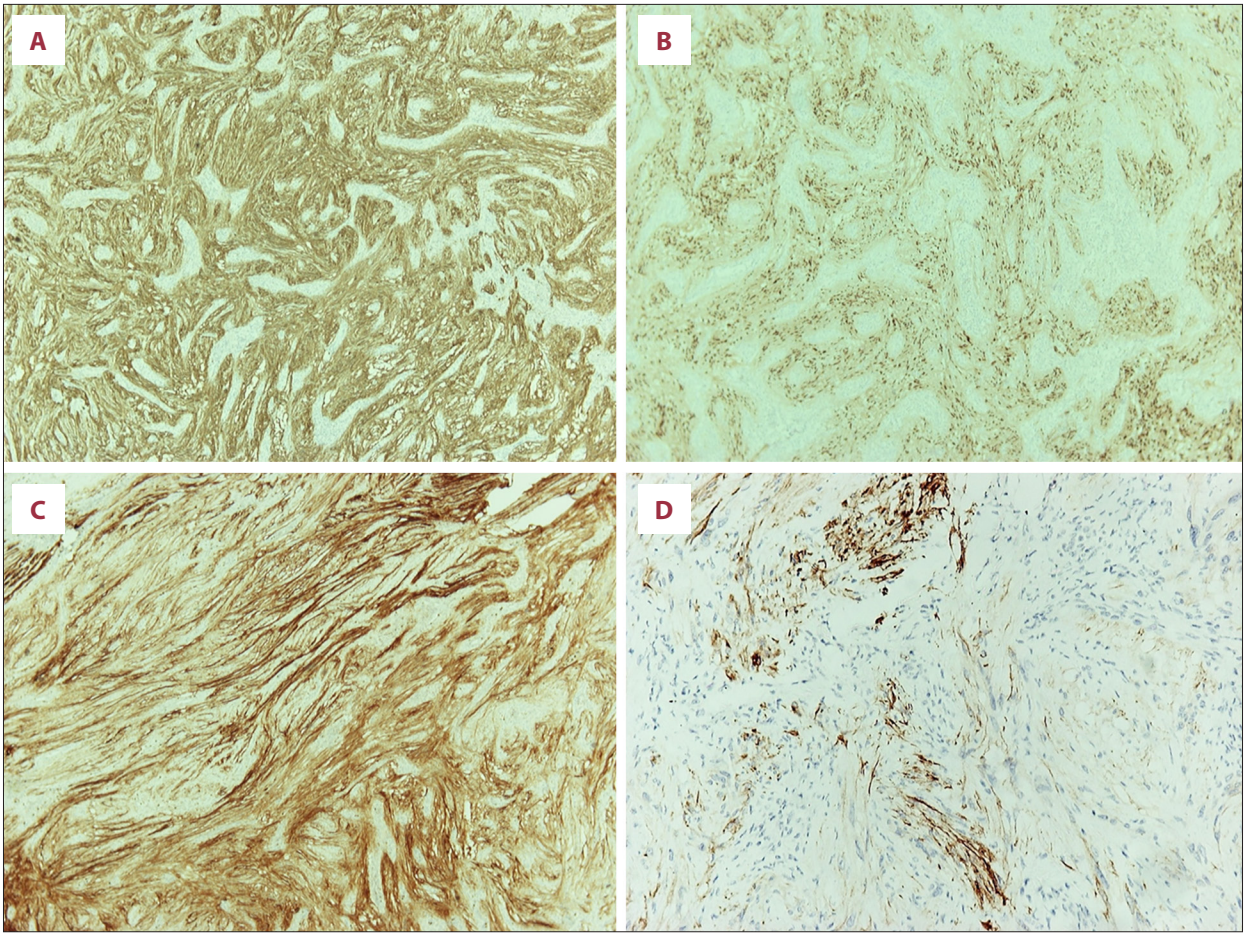

Figure 3. (**A-D**) Immunostaining analysis of the benign jejunal schwannoma in a 57-year-old woman. Positive immunostaining for protein S100 (**A**, 40× magnification), SOX10 (**B**, 40× magnification), CD56 (**C**, 40× magnification), and focally for GFAP (**D**, 40× magnification) in the neoplastic cells of our patient with jejunal schwannoma. A hot spot with the mitotic index Ki67/MIB1 is shown in the inset (**D**, 20× magnification).

were performed. The patient was scheduled for subsequent surveillance with abdominal CT scan to exclude recurrence.

Discussion

Schwannomas of the gastrointestinal tract belong to the class of the mesenchymal tumors, which are defined as different neoplasms from conventional schwannomas that develop in soft tissue or the central nervous system [2,5,7,9,10]. Schwannomas of the digestive tract are usually benign tumors with excellent prognosis [9,10]. They are thought to originate from the Auerbach plexus or the Meissner plexus, which are parts of the intestinal neural plexus [3,4]. This hypothesis has its root in the immunophenotypic similarities that both schwannomas and the cells of the myenteric plexus express, which are related to S-100 protein positivity [5].

The incidence of gastrointestinal schwannomas is very low and they are most commonly found in the stomach [3-5]. Reports have shown they most often occur in women aged 30-60 years old [2-5,7,12].

Due to the varying degrees of malignancy and the subsequent treatment plan, it is very important to differentiate gastrointestinal schwannomas from other submucosal tumors, such as gastrointestinal stromal tumors (GIST), leiomyomas, and leiomyosarcomas [5,11,12]. Although it is extremely rare, cases of malignant schwannomas of the gastrointestinal tract have been reported and diagnosed based on mitotic activity of the neoplasm [13].

The literature contains few reports of cases of small intestinal schwannomas cases. The symptomology appears clinically indolent, and acute presentation is rare. We searched the English literature in PubMed regarding small intestinal schwannomas, using keywords “small intestinal schwannomas; jejunal schwannomas” (**Table 1**).

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Table 1. A review of the literature on small intestinal schwannomas.

Article	Published year	No cases	Presentation	Histology report
Hasselfeldt-Nielsen, et al [27]	1987	1	Melena	Benign
Eskelinen, et al [23]	1992	1	Nonspecific symptoms*	Malignant
Zhou, et al [24]	1999	1	Melena	Benign
Iida, et al [21]	2003	1	Nonspecific symptoms*	Benign
Nagai, et al [19]	2003	1	Melena	Benign
Yilmaz, et al [13]	2004	1	Nonspecific symptoms*	Malignant
Zentar, et al [26]	2014	1	Incidental finding	Benign
Portale, et al [25]	2016	1	Incidental finding	Benign
Mekras, et al [2]	2018	3	Incidental finding	Benign
Roulston, et al [14]	2018	1	Ileal intussusception	Benign
Shu, et al [4]	2019	2	Melena	Benign
Fukushima, et al [3]	2019	1	Incidental finding	Benign
Hokama, et al [18]	2019	1	Melena	Benign
Peng, et al [20]	2022	3	Incidental finding, nonspecific symptoms*	Benign
Kumagai, et al [22]	2023	1	Melena	Benign

* Nonspecific symptoms include abdominal pain, abdominal distention, abdominal discomfort, vomiting, fatigue, anemia, and gastrointestinal bleeding [20-23].

The symptoms are nonspecific and vague [2-5,7,12,15]. Depending on the location of the tumor growth and development, the patient can present with atypical abdominal pain, melena, mass palpation, and, rarely, intestinal obstruction [1,6,14-16]. The presenting symptom of gastrointestinal bleeding could be due to the growing submucosal mass abundant in blood vessels, which at some point extends its vascular supply, leading to ischemic tissue formation that ulcerates and bleeds [3,11,17]. There have been case reports in which the main symptom was intestinal obstruction caused either by the tumor itself or by developing into intussusception [8,4]. Nevertheless, as in our case, the symptoms may be absent and the tumor is incidentally discovered by imaging [3].

Preoperative diagnosis of schwannoma is challenging because there are no disease-specific symptoms or imaging studies showing distinct features unique to this tumor. The most common modalities of imaging, such as CT, magnetic resonance imaging, ultrasound, and endoscopy, are useful for assessing the location of the tumor, the presence or the extent of the invasion, and revealing any complications [5,8,14-15,]. In addition, it has been reported that a jejunal mass, which proved to be schwannoma, had been identified by the use of video capsule endoscopy [18]. Among cases presenting with gastrointestinal bleeding, useful tools for workup appear to be blood scintigraphy or angiography [19].

Definitive diagnosis can only be made on the basis of histopathological and immunohistochemical features. Histopathologically, Schwannomas are solid homogeneous tumors that demonstrate spindle-shaped cells. Immunohistochemically, they appear positive for S-100 protein and negative for c-KIT, DOG1, CD34, SMA, and desmin. These immunohistochemical staining techniques are sufficient to confirm the diagnosis and differentiate schwannomas from other types of neoplasm [1-4,7,12].

The principal treatment for gastrointestinal schwannoma is complete surgical resection. This treatment approach is necessary due to the risk of underlying malignancy in any tumor-like finding in the gastrointestinal tract [2-4]. Taking into consideration the different size, location, the association with the surrounding tissues, and the dominant symptoms of the lesion, they are usually treated with endoscopic resection, laparoscopy, or open surgery [1]. Endoscopic resections are usually reserved for tumors less than 3 cm in diameter, with the risk of post-excisional bleeding, perforation, or the possibility of creating a fistula [1]. Open or the laparoscopic surgical resection are considered the best ways to assess curative treatment [1].

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This case report reminds us that it is important to include the diagnosis of schwannoma into the differential diagnosis of gastrointestinal submucosal tumors. However, to confirm the diagnosis by immunohistochemical methods, surgery appears to be the only appropriate treatment option.

Conclusions

Jejunal schwannoma is a rare type of tumor that originates from myenteric nerve plexus, with variable clinical presentation. The literature contains few reports of schwannomas in the small intestines. The symptomology appears clinically indolent or, rarely, with acute presentation. The nonspecific clinical manifestations and nonspecific imaging diagnostic standards contribute to misdiagnosis. Surgical resection of the tumor is the most effective approach to diagnose and treat jejunal schwannoma. Histological confirmation is achieved by positivity for S-100, the hallmark of schwannoma. Prognosis is generally good for patients with jejunal schwannoma, with a low risk of recurrence or malignant transformation.

Institution Where Work Was Done

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Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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