

Schwannoma of the Rectosigmoid Colon

Abstract

Schwannoma is a rare tumor in the colon which originates from the peripheral nerve plexus. Most of the cases have been asymptomatic but occasionally present as an obstructive mass. Abdominal investigations are effective in some cases, but usually, they are not informative. A significant number of cases have been detected after their operation by histopathology examination. Immune and histochemical staining shows the spindle cells that have been positive for S-100 and vimentin, but negative for CD34 and smooth muscle actin. If the diagnosis of Schwannoma is confirmed preoperatively, segmental resection is recommended. In this case report, we presented a 58-year-old woman with pelvic mass and normal colonoscopy that mimic extraluminal large uterine myoma with extraluminal pressure effect on the rectosigmoid.

Keywords: Colon, colonoscopy, rectum, schwannoma, sigmoid, surgery, tumor

Introduction

Schwannoma of the gastrointestinal (GI) tract is a rare spindle cell tumor originating from the peripheral nerve. Most cases present in the subcutaneous tissues of the extremities and the head and neck.^[1,2] Primary schwannoma of the colon and rectum, which is not related to systemic neurofibromatosis (Von Recklinghausen disease) has been an uncommon neoplasm from Auerbach's myenteric and Meissner's submucosal plexus.^[2] Most of the time, this tumor is asymptomatic, but sometimes it may present as obstructive adenocarcinoma symptoms such as obstruction, bleeding, pain, and tenesmus.^[2,3] In this case study, we describe a patient with rectosigmoid schwannoma presenting as a pelvic mass (like exophytic uterine mass).

Case Report

The patient was a 58-year-old woman with complaints of lower abdominal pain (cramp type), change in bowel movement, and sometimes abdominal distention. She did not have any drug history, psychosocial history including smoking or alcohol and any family history like genetic diseases. She did not have any sign of rectal bleeding or weight loss. She was referred to a gastroenterologist by her family physician and she had undergone

colonoscopy. In her colonoscopy, a sessile polypoid lesion, 0.3 cm in diameter was detected in the descending colon and removed by snare loop. Histopathological evaluation revealed hyperplastic polyp. Her pelvic ultrasonography had showed posterior uterine exophytic myoma with external pressure on the recto-sigmoid junction. Subsequent abdominopelvic computed tomography (CT) scan revealed a well-circumscribed slightly lobulated mass lesion, with almost homogenous density, arising from the proximal part of the rectum. Portal phase images revealed moderate enhancement of the mass with a density value of 66 HU. No calcification was detected. The lumen of the rectum was not identifiable at the level of the mass, and additionally; there was the suggestion for an extraluminal growth into the adjacent perirectal space. Several enlarged lymph nodes were also visualized. There was no liver or bone metastasis, and no peritoneal seeding or ascites was detected [Figures 1 and 2].

She was scheduled for trans-abdominal hysterectomy by a gynecologist, but she had postponed her operation because of the COVID pandemic situation for 5 months. Finally, laparotomy was done and a partial circumferential mass in the rectosigmoid junction with a length of 5 cm without any adhesion to the uterus or other structures was detected by the gynecologist surgeon in

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Shahid Faghihi Hospital. The gynecology team called the colorectal surgeon due to operation findings. The anterior resection with colorectal anastomosis was done by the colorectal surgery team. She was admitted to the Colorectal Surgery Ward without any postoperative complications and discharged after 5 days.

Histopathological and immunohistochemistry (IHC) examination showed Schwannoma mass of the rectosigmoid colon due to positive S100 and SMA and negative CD117 and DOG1 test. Her pathology study and imaging were reviewed again and IHC confirmed the diagnosis of Schwannoma colon tumor.^[4] Histopathological examination of the resected mass showed a spindle cell neoplasm with hypo- and hyper-cellular areas and thick wall hyalinized blood vessels. Rare mitotic figures were present. The subsequent IHC examination showed strong positivity of S100 and negativity for CD34, cytokeratin, smooth muscle actin, C-KIT, and DOG1. The ki-67 showed 3%–4% immunolabeling. According to

morphology and IHC results, the diagnosis of Schwannoma was made [Figures 3-6]. She was visited 2 weeks and 2 months after her operation without any complications.

Discussion

Peripheral nerve sheath tumors are found in 2%–6% of stromal tumors of the GI tract, but solitary schwannomas of the rectosigmoid area have been rare. The most common site of GI Schwannoma is the stomach that was diagnosed accidentally on screening test and the least incidence is found in the colon. They had the same incidence in men and women with a median age of 65-year-old in previous case reports, but according to a systematic literature review by Bohlok *et al.* and his collages, colorectal schwannoma has been diagnosed slightly more in female patients (59%) with a different age range from 14 to 95 years.^[3] The size of this tumor has been reported from <1 cm as a submucosal polyp to large tumors near 28 cm that present with a palpable abdominal mass.^[3,5] It is usually asymptomatic but can produce nonspecific or GI symptoms such as constipation, bleeding, obstruction, nonspecific pain, and even intussusception in children as a case report.^[6]



Figure 1: Axial contrast-enhanced abdominal computed tomography scan through the pelvic cavity demonstrates a 67 mm × 51 mm almost homogeneous enhancing mass (asterisk) arising from the wall of the proximal part of the rectum. Several enlarged lymph nodes are also detectable (black arrows)

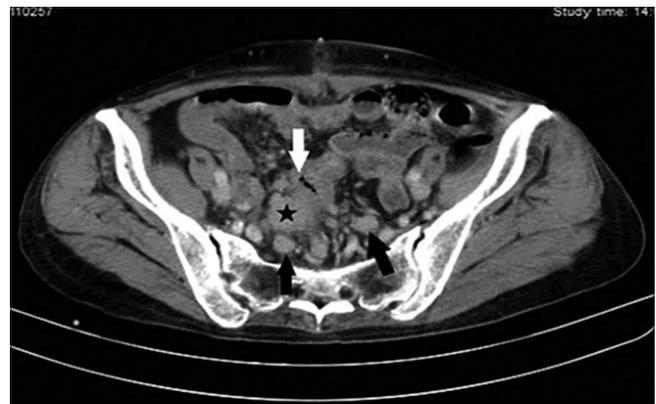


Figure 2: Axial section through the more superior part reveals the mass arising from the rectal wall (white arrow)

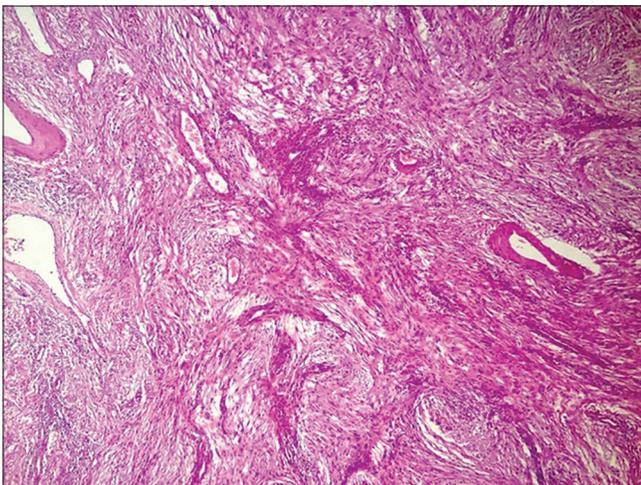


Figure 3: Histopathological examination of the mass shows hypo- and hyper-cellular areas with thick wall hyalinized blood vessels, H and E, ×200

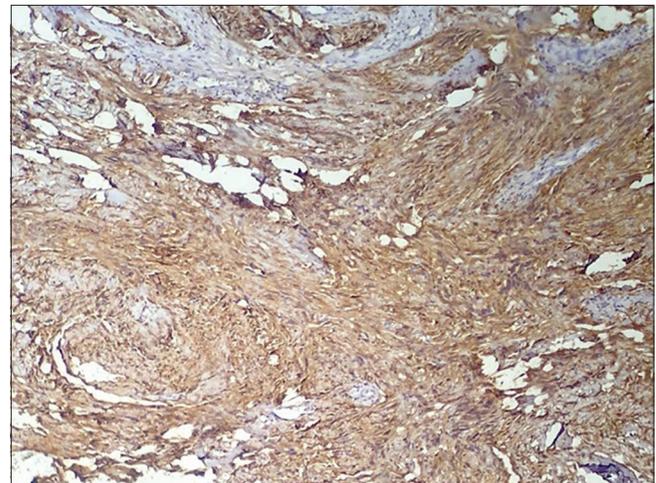


Figure 4: Strong and diffuse S100 immunostaining

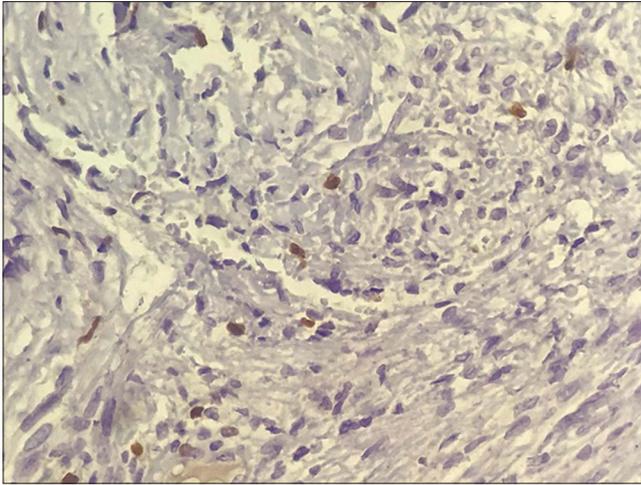


Figure 5: Ki-67 immunolabeling showed 3%–4% proliferation index

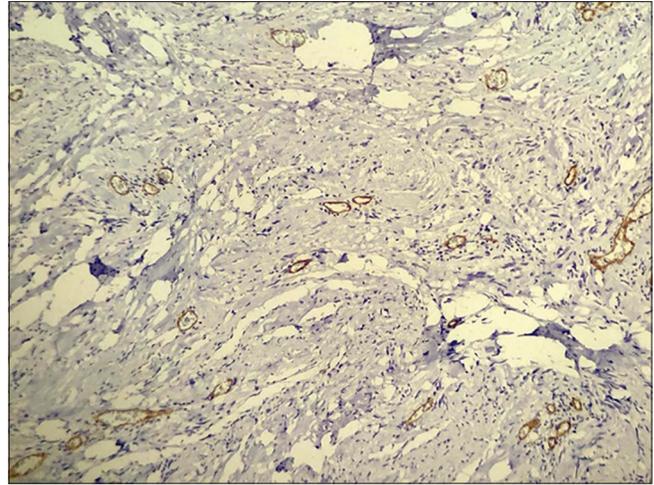


Figure 6: Immunostaining of CD34 which is negative in the tumor cells in contrast to staining of the vessels

There is no evidence of malignant metastases or connection with neurofibromatosis 1 or 2 as well. Visceral schwannomas have been uncommon, and preoperative diagnosis remains challenging. In terms of radiologic evaluation, the appearance is usually nonspecific, mimicking more common GI lesions, especially those of the mesenchymal origin.^[3,7,8] They appear as nonspecific hypoechoic submucosal lesions on endoscopic ultrasonography. Most of the schwannomas are identified as a well-defined submucosal lesion with homogeneous density on both unenhanced and contrast-enhanced abdominal CT scans.^[3] The majority of the lesions tend to have mild to moderate enhancement upon injection of contrast.^[7] The degree of the enhancement depends on the phase of the CT scan; while most of the reported cases show low enhancement on the arterial and portal phase, more conspicuous enhancement has been observed in the equilibrium phase.^[3,8] Interestingly, hemorrhage, cystic changes, necrosis, or calcifications, which are more commonly observed in mesenchymal tumors such as GI stromal tumor, are rather absent in schwannomas, thus resulting in the aforementioned homogeneous CT appearance. While lymphadenopathy is not a common associated feature,^[8] enlarged lymph nodes have also been reported in CT scans as in our case and are believed to be the result of cytokine-induced chemokinesis of the lymphocytes or an inflammatory reaction rather than tumor metastasis.^[7]

Surgical excision is essential to confirm the diagnosis as pathology is the “gold standard.”^[9] Submucosal or deep biopsy sometimes helps to establish a preoperative diagnosis.^[3] Schwannomas of the colon were misdiagnosed as GI stromal tumors such as leiomyomas and leiomyosarcomas, but with advances in intracerebral hemorrhage staining, the detection of colon schwannoma has been easier.^[10] Colorectal schwannomas are divided into three clinicopathologic types: spindle cell, epithelioid, and plexiform.^[11] Schwannomas often have significant amounts

of the myxoid matrix that is positive with alcian blue at pH 2.5. In addition, it may show only vague rudimentary nuclear palisading and compact cell bundles,” in contrast to the distinct Antoni A, Antoni B and Verocay bodies of soft-tissue schwannomas.^[12] Schwannoma shows diffuse strong positivity for S-100 and vimentin and variable positivity for glial fibrillary acidic protein; it is also negative for CD34, CD117, desmin, c-Kit, and actin.^[4,11]

Complete surgical resection with adequate free margins remains the treatment of choice, but radical resection is unnecessary to manage the tumor with aggressive lymph node dissection.^[10,13] The procedure depends on the size and location of the tumor. In accordance with some articles, the minimally invasive approaches by laparoscopy, robotic surgery, and even endoscopic resection have been accepted techniques to remove Schwannoma of the large bowel. In addition, transanal endoscopic resection is another option for rectal involvement in the distal part.^[9,13-15] Additional resection should be considered for very unusual cases with coincident malignant tissue.^[16] The use of radiotherapy and adjuvant chemotherapy have not been advised because of benign behavior presentations.^[17,18] There is a rare incidence of lymph node involvement with loco-regional and liver metastases in aggressive tumors (2%) depending on the large size (more than 5 cm) with multiple mitoses in mitotic index calculation.^[3,19] The patient gave us inform the consent for publication as well.

There is no conflict of interest. In addition, the authors of this case report are affiliated to Shiraz University of Medical Sciences. The patients signed the consent form of this case report as well.

Conclusion

Schwannoma of the colon and rectum is a rare tumor. The definitive diagnosis is based on IHC of the operative specimen. Schwannoma stains strongly positive for S100

and the mitotic index should be calculated to exclude malignant lesions. Surgical segmental resection is the treatment of choice without complete mesocolic or total mesorectal excision.

We used SCARE guideline 2020 to write this case report.^[20]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient have given her consent for her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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