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Case Report

SCHWANNOMA OF THE ASCENDING COLON: AN UNEXPECTED CAUSE OF LOWER GASTROINTESTINAL BLEEDING

SCHWANNOMA DE COLON ASCENDENTE: UNA CAUSA INESPERADA DE HEMORRAGIA DIGESTIVA BAJA

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Abstract

Schwannomas of the gastrointestinal tract are spindle cell tumors originating from peripheral nerve lining Schwann cells, accounting for 2–6% of all mesenchymal tumor and are benign in 98% of cases. Most commonly, they are asymptomatic and discovered incidentallyin elderly. The preoperative diagnosis is challenging because clinical manifestations and CT findings are nonspecific, and biopsies during colonoscopy are in most cases non conclusive, as in our patient, who presented atypical abdominal pain and hematochezia. The best therapeutic option is a complete surgical resection, which will give the definitive diagnosis by confirming S-100 proteins on the immunohistochemical analysis.

Resumen

Los schwannomas del tracto gastrointestinal son tumores de células fusiformes que se originan en las células de Schwann del revestimiento de los nervios periféricos, representan del 2 al 6 % de todos los tumores mesenquimatosos y son benignos en el 98 % de los casos. Más comúnmente, son asintomáticos y se descubren incidentalmente en ancianos. El diagnóstico preoperatorio es desafiante porque las manifestaciones clínicas, así como los hallazgos colonoscópicos y tomográficos, son inespecíficos, como en nuestro paciente, quien presentó dolor abdominal atípico y hematoquecia. La mejor opción terapéutica es la resección quirúrgica completa, que dará el diagnóstico definitivo a partir del examen inmunohistopatológico de la pieza operatoria, característicamente positividad para proteínas S100.

Keywords: Schwannoma, colo-rectal, hematochezia, S100 protein.

Introduction

Neurilemmoma or schwannomas of the gastrointestinal tract are spindle cell tumors originating from peripheral nerve lining Schwann cells. Most frequently, they originate from Auerbach's plexus, [1] and represent a very rare entity, accounting for approximately 2–6% of all mesenchymal tumor.[2,3] They occur in decreasing frequency in the stomach (60-70%), small bowel (10-15%), and finally the right colon and cecum (3%), [2] followed by the sigmoid colon, rectum, left colon and the transverse colon.[4,5] They occur slightly more in female patients (59%), with a mean age of 60–65 years. [4]

Case Report

A 70-year-old man, orally anticoagulated with Rivaroxaban, presented with a nonspecific pain in the right abdomen and isolated episodes of hematochezia.

A screening colonoscopy was performed and showed a diverticulosis of the sigmoid colon and a 3.5 cm diameter submucosal spherical hard tumor with central ulceration in the proximal ascending colon (figure 1), from which biopsies were difficult to take due to the hardness of the lesion and the high risk of perforation.

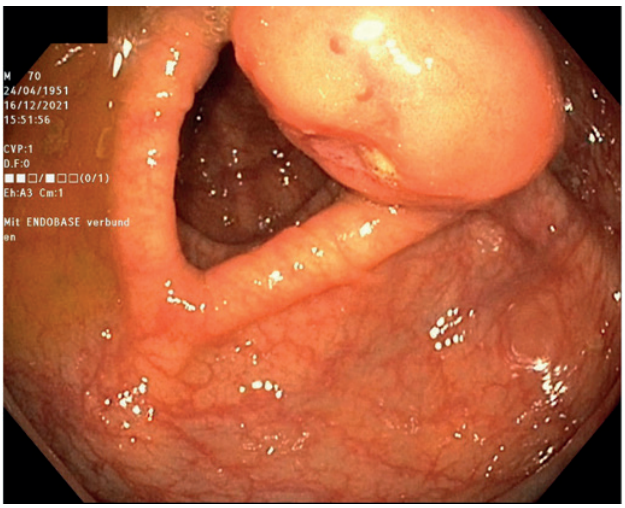

Figure 1: Endoscopic view showing a 3.5 cm sized globular submucosal hard tumor with central erosion located at the ascending colon.

Laboratory examination showed no anemia and a normal value of carcinoembryonic antigen (CEA). The other laboratory test results were within normal ranges.

From the available biopsy material, no definitive histological diagnosis could be made. After multidisciplinary team discussion, a differential diagnosis of neuroendocrine tumor or mesenchymal tumor of the colon (gastrointestinal stromal tumor and leiomyoma) were suggested. For this reason, the colonoscopy was repeated to take new biopsies (figure 2) with inconclusive findings again.

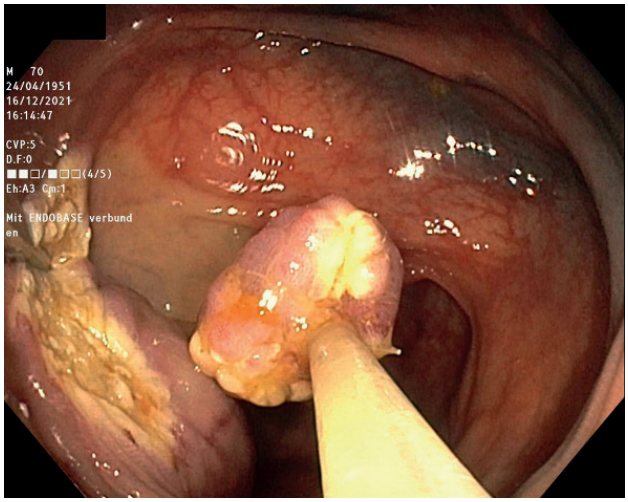

Figure 2: Endoscopic partial removal of the tumor.

The abdominal computerized tomography (CT) confirmed a not complicated diverticulosis of the colon sigmoideum, as well as in the proximal ascending colon a strong contrast enhancing mass with intralumunal growth (2,5 x 1,9 x 3,2 cm), without direct contact with the ileocecal valve or infiltration of the pericolic fatty tissue (figure 3). No pathological lymph nodes or liver metastasis were noticed. Chest CT scan showed no intrathoracic metastasis.

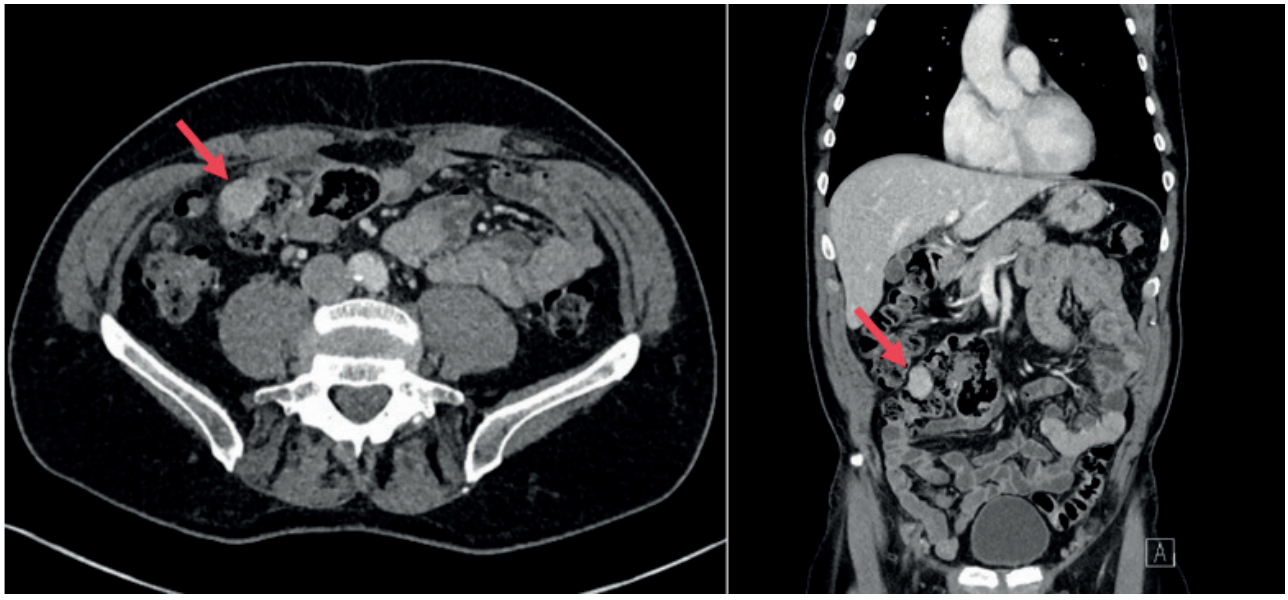

Figure 3: Abdominopelvic computerized tomography demonstrating a well-circumscribed and contrast-enhancing lesion (2.5 x 1.9 x 3.2 cm), round-shaped mass with intraluminal growth, at the ascending colon in axial (left) and coronal (right) views.

As the tumor could only be partially removed endoscopically with inconclusive histopathological finding, a surgical removal was decided.

The procedure started laparoscopically. The ink marked lesion was found in the hepatic flexure of the colon, and no peritoneal carcinomatosis or liver metastasis were noted. After releasing the hepatic flexure and mobilization of the right colon, a transversal mini-laparotomy was made in the right abdomen, for subsequent exteriorization of the colon. A wedge resection of the lesion was performed with closure of the intestinal defect with continuous 4/0 PDS suture.

The histopathological examination of the resected colonic specimen reported an ulcerated mass with acute granulating inflammation, compatible with a 2cm schwannoma, with proliferative activity (Ki 67) below 3%, without signs of malignancy or involvement of the resection margins (R0 resection). The tumor tissue was immunohistochemically positive for S100, SOX-10 and vimentin and negative for CD117, DOG-1, STAT6, SMA, Desmin, CD34 and MNF116 (figure 4).

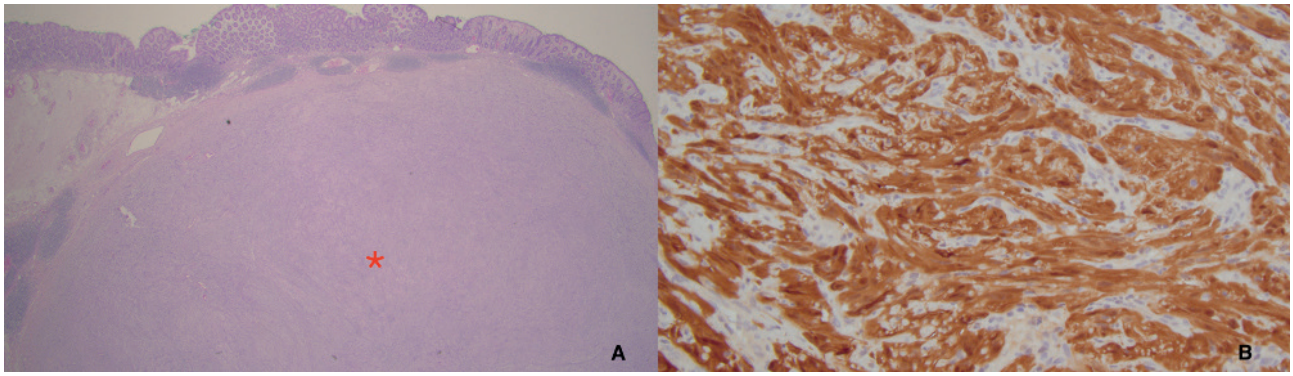

Figure 4: Microscopic findings of the surgical specimen: (A) Hematoxylin-eosin staining showing a spiky tumor tissue with elongated bland cell nuclei and abundant fibrillar cytoplasm. Cell nuclei partially arranged on a palisade, without necrosis or mitoses. (B) Immunohistochemically positive for S100. (Pictures by courtesy of Urs Lüthi, MD, Pathologie Länggasse, Bern, Switzerland).

The postoperative course was uneventful, and the patient could be discharged on the third postoperative day.

Discussión

Mostly schwannomas are asymptomatic,[5] and are discovered on screening endoscopy or incidentally on abdominal imaging performed for another reason. [4] However, just like any other gastrointestinal tumors, they can present with symptoms such as abdominal pain, as in our case, tenesmus, rectal bleeding or melena.[3,6] Sometimes these tumors manifest as colonic obstruction or intussusception.[3] No specific tumor marker exists either.[3]

Schwannoma is frequently diagnosed as a submucosal mass or polyp with a smooth surface but in rare cases can ulcerate into the mucosa.[4]

Histopathological examination of preoperative colonoscopic mucosal biopsies alone, provides usually limited information. Only 15% of the colonic schwannomas are diagnosed on preoperative endoscopic biopsy.[3]

For the differential diagnosis of gastrointestinal schwannomas, abdominal CT scan can help to differentiate between schwannomas and other mesenchymal tumors [4], which did not happen in our case. Schwannomas usually present as exophytic masses with homogeneous enhancement and cystic change, necrosis, or calcification within tumors are uncommon.[3]

The definitive diagnosis is made on immunohistopathologic examination of the operative specimen.[4]

Macroscopically, schwannomas tend to be lobulated well-defined tumors with a cystic pattern and, rarely, may be hard, solid, ulcerated or calcified. [4,5]

Histopathological features of schwannomas are mainly elongated bipolar spindle cells with variable cellularity and sometimes peripheral cuff-like lymphocyte infiltration is exhibited around the tumor, which helps to differentiate schwannomas from other spindle-cell tumors.[3] Furthermore, they stain positive for S100 proteins, and occasionally for vimentin, and stain negative for SMA, Desmin, CD117, and P53.[4]

Even though schwannomas are reported as benign in more than 98% of cases, malignant transformation can occur in 2% of cases,[2] with the occurrence of loco-regional or distant metastasis in aggressive tumors. [4] High mitosis rate (>5 mitoses per field), high Ki-67 proliferative index (≥5 %)5, and large tumor size (larger than 5 cm) are considered to be associated with an higher risk of metastasis and/or recurrence.[4,3]

The mainstay of treatment is a complete surgical resection with negative margins.[3] The surgical approach depends on the size, location and histopathological pattern of the tumor.[6]

Endoscopic resection is often suggested for tumors <3 cm in diameter. Commonly used techniques include endoscopic mucosal resection (EMR) and endoscopic full thickness resection (EFTR). However, since the tumor grows under the mucosa, the use of endoscopic resection increases the risk of bleeding, perforation and gastrointestinal fistula.[2]

Oncologic resection is not mandatory.[5] In reported cases, the observed high frequency of radical resection is due to the absence of accurate preoperative diagnosis.[4] When diagnosed preoperatively, schwannomas were resected either endoscopically or with a wedge resection.[4]

Routine adjuvant radiotherapy or chemotherapy is not recommended.[5] For cases with malignant transformation, the current guidelines remain controversial und there is still a lack of evidence.[1] Bevacizumab, an antibody against vascular endothelial growth factor (anti-VEGF), is one of the few drugs used for the treatment of malignant schwannomas and has been used in vestibular schwannomas. Currently, the effectiveness of molecular therapy in colonic schwannomas is unclear because of the very low number of reported cases. Further molecular therapy research is mandatory to determine its efficacy und the rationality of its use in the treatment of malignant schwannomas.

Conclusion

Colonic schwannoma is a rare, usually benign tumor,[4] whose preoperative diagnosis is challenging because clinical manifestations, as well as colonoscopic and CT findings, are nonspecific.[3]
Conclusive diagnosis can be made by confirming S-100 proteins in the immunohistochemical analysis of the operative specimen.[4,3] Given the risk of malignant transformation, surgery is the mainstay of treatment,[1] and as for other mesenchymal tumors, wedge resection rather than classic regional resection is advised.[4]

Author's Statement

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— Consent: Consent of the patient was obtained for publication of this case.
— Ethical Responsibilities: Protection of people and animals. The authors declare that no experiments have been performed on humans or animals for this research. Data confidentiality. The authors declared that they have followed the protocols of their work center regarding the publication of patient data. Right to privacy and informed consent. The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.
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