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Case Report

Abdominal Schwannomas: Case Report With Literature Review

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Schwannomas are rare tumors that arise from Schwann cells in neural sheaths. They are commonly found in the central nervous system, spinal cord, or peripheral nerves of the body. Occasionally, they occur in the gastrointestinal tract, with the stomach being the most common site. However, colorectal and retroperitoneal schwannomas are very rare. Preoperative diagnosis is often difficult and definitive treatment entails surgical excision. We herein present 3 cases of intraabdominal schwannomas.

Key words: Benign – schwannoma – tumor

Gastrointestinal and retroperitoneal schwannomas are rare benign tumors with reported malignant potential.[1–3] The stomach is the most common site of gastrointestinal schwannoma, while the colon and rectum are relatively uncommon.[4–6] Intraabdominal schwannomas occur equally in men and women, with a wide age range.[1] Diagnosis is usually made only after excision and histologic examination owing to nonspecific radiologic and endoscopic appearances. Definitive treatment entails complete surgical excision with negative margines. We herein present 3 cases of intra-abdominal schwannoma.

Case Report 1

A 63-year-old lady with past medical history of hypertension and previous hysterectomy for fibroids presented to us with 6-month-duration of early satiety and loss of appetite. A computerized tomography (CT) scan of the abdomen/pelvis was performed, and a sigmoid polyp was detected. Colonoscopy was done, and it showed a large 4-cm, broad-base polyp in the upper rectum (Fig. 1). Biopsy of the polyp showed mucosal tissue with mild hyperplastic changes only. Magnetic resonance imaging (MRI) of the rectum revealed extension of the rectal lesion beyond the serosa with possible lymph node metastasis. The patient was then scheduled for anterior resection surgery. Intraoperatively, the low rectal tumor was well circumscribed without local invasion. Postoperatively, the patient had an unremarkable recovery, and she was discharged home well on the fourth day after surgery.

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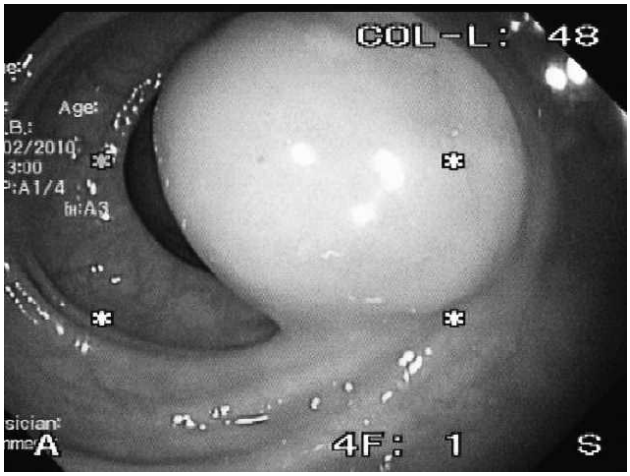

Fig. 1 Submucosal lesion seen in upper rectum on endoscopy.

Microscopically, hematoxylin and eosin (H&E)-stained sections of the lesion show a predominantly well-circumscribed, submucosal, moderately cellular, spindle-cell tumor arranged in interlacing bundles and short fascicles and intermixed with wavy collagen fibers. Immunohistochemical studies revealed positive diffuse staining for S100, while CD117, CD34, and smooth-muscle actin/desmin were negative (Fig. 2). All the lymph nodes resected were negative. A definitive postoperative diagnosis of rectal schwannoma was thus made. She is well at 3 years follow-up.

Case Report 2

A 43-year-old gentleman presented with abdominal discomfort with no complaints of vomiting or any other constitutional symptoms. Physical examination was unremarkable. He underwent a CT scan of the abdomen and pelvis, which revealed a 3.1×3.0-cm, well-defined, retroperitoneal soft tissue mass in close proximity to the superior mesenteric vessels with a feeding vessel from the superior mesenteric artery (SMA) and venous drainage to the superior mesenteric vein (SMV) (Fig. 3). This mass lesion was distinct from the pancreas and bowel loops.

The lesion was biopsied under CT guidance. It was strongly positive for S100 and negative for CD117, SMA, and desmin and thus confirmed to be a schwannoma.

The patient underwent laparoscopic excision of the mass. Postoperative recovery was uneventful. He is well at 15 months follow-up.

Case Report 3

A 52-year-old lady presented with upper abdominal discomfort. Physical examination was unremarkable. CT scan revealed a large polypoidal lesion in the lumen of the first part of the duodenum, about 27 mm in diameter. It was well circumscribed, rounded, and showed contrast enhancement. The rest of the imaged bowel was normal. She was then subjected to an upper gastrointestinal endoscopy and endoscopic ultrasonography (EUS), which confirmed a 3-cm submucosal nodule. The biopsy was nondiagnostic.

She underwent distal partial gastrectomy with duodenectomy and a Roux-en-Y reconstruction. Recovery was uneventful and she was discharged on the fifth postoperative day. The histology was reported as schwannoma with uninvolved surgical margins. It was positive for S100 and negative for CD117, CD34 and SMA. She is well at 12-month follow-up.

Discussion

Schwannomas are rare tumors that arise from Schwann cells in neural sheaths. They are usually found in the central nervous system, spinal cord, or peripheral nerves of the body, with the most common type being acoustic neuroma.[7] Occasionally, schwannomas occur in the gastrointestinal tract. Together with leiomyoma, leiomyosarcoma, and gastrointestinal stromal tumors (GIST), they constitute mesenchymal tumors of the gastrointestinal tract.[8] Gastrointestinal schwannomas account for 0.4% to 1% of all submucosal tumors of the gastrointestinal tract[9–11] and most commonly occur in the stomach.[4] Large intestine and retroperitoneal schwannomas on the other hand are rare.[5,6] Gastrointestinal schwannomas occur equally in men and women, with a wide age range of 18 to 87 years[1] and median age of 65 years.[5] They generally run a benign course.[12–17] However, isolated cases of "malignant schwannomas" have been reported.[2,18] Whether these malignant tumors arise *de novo* or from benign schwannomas remains controversial.[19] Currently, such malignant tumors with neural differentiation are regarded by most pathologists as distinct from gastrointestinal schwannomas and are termed gastrointestinal autonomic nerve tumors (GANTs).[20]

Colonic schwannomas generally grow slowly and are mostly asymptomatic.[21] Occasionally, they may cause rectal bleeding, intestinal obstruction, defeca-

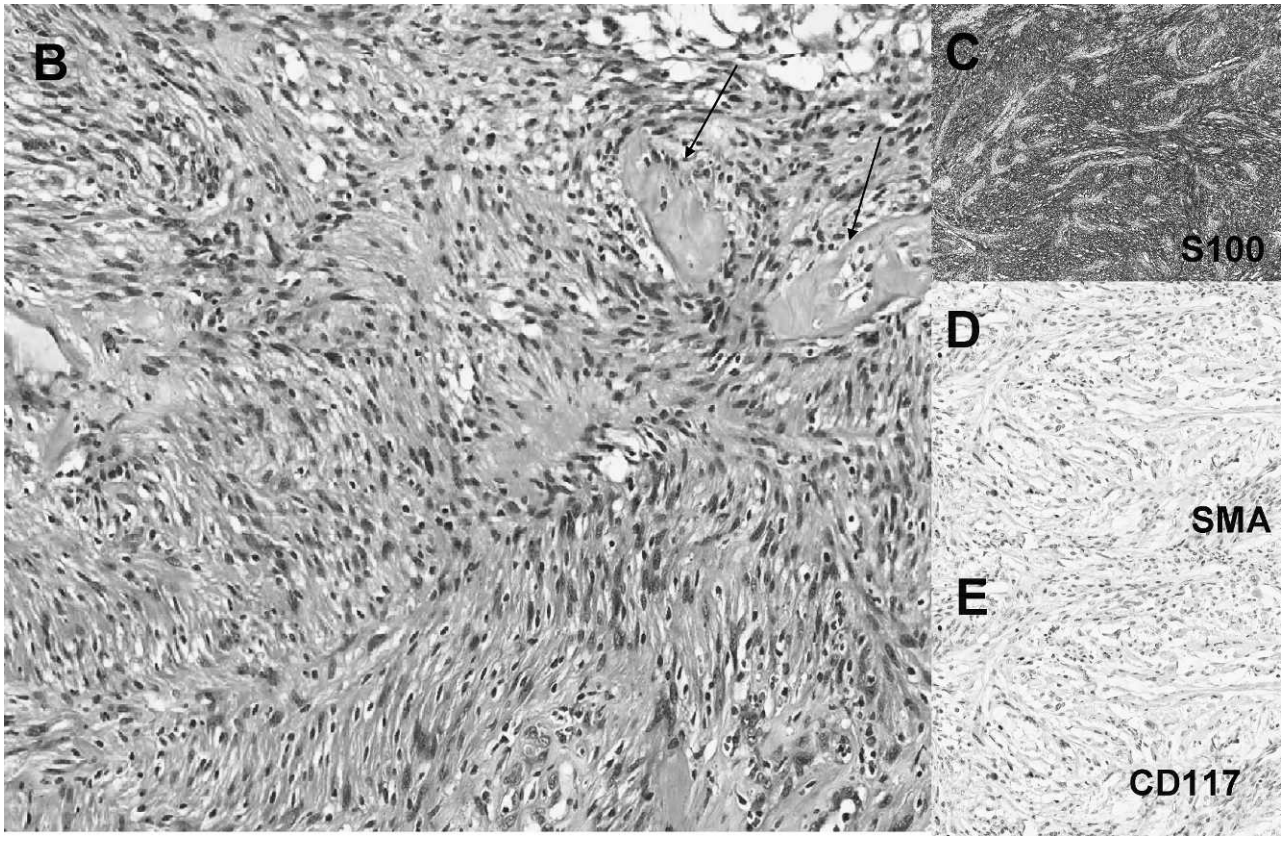

Fig. 2 High power showing fascicles of bland spindle cells with palisading and hyalinized blood vessels (arrows). (H&E, ×40). (C–E.) Inset shows positive S100 immunohistochemical staining confirming the diagnosis of Schwannoma, whilst the other markers, smooth-muscle actin and CD117, are negative, excluding smooth-muscle tumor and GIST.

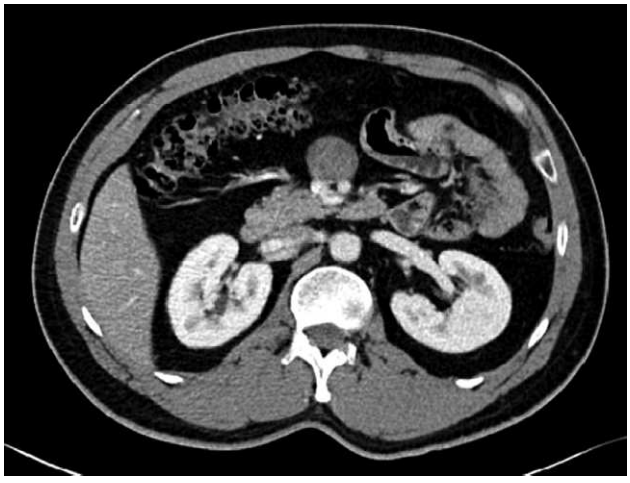

Fig. 3 CT image showing the mass lesion in the retroperitoneum just inferior to the pancreas and in close proximity to the superior mesenteric vessels.

tion disorders, or pain.[5,22] Retroperitoneal schwannomas may present with vague abdominal symptoms depending on their size and location. Upper gastrointestinal schwannomas may present with vomiting, early satiety, abdominal bloatedness, or intestinal perforation. However, these symptoms are not specific to schwannomas. Schwannomas can be discovered incidentally during laparotomy and on radiologic scan.[23] In all 3 cases, the patients had nonspecific complaints, and the lesions were detected on CT. Computed tomography may not be diagnostic, and final diagnosis is often made postoperatively by specimen microscopic examination.

Accurate preoperative diagnosis of gastrointestinal schwannomas is often difficult owing to the rarity of this condition. It is also in part due to the lack of pathognomonic endoscopic features of gastrointestinal schwannomas.[24,25] They usually appear as submucosal lesions, indistinguishable from other mesenchymal tumors. Furthermore,

being typically hard and solid tumors, attempts to obtain good biopsy samples are often technically challenging,[26] as was the case with our patients. EUS examination is a useful tool in the preoperative assessment of gastrointestinal submucosal tumors.[27–29] Besides allowing for accurate EUS-guided fine-needle aspiration or biopsy of the lesion, it helps to define the borders of the tumor and its localization within the gastrointestinal wall. However, there are no typical EUS features of gastrointestinal schwannomas because of their plexiform pattern of growth.

Histologically, schwannomas are made up of spindle-shaped cells with intertwining cytoplasmic extensions, seen on both light and electron microscopy. Two histologic subtypes have been further described: (1) Antoni type A with densely packed spindle cells (Verocay bodies), (2) and Antoni type B with loosely organized spindle cells (absence of Verocay bodies) in myxoid stroma.[5,22] Immunohistochemical studies are required to differentiate between the different types of spindle-cell tumors. Positive desmin and muscle actin stains indicate smooth-muscle lesions like leiomyoma or leiomyosarcoma, while positive CD34 and CD117 indicate GIST. Strongly positive S100 specimens would point towards a schwannoma. In the case of our patients, pathologic examination of the lesion revealed spindle cells, strongly positive for S100 and negative for CD117, CD34, and smooth-muscle actin/desmin, confirming the diagnosis of schwannoma.

Although benign, definitive treatment of gastrointestinal schwannomas in fit patients entails surgical excision with clear margins, as it is frequently impossible to distinguish these tumors from other mesenchymal tumors, which are malignant or have malignant potential. In addition, the response to chemotherapy and radiotherapy remains uncertain.[30] The surgical approach depends on the location and size of the tumor. As the lesion is usually well encapsulated and does not exhibit aggressive malignant behavior, it can be amenable to minimally invasive modalities, besides commonly used open surgical techniques. This includes transanal endoscopic microsurgery (TEM), whereby local excision is accomplished through a rectal expander[31,32] with or without the use of an ultrasonically activated scapel.[33] TEM was not offered to our patient (Case 1) owing to suspicion of lymph node metastasis on MRI imaging. We also did not attempt laparoscopic resection in the first case in view of previous pelvic surgery. However, the patient with the retroperitoneal mass underwent laparoscopic excision of the same. Endoscopic submucosal dissection can be performed for gastric schwannomas with a confirmed diagnosis on histology. We performed a local resection to achieve negative margin for a preoperatively suspected duodenal GIST and final histology revealed a schwannoma.

In conclusion, gastrointestinal schwannoma is a rare benign tumor of the gastrointestinal tract that is usually asymptomatic. Preoperative diagnosis is often difficult, and definitive treatment involves surgical resection with clear margins. The outcome after surgery is excellent as these lesions generally run a benign course.

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