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CASE REPORT | STOMACH

Solitary Fibrous Tumor of the Stomach

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ABSTRACT

Solitary fibrous tumor is a rare mesenchymal neoplasm that usually originates from the pleura, but has been reported in other extrapleural locations. We report a rare case of a solitary fibrous tumor of the stomach, which was successfully treated with endoscopic mucosal resection.

INTRODUCTION

Solitary fibrous tumors (SFTs) were first described in pleura in 1931.[1] Since then, nonpleural SFTs have been described as originating from almost every anatomic location of the human body, but reports of SFTs in the abdominal cavity are rare.[2] Five SFT cases arising from the stomach have been reported as of November 2016.[3-7]

CASE REPORT

A 55-year-old woman was referred for an upper endoscopy for new-onset dyspepsia and gastroesophageal reflux symptoms. The patient had a history of *Helicobacter pylori* gastritis, which had been successfully treated 1 year prior with triple therapy. Her physical examination was unremarkable. Initial biochemical and hematological laboratory studies were within normal limits.

Esophagogastroduodenoscopy (EGD) revealed a 1-cm nodule in the gastric antrum along the greater curvature with normal overlying mucosa (Figure 1). A contrast-enhanced computed tomography scan of the abdomen did not detect any gastric lesions, enlarged lymph nodes, or abnormal findings in other organs. On endoscopic ultrasound (EUS), the lesion appeared to be isoechoic with hyperechoic foci measuring 7.1 × 6.7 mm (Figure 2). The endoscopy and EUS findings indicated that the lesion was subepithelial. An uneventful endoscopic mucosal resection (EMR) of the lesion was performed using the Captivator™ EMR banding kit (Boston Scientific, Natick, MA; Figure 3). The patient was discharged the same day of the procedure on proton pump inhibitor (PPI) therapy for her dyspepsia.

Pathologic analysis of the resected specimen revealed negative margins and normal gastric mucosa with an underlying subepithelial tumor. The tumor was composed of bland and uniform oval to spindle cells dispersed along thin parallel collagen bands. The cells had minimal cytoplasm, small elongated nuclei, and indistinct nucleoli. Additionally, staghorn-type vessels and perivascular sclerosis were also present (Figure 4). Immunohistochemical staining was positive for CD34 and bcl-2 and was negative for S-100, DOG-1, CD-117, desmin, and ALK. These findings were consistent with the diagnosis of a gastric SFT. The patient was immediately informed about the diagnosis and was educated regarding the long-term follow-up of her lesion. At the 6-month follow-up, the patient had improved clinically and PPI was tapered off. Repeat upper endoscopy showed no recurrence of the gastric SFT lesion.

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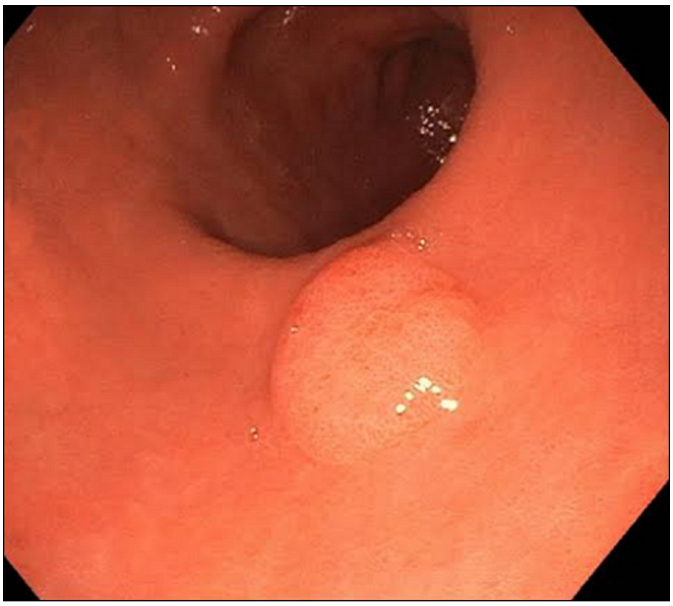

Figure 1. EGD demonstrating a 1-cm diameter nodule in the gastric antrum along the greater curvature.

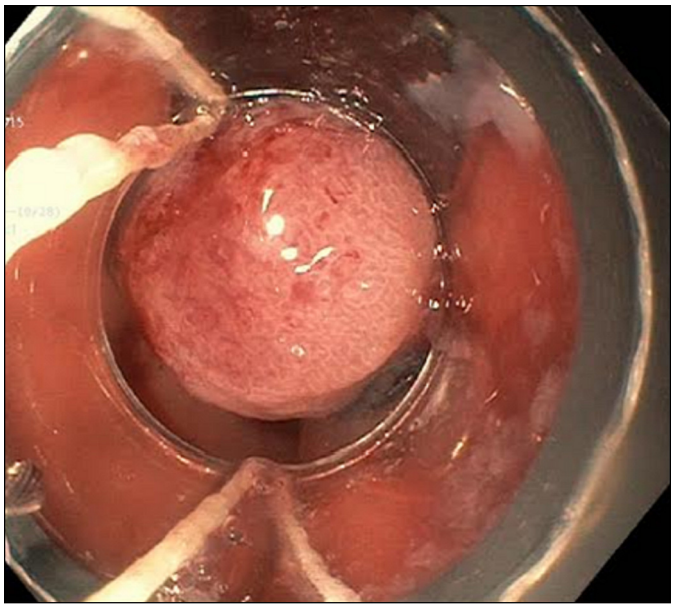

Figure 3. Endoscopic mucosal resection of the gastric nodule.

DISCUSSION

SFT is a rare mesenchymal neoplasm often originating from the pleura, but it has been reported in other extrapleural locations such as the pelvis, abdomen, retroperitoneum, buccal space, maxillary sinus, liver, pancreas, suprarenal region, and kidneys.[1-3] Although the true incidence is unknown, pleural SFTs have been estimated to occur with a frequency of 2.8 per 100,000 individuals.[3] It is frequently encountered in middle-aged people, and it is reported with an equal distribution between genders.[4] The symptoms are usually non-specific and vary depending on the lesions. The diameter of SFTs is usually >8 cm.[4] Unfortunately, tumor markers are not specific and sensitive for SFT. A high expression of serum insulin-like growth factor-II was reported in patients presenting with hypoglycemia.[2-4]

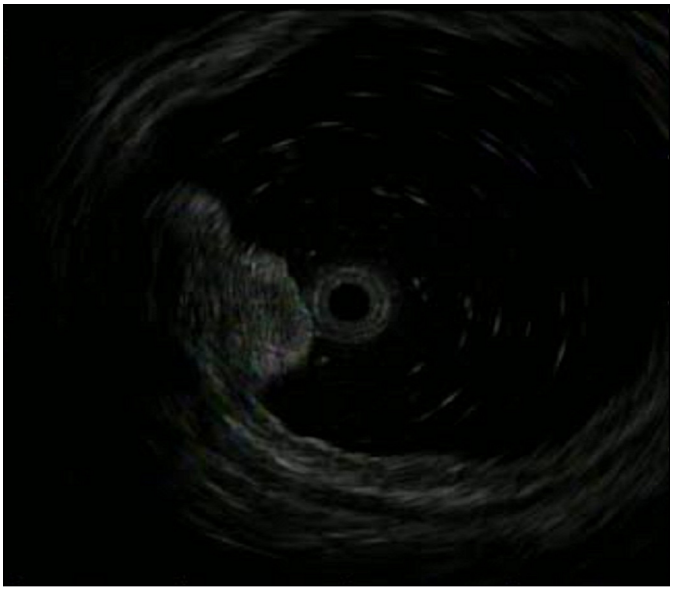

Figure 2. EUS showing an isoechoic, subepithelial gastric nodule with hyperechoic foci.

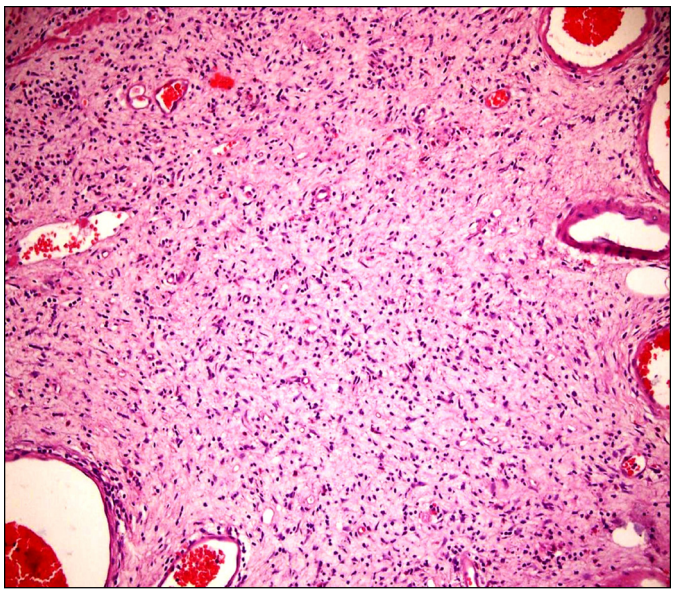

Figure 4. Histopathologic analysis of gastric subepithelial tumor showing bland, uniform oval to spindle cells dispersed along thin parallel collagen bands. The tumor cells had minimal cytoplasm, small elongated nuclei, and indistinct nucleoli. Staghorn vessels with perivascular sclerosis were also present (hematoxylin and eosin stain, 200x).

SFT with the involvement of stomach is a rare clinicopathologic entity. To our knowledge, only 5 cases of gastric SFTs have been reported to date.[5-9] The data on previously reported cases is summarized in Table 1. Three patients with gastric SFTs were female and 2 were male, with a median age of 56 years at the time of diagnosis and a median tumor size of 5.3 cm. Gastric SFTs were diagnosed incidentally in 3 cases. Patients with intra-abdominal SFT most commonly present with a palpable abdominal mass and may complain of vomiting, abdominal pain, or weight loss. However, incidental SFTs, as in the present case, are small tumors that are typically asymptomatic.[4-9] The differential diagnosis of gastric SFT includes gastrointestinal stromal tumor, calcifying fibrous tumor, fibromatosis, schwannoma, leiomyoma, leiomyosarcoma, inflammatory myofibroblastic tumor, fibrosarcoma, malignant fibrous histiocytoma, hemangiopericytoma, synovial sarcoma, and malignant mesenchymoma.[2-4]

Immunohistochemically, SFTs commonly express CD34, CD99, bcl-2, and STAT-6. They are usually negative for S-100 and epithelial membrane antigen (EMA).[10] Although there are no pathognomonic radiologic features that are specific for this tumor type, computed tomography (CT) and magnetic resonance imaging (MRI) are frequently used to diagnose SFTs.[2] The lesions often appear homogenous on computed tomography and are enhanced after contrast administration. On MRI, SFTs typically display low T1 signal intensity and variable T2 signal intensity, and they enhance intensely after intravenous gadolinium administration.[2,3,11] Needle aspiration biopsy for SFTs usually provides inconclusive results. However, histopathologic and immunohistochemistry findings are key to establish a definitive diagnosis. In our review of gastric cases, all 5 tumors were positive for CD34 and 3 were positive for vimentin. All 5 patients were negative for SMA, and desmin stained negative in 4 patients. In most of the patients with gastric SFTs, the diagnosis was established by a combination of clinical suspicion, imaging findings, and histopathology, and was confirmed with immunohistochemical staining.

SFT can be an aggressive disease, and relapses can occur several years from diagnosis.[11] The malignant potential of SFTs can be assessed on the basis of mitotic activity (>4 mitotic figures per 10 high-power fields), the presence of necrosis or hemorrhage, tumor size >10 cm, increased cellularity, nuclear pleomorphism, and stromal or vascular invasion.[2] Assessment of metastatic disease in SFTs is primarily done with CT, MRI, and positron emission tomography/CT. However, accurate prediction criteria for clinical behavior of SFTs are not available to date. One study demonstrated that 10–20% of SFTs show malignant histologic features and propensity for local recurrence or metastasis.[2] According to a retrospective analysis of 33 patients, histologic features of malignancy were found in 54% of extrathoracic SFTs, and the

Table 1. Clinical characteristics of the 5 reported cases of SFT originating from the stomach.

Patient Age (years)	Gender	Size (cm)	Immunohistochemical Analysis (positive/negative)	Mitotic Count	Local Recurrence/ Metastasis	Treatment	Status at Last Follow-up	Reference
77	Female	3	CD34, vimentin, SMA/desmin, pancytokeratin, S-100, CD31, bcl-2, EMA	1/10 HPF	No	Careful monitoring	NED	[5]
70	Male	8.5 × 7 × 6	CD34, vimentin/pancytokeratin, desmin, SMA, S-100, CD99, CD117	0/10 HPF	No	Wedge resection of the stomach	NED	[6]
26	Male	5.4 × 5.2 × 4	CD34, S-100/CD117, SMA, desmin	0/10 HPF	No	Wedge resection of the stomach	NED	[7]
43	Female	2.7 × 2 × 1.5	CD34, bcl-2, MIC-2/SMA, CD117, DOG-1, ALK-1, and EML4-ALK	0/10 HPF	No	Wedge resection of the stomach by LECS	NED	[8]
65	Female	2.5 × 2.3 × 1	CD34, vimentin/desmin, DOG1, PDGFRA, SMA, S-100, CD99, CD117	less than 3/10 HPF	No	Surgical excision	NED	[9]

Abbreviations: ALK-1, activin receptor-like kinase 1; bcl-2, B-cell lymphoma protein 2; DOG1, discovered on GIST-1; EMA, epithelial membrane antigen; EML4-ALK, echinoderm microtubule-associated protein-like 4-anaplastic lymphoma kinase; HPF, high-power field; LECS, laparoscopic and endoscopic cooperative surgery; NED, no evidence of disease; PDGFRA, platelet-derived growth factor receptor alpha; SFT, solitary fibrous tumor; SMA, smooth muscle actin.

rate of metastasis was 39%.[12] A recent retrospective study of ncluded 139 patients with pathologically proven SFT showed that 49 of them developed metastases.[13] The tendency of metastatic disease was shown to be higher in patients with extrathoracic SFT than those with thoracic SFT.[13] Various sites of metastasis have been described in the literature, including lungs, mediastinum, liver, bone, pancreas, kidney, mesentery, and retroperitoneum.[9-13] Generally, malignant SFTs have a poor prognosis.[11-13] Therefore, it is a matter of urgency to reach a consensus to clarify the malignant propensity of SFTs to diagnose such patients early and institute an effective treatment. In the present review, all 5 cases of gastric SFT were benign on histopathologic analysis.

In the management of localized SFT lesions, complete en bloc surgical resection is the mainstay of treatment.[13] The standard of care is complete surgical resection with negative margins even for high-risk tumors, given the low overall metastatic potential. In advanced disease, traditional cytotoxic sarcoma chemotherapy has been used with limited benefit to patient outcomes. In newer case series, vascular endothelial growth factor inhibitors and other tyrosine kinase-inhibiting molecular agents such as bevacizumab, sunitinib, pazopinib, and sorafenib suggest some promise for therapy of advanced SFT.[14] However, there is no standard therapeutic strategy for advanced SFT lesions.

Our patient's gastric SFTs were treated with surgical resection. The curative wedge resection of the stomach was performed in 3 of 5 cases.[5-9] The patients showed a remarkable response with no recurrence on follow-up, even without any specific maintenance therapy. In our patient, EMR was used to resect the SFT lesion. Therefore, to our knowledge, our case represents the first report of an exceptionally small gastric SFT indicated by EUS, confirmed with pathologic analysis, and successfully treated with EMR. Hence, curative surgical or endoscopic resection should be recommended in patients with gastric SFT.

Gastroenterologists should consider the possibility of gastric SFT when an exceptionally small subepithelial nodule is encountered during routine endoscopy. Considering unpredictable biological behavior and the possibility of recurrence, a long-term clinical and endoscopic follow-up may be necessary as standard guidelines are not available at the present time.

DISCLOSURES

Author contributions: F. Inayat reviewed the literature, designed the study, wrote the manuscript, and is the article guarantor. Q. Hussain and A. Hurairah reviewed the manuscript. K. Shafique acquired the histopathologic image. EB Grossman reviewed the final version of the manuscript.

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