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Case Report

CASE REPORT

Gastric schwannoma—a rare benign mimic of gastrointestinal stromal tumor

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Abstract

Schwannomas most commonly manifest as acoustic neuromas in the vestibulo-cochlear nerve (CN VIII). These may occur unilaterally as primary tumors, or bilaterally secondary to neurofibromatosis type 2. More rarely, they present in extracranial sites, including the gastrointestinal tract. Gastrointestinal schwannomas are believed to arise from Auerbach's plexus in the muscularis propria, and are classified as mesenchymal tumors. Here, we report a rare case of a 49-year-old woman who had surgical resection of a gastric mass which was eventually diagnosed as gastric schwannoma.

INTRODUCTION

Mesenchymal tumors of the gastrointestinal (GI) tract are a group of tumors that can arise from an array of different tissue types. These include leiomyomas, GI stromal tumors (GIST), lipomas, hemangiomas and the aforementioned, schwannomas [1]. Each of these tumors has defining characteristics, but can often be misdiagnosed. Schwannomas, for example, can be easily mistaken for a GIST or a leiomyoma, when visualized on upper endoscopy [2]. What makes each tumor distinct are their histologic features, and more specifically, what immunohistochemical (IHC) markers they stain positively to. It is important to make this distinction, because GISTs have the potential for malignant conversion, while Schwannomas are benign and carry a good prognosis [3].

CASE REPORT

A 49-year-old woman with a medical history of hypertension, obesity and chronic kidney disease presented to the gastroenterology clinic complaining of heartburn, nausea, decreased appetite, 10 pound weight loss and sore throat of 4 months duration. The patient was found to have macrocytic anemia with hemoglobin of 9.2 g/dL and mean corpuscular volume (MCV) of 107.5, most likely attributable to her vegetarian diet. On this initial visit, the patient was started on a proton pump inhibitor (PPI) 40 mg orally once daily for 8 weeks. Twelve weeks later, the patient returned to the clinic with mild improvement of symptoms, but disclosed that she had an episode of bloody emesis. In response to this finding, an esophagogastroduodenoscopy (EGD) was scheduled which revealed mild chronic gastritis, negative for *Helicobacter pylori* and a submucosal mass in the gastric body (Fig. 1). Biopsy of the mass showed no evidence of malignancy. A presumptive diagnosis of GIST was made. Computerized tomography (CT) scan of the abdomen was obtained to further evaluate the gastric mass which showed an exophytic $3.2 \times 3.7 \times 4.1\,cm^3$ solid homogeneous mass along the greater curvature of the stomach. An endoscopic ultrasound (EUS) was scheduled to further evaluate the depth of the lesion. EUS showed a $3.5 \times 3.0\,cm^2$ lesion in the body of the stomach, originating in the muscularis propria (Fig. 2). Fine needle aspiration (FNA) was attempted, but was unable to be

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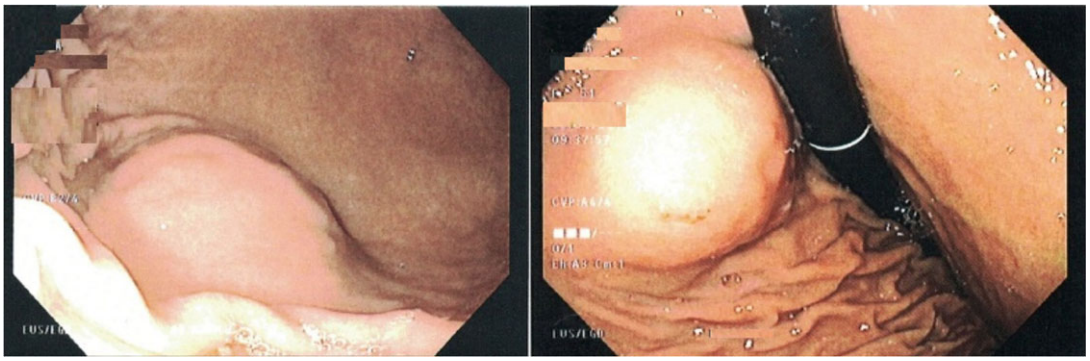

Figure 1: Esophagogastroduodenoscopy (EGD) showing a submucosal mass in the gastric body

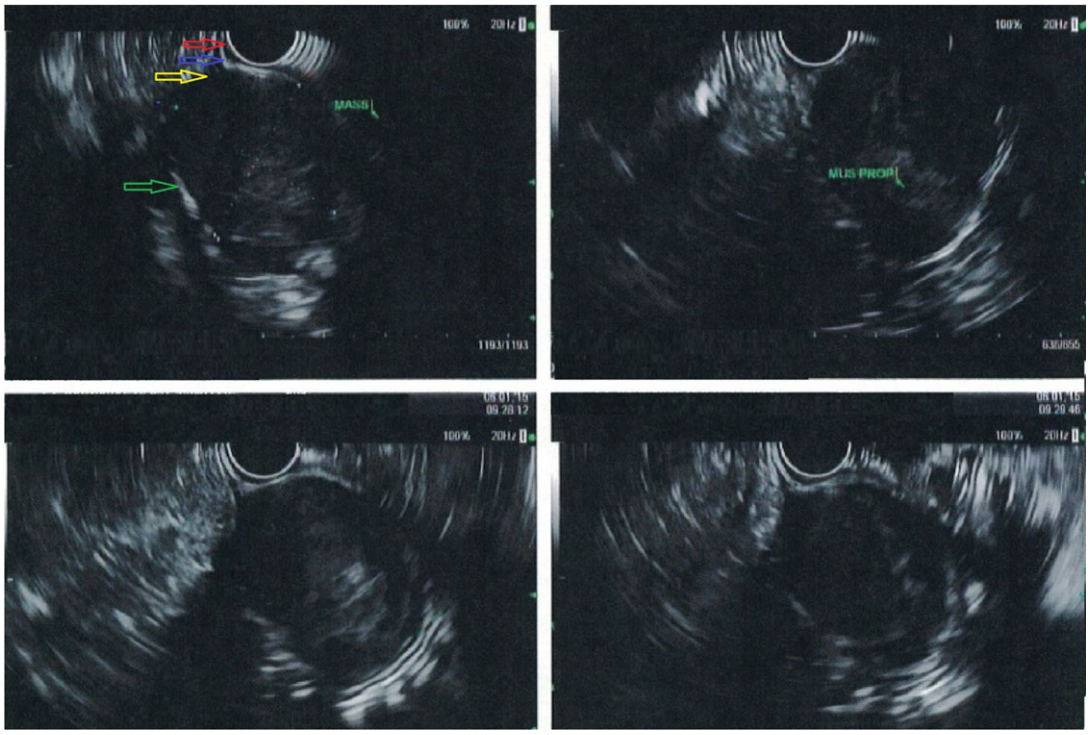

Figure 2: Endoscopic ultrasound (EUS) showing a 3.5 × 3 cm^2 mass arising from muscularis propria in the body of the stomach (yellow arrow). The other layers identified are muscularis mucosa (red arrow), submucosa (blue arrow) and serosa (green arrow)

accomplished due to intervening blood vessels. Due to the failed FNA and size of the mass, the patient was referred to surgical clinic for possible surgical resection. The patient underwent successful resection of the mass and the specimen was sent to pathology. To our surprise, the results were negative for GIST or leiomyoma. Rather, the specimen stained positive for S-100 confirming the diagnosis of gastric schwannoma. Biopsy shows the location of the schwannoma with relation to the overlying epithelial layer (Fig. 3), schwannoma demonstrating spindle cell histology (Fig. 4) and spindle cells stained under S-100 confirming schwannoma (Fig. 5). Patient was followed up in the gastroenterology clinic 8 weeks after the surgery at which point patient denied any GI symptoms.

DISCUSSION

GI schwannomas are a rare entity, comprising only 0.2% of all gastric tumors [4]. A recent study compiled 51 documented cases of gastric schwannomas in the United States from 1970 to 1999. Their results showed a female to male ratio of ~1.4–1.0, with a mean age at presentation of 58 [5]. These findings are consistent with our case, in which a gastric schwannoma presented in a 50-year-old female. Symptoms, when present, can range from mild dyspepsia, to hematemesis and weight loss. The bleeding is thought to occur due to ulceration of the mass from reduced tolerance to gastric acid [6].

Gastric Schwannomas are most often encountered as incidental findings on upper endoscopy. Their appearance is identical to other mesenchymal tumors in that they are well circumscribed with intact overlying mucosa, and can grow in an exophytic or endoluminal fashion [1]. They may also be encountered by other diagnostic modalities including CT scan. When visualized on CT scan, gastric schwannomas are round, well defined, mural masses with homogeneous attenuation [7]. EUS is another diagnostic modality, which may also serve as a therapeutic tool. Through EUS, one can obtain a biopsy through

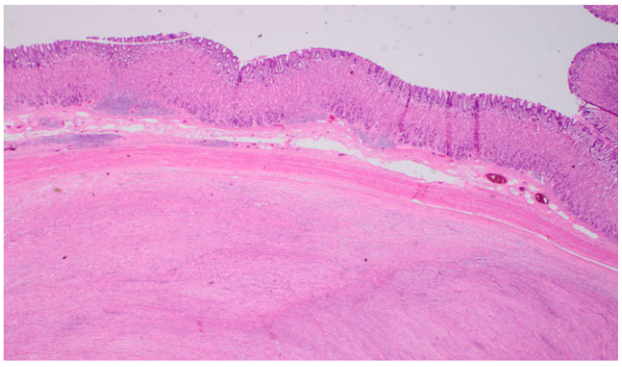

Figure 3: Biopsy (hematoxylin and eosin stain) showing schwannoma with relation to epithelial layer

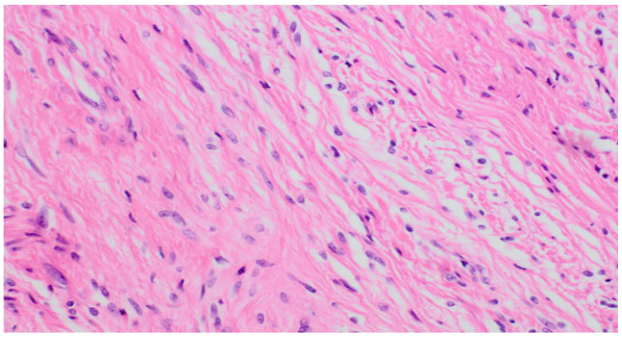

Figure 4: Biopsy (hematoxylin and eosin stain) with schwannoma demonstrating spindle cell histology

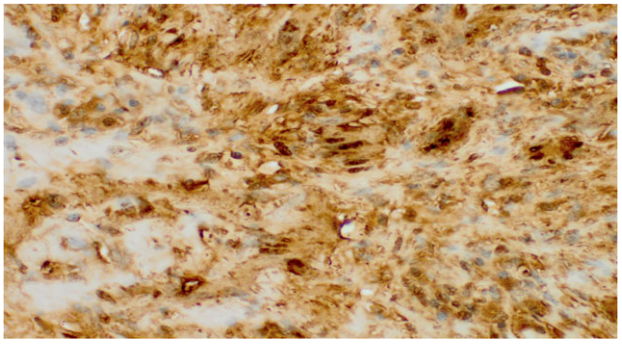

Figure 5: Biopsy showing schwannoma staining positive for S-100 stain

FNA, and assess the depth of the mass. Depending on the depth and size of the mass, one can safely treat, through endoscopic resection [8].

Histologically, GI schwannomas are characterized by spindle cells with wavy cigar shaped nuclei and collagen. They seldom present with the classic Antoni A and Antoni B patterns that are typical of acoustic neuromas [9]. Interestingly enough, spindle cells are also characteristic of GI leiomyomas and GISTs. This, coupled with their near identical appearance on upper endoscopy, underscores the importance of IHC markers in making a definitive diagnosis. These markers include S-100, CD117 (c-kit), desmin and smooth muscle actin (SMA). GISTs stain positively to CD117 (c-kit); leiomyomas stain positively to desmin and SMA; and schwannomas stain positively to S-100 [10]. Treatment of GI schwannomas depends on the size and depth of the mass. New advances in gastroenterology have increased the role that endoscopy plays in the treatment of GI tumors. Techniques such as endoscopic submucosal excavation, endoscopic full thickness resection and submucosal tunneling endoscopic resection provide a non-invasive, safe way to treat sub-epithelial tumors [11]. Despite these new developments, tumors that lie within the muscularis propria, or are >3 cm in size require surgical intervention, due to their high risk of perforation [8, 11]. Considering the fact that schwannomas originate in the Auerbach plexus of the muscularis propria, surgical resection is generally the treatment of choice. Our patient's schwannoma indeed originated in the muscularis propria, and measured >3 cm, making surgical resection the obvious choice. Recurrence of these tumors is also rare. In 2015, an intensive review of literature gathered 137 cases of gastric schwannomas. Of those cases, 105 were followed up, none of which showed recurrence up to 36 months post date of diagnosis [12]. Tumors that do demonstrate recurrence have been associated with incomplete resection at time of treatment [13].

CONCLUSION

Mesenchymal tumors of the GI tract have long been synonymous with GISTs, or leiomyomas. Although rare, schwannomas should be added to the differential diagnosis when encountering submucosal tumors on endoscopy or colonoscopy. Thanks to the development of IHC, the distinction between mesenchymal tumors can easily be made within minutes. Due to their benign nature and rare recurrence, the diagnosis of gastric schwannoma should provide the patient reprieve at time of discovery.

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CONFLICT OF INTEREST STATEMENT

None of the authors have any form of conflicts of interest.

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ETHICAL APPROVAL

This case report was approved by the institute's Institutional Review Board as per its policy.

CONSENT

Consent for participation was obtained from this patient.

GUARANTOR

Tagore Sunkara, MD.

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