

Gastric schwannoma: a benign tumor often misdiagnosed as gastrointestinal stromal tumor

Apurva S. Shah,¹ Pravin M. Rath,¹
Vaibhav S. Somani,¹ Astha M. Mulani²

¹Department of Gastroenterology; and

²Department of Obstetrics and
Gynaecology, Bombay Hospital and
Medical Research Institute, Mumbai, India

Abstract

Gastric schwannomas are rare mesenchymal tumors that arise from the nerve plexus of gut wall. They present with nonspecific symptoms and are often detected incidentally. Preoperative investigation is not pathognomonic and many are therefore misdiagnosed as gastrointestinal stromal tumors. We report a rare case of a 37-year old woman who underwent laparotomy for complex bilateral ovarian cyst with resection of gastric-gastrointestinal stromal tumor preoperatively, but confirmed to have a gastric schwannoma postoperatively. This case underscores the differential diagnosis of submucosal, exophytic gastric mass as schwannoma.

Introduction

Mesenchymal tumors of the gastrointestinal (GI) tract are mainly comprised of a spectrum of spindle cell tumors which include gastrointestinal stromal tumors (GISTs), leiomyoma or leiomyosarcoma, and schwannomas.¹ Among these neoplasms, GISTs are the most common and a great majority (60-70%) of them occur in the stomach.^{2,3} Schwannomas, contrastingly, are generally slow growing asymptomatic neoplasms that rarely occur in the GI tract. However, if found, the most common site is the stomach, accounting for 0.2% of all gastric tumors and 4% of all benign gastric neoplasms.⁴ Owing to their typical presentation as submucosal neoplasms with spindle cell histology, gastric schwannomas and GISTs appear grossly similar.^{2,5}

Both gastric schwannomas and GISTs occur predominantly in middle-aged persons and have no distinct clinical features.^{1,5,6} However, the prognosis for gastric schwannomas and GISTs is very different. As reported by Daimaru *et al.*, in 1988, gastric schwannomas are benign tumors with an excellent prognosis whereas 10-30% of GISTs have malignant behaviour. Hence, it is important to make an accurate diagnosis to optimally guide treat-

ment options. Computed tomography (CT) and upper gastrointestinal endoscopy are the mainstays of investigation although neither of these investigations is pathognomonic. Ultimately, the definitive diagnoses of GISTs and gastric schwannomas require immunohistochemical studies, which only can be performed on the surgical specimen.

In this paper we report a rare case of a 37-year old woman who underwent laparotomy for bilateral ovarian complex cyst with resection of gastric GIST preoperatively but confirmed to have a gastric schwannoma postoperatively.

Case Report

A 37-year old female was referred to gastroenterology department for an incidentally detected gastric mass. The reason for her hospitalization was per vaginal bleeding and lower abdominal discomfort since last two weeks. She was asymptomatic for gastric lesion and physical examination of the abdomen revealed no signs of tenderness or of a palpable mass. Her medical history was non-significant. She had history of previous two caesarean sections and umbilical hernia.

Routine blood investigations were unremarkable. She had undergone contrast-enhanced CT of abdomen and found to have complex bilateral ovarian cysts with focal nodular mildly enhancing mass lesion arising from fundus and body of stomach about 3.8 cm, indenting the left lobe of liver without obvious infiltration. Upper gastrointestinal scopy showed a large submucosal mass of approximately 4-5 cm in the fundus and body of stomach along greater curvature with ulcer at the summit of mass (Figure 1). Biopsy specimens obtained at the endoscopy yielded nonspecific signs of mild inactive chronic inflammation and gastric ulcer without evidence of a dysplasia or malignancy. Patient underwent CT guided biopsy of gastric mass at our centre. Biopsy specimen showed spindle cell neoplasm. Patient was counseled for surgical options considering GIST as a provisional diagnosis and offered an elective laparotomy. After an informed consent, patient had undergone

Correspondence: Apurva S. Shah, Endoscopy Department, 2nd floor, MRC wing, Bombay Hospital and Medical Research Institute, Marine Lines-12, Mumbai-400020, India.

Mobile: +91.9920892248.

E-mail: apurvashah411@gmail.com

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exploratory laparotomy for total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy and 5x5 cm gastric tumor resection and umbilical hernia repair.

Macroscopically, the nodular gastric mass covered by mucosa measured 5x5x4 cm with cut surface shows firm lobulated yellowish white tumor abutting the serosa (Figure 2). Microscopically, submucosal tumor composed of spindle cells arranged in interlacing fascicles separated by myxoid or hyalinised stroma. Neoplastic cells contain ill-defined eosinophilic cytoplasm and slender elongated often wavy, bland appearing nuclei (Figure 3A). The neoplastic cells were immunoreactive with S-100 protein and vimentin (Figure 3B), but lacked immunoreactivity with CD-117, DOG-1, CD-34, smooth-muscle actin and desmin. The histopathologic features and immunohistochemical staining pattern were consistent with gastric schwannoma. Bilateral adnexae had showed serous borderline cystic tumor (low malignant potential) with micropapillary pattern.

The postoperative period was uneventful and one-month follow-up was unremarkable.

Table 1. Immunohistochemistry for differentiating schwannoma and gastrointestinal stromal tumor.

Tumor	S-100	Vimentin	GFAP	CD-117	DOG-1	CD-34
Schwannoma	+	+	+	-	-	-
GIST	-	-	-	+	+	+/-

GFAP, glial fibrillary acidic protein; GIST, gastrointestinal stromal tumor.

Discussion and Conclusions

Gastrointestinal schwannoma is a rare gastrointestinal mesenchymal tumor, first described by Daimaru *et al.* in 1988.⁷ They are thought to arise from the nerve plexus of gut wall, in contrast to conventional schwannomas, which arise from peripheral nerve of skin, connective tissue and internal organs.⁷ Gastric schwannoma can occur at any age but are most frequently noted in fifth and sixth decades with female predominance.⁷ This case was seen in fourth decade and detected incidentally. They cause nonspecific symptoms of pain, epigastric mass and bleeding although many are detected incidentally. *Malignant transformation of a gastric schwannoma is very rare*, only few cases have been reported in literature.⁸ Endoscopy and imaging modalities cannot differentiate gastric mesenchymal tumors as all of them present as solitary submucosal mass. Majority are submucosal in origin, located in body of stomach with size ranging from 0.5 to 11 cm in earlier reported cases. In this case tumor was located in fundus and body of stomach and was of 5×5×4 cm size.

Due to the difficulty of establishing a definite preoperative diagnosis, but also in order to prevent possible complications such as bleeding or pyloric stenosis, surgical resection should be considered the treatment of choice in patients with gastric schwannoma.⁹ The diagnosis of schwannoma is based on tissue histology with further confirmation by immuno-histochemical markers. Immunohistochemistry shows strong nuclear and cytoplasmic staining pattern for S100 protein, vimentin and glial fibrillary acidic protein with consistent negativity for CD-117, DOG-1, CD-34 and smooth-muscle actin (Table 1).

The differentiation between gastric schwannomas and GISTs can be difficult preoperatively, as imaging studies, such as sonography, endoscopy, and CT scan have not shown any distinct features unique to these neoplasms. In 2005, Levy *et al.* reported that gastric schwannomas are uniquely different from other schwannomas in that they show homogeneous attenuation on CT and that degenerative changes such as cystic changes are uncommon.¹⁰ the homogenous enhancement pattern may aid in differentiation of gastric schwannomas from GISTs, which frequently show heterogeneous enhancement due to degenerative changes. Before the recognition of S-100 antigen and c-kit antigen in gastric schwannomas and GISTs, respectively, these neoplasms were most often classified as leiomyoma, leiomyosarcoma, or gastrointestinal autonomic nerve tumor.^{1,2,6,7} Currently, a complete surgical resection of the tumor is the only effective method of treatment and the prognosis after tumor resection is excellent.

In conclusion, gastric schwannoma is uncommon generally benign mesenchymal tumor, which is to be differentiated from GIST

as schwannoma carries a good prognosis as compared to more aggressive GIST, which has malignant potential.

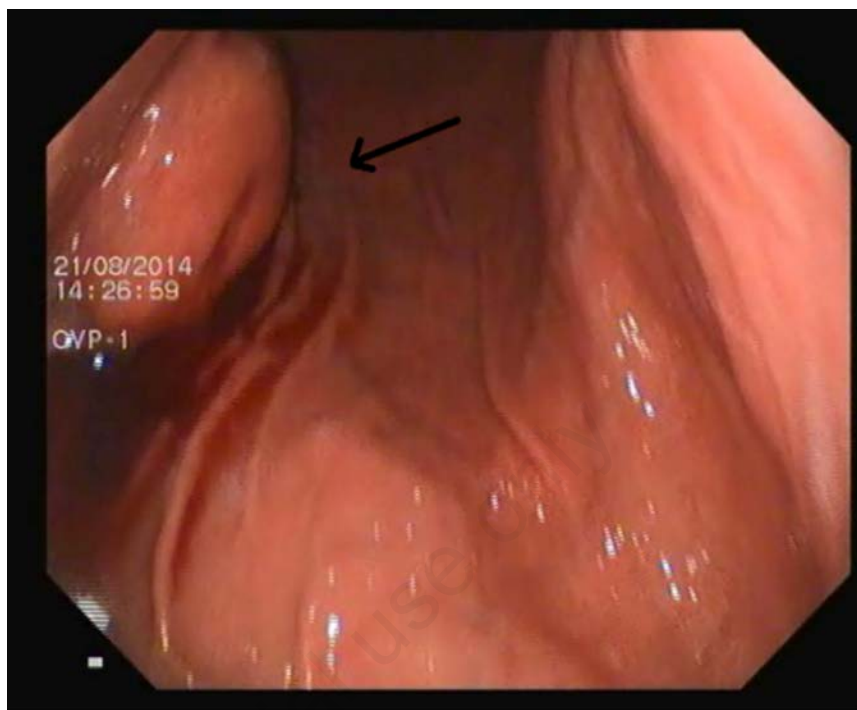


Figure 1. Endoscopy showing submucosal mass in body of stomach.



Figure 2. Macroscopic view of the resected specimen showing a yellow-white, solid and well circumscribed exophytic tumor with a rubbery and jelly cut surface measuring 5×5×4 cm, located at body of the stomach.

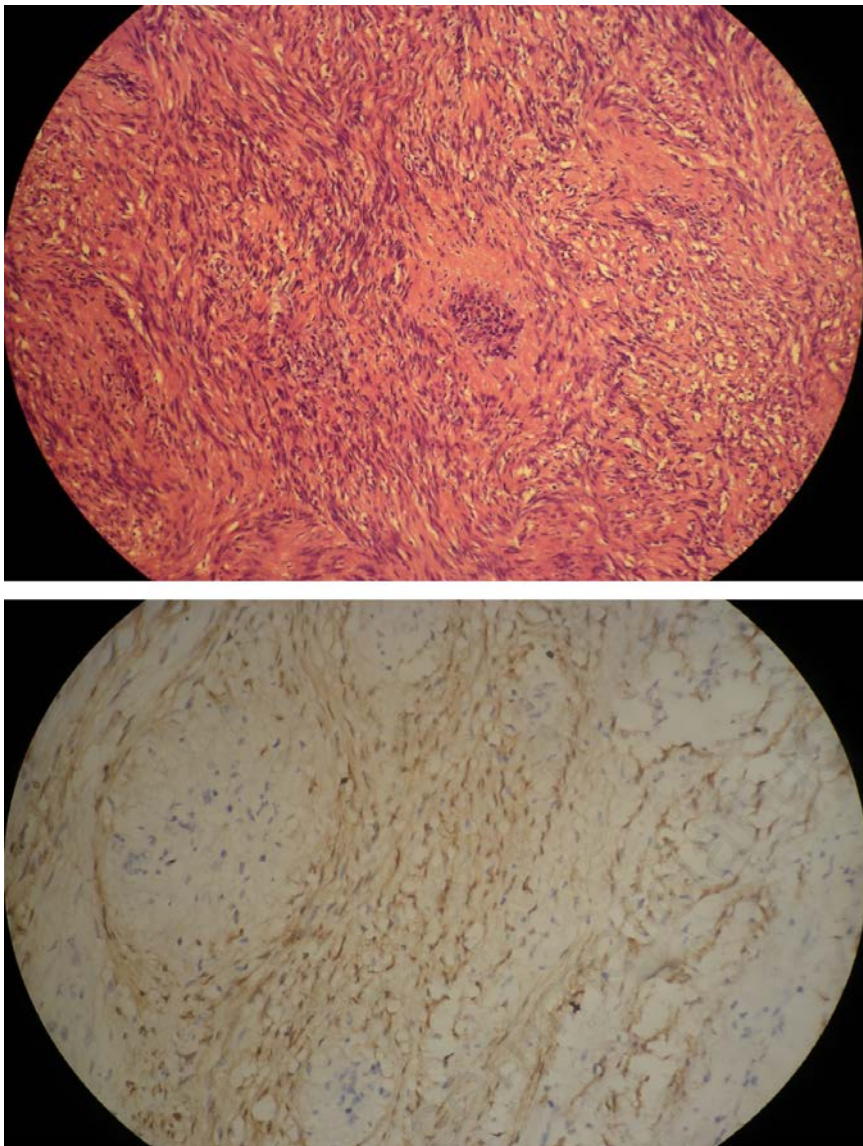


Figure 3. A) Hematoxylin and eosin: spindle cells in fascicles; B) IHC-S-100 stain-400x: tumor cells positive for s-100 marker.

Key message

Gastric schwannoma is often misdiagnosed as GIST as both are submucosal spindle cell tumors.

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