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Gastric schwannoma: a case report and review of literature

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ABSTRACT

Background: Schwannoma is a rare mesenchymal tumor, that arise from the Schwann cells of the nerve sheath. Schwannoma in the gastrointestinal tract is a very uncommon finding, with the stomach being the most common location. Preoperative diagnosis is difficult due to the wide variety of other mesenchymal tumors, definitive diagnosis is after histopathological examination and immunohistochemical staining of the specimen.

Case presentation: We are presenting a rare case of stomach schwannoma, in a 50-year-old male who presented with urological complaints. He had a CT scan that incidentally showed a polypoidal gastric mass.

Discussion: Schwannomas generally present asymptotically; however, in some cases they can cause abdominal discomfort, pain or digestive symptoms. The definitive diagnosis of gastric schwannomas is determined by pathological examination of the surgically removed specimen.

Conclusion: Schwannoma usually runs a benign course with excellent prognosis, yet there are very few reported cases of Malignant GI schwannoma. Benign or malignant, until now the treatment of choice is surgical excision with free margin.

Keywords: gastric schwannoma, benign schwannoma, malignant schwannoma, excellent prognosis.

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Introduction

Mesenchymal tumors of the gastrointestinal tract (GIT) are rare tumors which arises from the spindle cells, including GIST the most common type, leiomyoma, leiomyosarcomas or other neuroectodermal neoplasms¹. In the GIT, gastrointestinal stromal tumours (GISTs) constitute the largest group of mesenchymal tumours, whereas schwannomas are rare². Schwannomas are representing 2-6% of all mesenchymal tumors³. The most common site in the GIT is the stomach with 60-70% and 0.2% of all gastric tumors^{4,5,6}. In the colon and rectum, it represents 3%^{7,8} while it is very rare in the small intestine and esophagus¹. It was first reported by Daimaru et al., in 1988 as a benign tumor with good prognosis⁹. Only 8 cases of malignant schwannomas have been reported in the literature¹⁰ schwannomas usually present asymptotically however in some cases they can cause abdominal pain, GI bleeding or change in bowel habits. Most of the time it is incidentally discovered on clinical imaging like Computerized tomography (CT) while investigating other pathology, or while operating^{2,11}.

During investigation with endoscopy the tumor is usually submucosal with no specific features to differentiate it from other mesenchymal tumors, and biopsy is not usually feasible or inconclusive¹². Computed tomography (CT) and upper gastrointestinal endoscopy are the mainstays of investigation although neither of these investigations are pathognomonic. Ultimately, the definitive diagnoses of GISTs and gastric schwannomas require immunohistochemical studies, which only can be performed on the surgical specimen¹³.

Case Report

A 50-year-old male referred to our clinic with incidentally detected gastric mass on abdominal CT. the reason for his CT was hematuria with back pain for the past year associated with history of kidney stones for which he had a CT scan of his abdomen. He was asymptomatic for the gastric lesion and his abdominal examination

revealed no signs of tenderness and no palpable masses. His past medical and surgical history was not significant and the routine blood investigations were unremarkable.

Due to his urinary symptoms he was investigated with urinary tract CT scan, that showed and incidental well-defined soft tissue density lesion with smooth margins, measuring about 18 x 14 mm is noted along the greater curve of the stomach in the region of the gastric fundus, the lesion was partly projecting into the gastric lumen and partly exophytic with no comment on homogeneity as the CT was without contrast (image 1)

upper GI endoscopy showed large subepithelial lesion in the body of the stomach along the greater curvature and the overlying mucosa is intact. (image 2), Endoscopic ultrasound showed subepithelial lesion in the gastric body measuring 3 cm in size. The lesion was hypoechoic and originating from the third layer of the gastric wall (submucosa) and the elastography showed that the lesion is hard and with fine needle biopsy done (image 3). Biopsy specimen showed spindle cell lesion/neoplasm with slightly atypical nuclei that express S100 and SOX10.

The case was discussed in the multidisciplinary meeting and the decision was to proceed with surgical resection. The patient was consented for laparoscopic wedge resection of gastric lesion. Intra-operatively the exophytic part of the lesion was seen after releasing the greater omentum along the posterior wall of the stomach, but given the pre-operative OGD; the stomach was opened in order to visualize the endophytic part of the lesion to include it completely in the resection (image 5).

The final pathology was schwannoma completely excised and the immunohistochemical staining was positive for S100 and negative for SMA, CD34, CD117, DOG1 and Ki67 was less than 5%. We conducted a literature review using pubmed research engine and two separate researches used the following keywords: schwannoma, GIT and schwannoma, gastric

schwannoma. We reviewed 50 articles and included 9 in our review table with the largest number of schwannomas of the GIT (table 1).

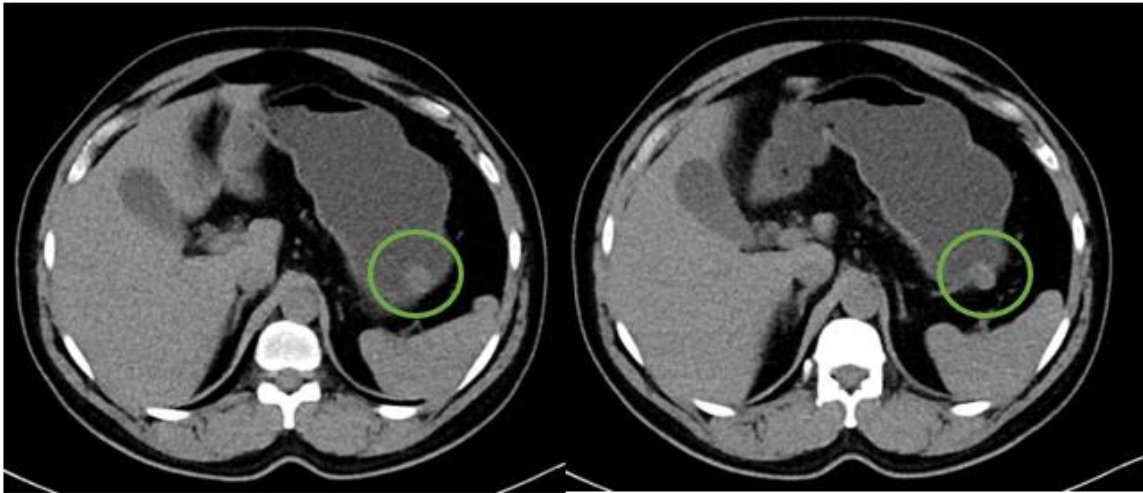


Image 1: CT abdomen and pelvis with contrast showing well-defined soft tissue density lesion with smooth margins(circle), measuring about 18 x 14 mm is noted along the greater curve of the stomach in the region of the gastric fundus, the lesion was partly projecting into the gastric lumen and partly exophytic with no comment on homogeneity as the CT was without contrast.



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Image 2: endoscopy showing large subepithelial lesion in the body of the stomach along the greater curvature and the overlying mucosa is intact.

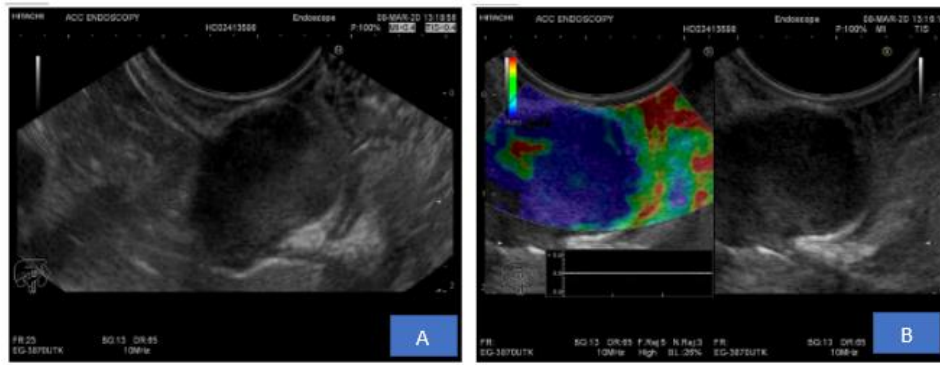


Image 3: A. subepithelial lesion in the gastric body measuring 3 cm in size. The lesion was hypoechoic and originating from the third layer of the gastric wall (submucosa). B. Elastography: the lesion was homogeneously blue (hard elasticity)

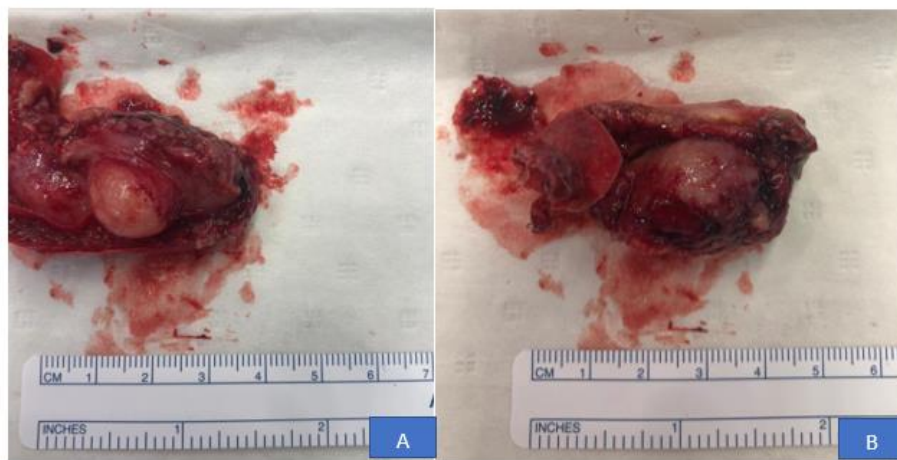


Image 4: intra operative specimen after resection: A: showed the exophytic part of the lesion that was seen at the posterior wall of the stomach neat the greater curvature. B: showed the endophytic part of the lesion that was taken completely with the specimen.

Table 1: literature review articles looking at GIT schwannoma

Author	Year	Type of study	N of cases	Mean age	Location of tumor					Management	Outcome	Follow-up (months)
					Stomach	large bowel	Rec-tum	Small bowel	Esopha-gus			
Levy AD et al	2005	Retrospective	8	70	5			2	1	Complete surgical excision	Full recovery	Not mentioned
Hou YY et al	2006	Retrospective	33	52.6	24	2	3		4	Enucleation to radical surgery	No recurrence or metastasis	6-156
Zheng L et al	2014	Retrospective	29	63.5	29					Complete surgical excision	Local recurrence in one patient and liver metastasis	57
Miettinen M et al	2001	Retrospective	20	63		18	1			Colectomies and one enucleation	No recurrence	81
Kwon MS et al	2002	Retrospective	12	56.5	11				1	11 subtotal gastrectomy and 1 excision	No recurrence and no metastasis	60
Prevot S et al	1999	Retrospective	5	56-74	3	1			1	Surgical resection	No recurrence and no metastasis	Not mentioned
Voltaggio L et al	2012	Retrospective	51		51					Partial gastrectomy and gastric wedge excision	No recurrence and no metastasis	11 years
Tao LP et al	2018	Retrospective	12	52	12					Partial gastrectomy and wide local excision	No recurrence and no metastasis	2.8 years
Li B et al	2014	Retrospective	6	58	6					Endoscopic resection	No recurrence and no metastasis	7.4

Discussion

Schwannoma occurs commonly in the peripheral nerves, connection tissue and central nervous system, the most common form is acoustic neuroma^{9,14}. In the GIT Schwannoma is rare as it arises from the Schwann cells of the gut neural plexus, and considered one of the mesenchymal tumors¹. It represents up to 1% of submucosal tumors of the GIT^{2,15,16} and 2.9% of gastrointestinal mesenchymal tumors³. It most commonly occurs in the stomach but it could happen anywhere in the gut or retroperitoneum in a very less common rate^{4,5,6,17}. Table 1 includes the literature review for all articles that looked at schwannoma of the GIT with the largest study including 51 cases and the smallest study including 5 cases.

The incidence of GI Schwannoma between men and women was the same in previous studies¹⁰. Yet Bohlok et al reported more female predominance of GI Schwannoma over males¹¹. Inagawa et al and Tao et al also showed more cases of intestinal schwannomas among female patients^{12,33}. Schwannoma has been reported in different age groups with incidence among patients in the 6th decade and older^{7,9}.

The clinical presentation of mesenchymal tumors in general including Schwannoma is non-specific and depends on the location of the tumor, symptoms range from abdominal discomfort, pain melena, bleeding, change in bowel habits and tenesmus (and most of the time the tumor is detected incidentally while investigating the symptoms or other pathology which is the case of our patient presented here^{12,19,32}).

Pre-operative diagnosis of Schwannoma or any of the mesenchymal is very challenging. Different modalities of investigations are being used like endoscopy, EUS, CT, MRI, FNA biopsy.

CT scan usually shows an exophytic mass protruding from the wall of the stomach and homogeneous enhancement goes in favor of schwannoma, while cystic and degenerative changes with heterogeneous enhancement is more common with GIST^{1,20}. The CT of our patient was

showing an endophytic and an exophytic part for the lesion but it was difficult to determine the type of tumor per operatively (image 1). Endoscopy usually shows a submucosal tumor with intact mucosa without the ability to distinguish schwannoma from other mesenchymal tumors²¹. The case that we presented had a similar picture in the endoscopy and EUS with the mucosa being intact and the tumor arising from the third layer of gastric wall (image 2 & 3).

EUS with FNA and immunohistochemical staining was studied to diagnose GIST perioperatively and has shown improved accuracy in pre-operative diagnosis, but it has not been studied enough in cases of Schwannoma²². The case presented here had EUS with FNA and immunohistochemical staining was positive for S100. Which could be a promising route in achieving pre-operative diagnosis. Also, nonspecific enolase (NSE) which is an acid protease that is concentrated in Neurons and neural tumor cells, found to be often positive in most GI schwannoma^{23,36}. Zhenbo Shu et al. mentioned in their study that it could be a potential tumor marker, but it needs further studying²⁴. Due to the rarity of the disease it is difficult to unify the diagnostic method or increase the accuracy of pre-operative detection of schwannomas however these two markers (NSE and S100) are promising markers that may help in distinguishing schwannoma pre-operatively.

The definitive diagnosis of Schwannoma is post-surgical excision, histopathological examination and staining. The tumor is usually well circumscribed exophytic lesion without ulceration or calcifications and feels rubbery^{25,33,34}. Image 4 shows the intra-operative specimen after excision and it was firm and rubbery in consistency.

Under the microscope it is composed of interlacing spindle shape cells, with staining of the nucleus and cytoplasm for S100 and are often positive for vimentin and glial fibrillary acidic protein (GFAP), however Schwannoma stain negative for CD117 CD34, actin, desmin, and SMA, which distinguish Schwannoma from GIST and other mesenchymal tumors^{3,7,11,26,27,32,33,35,36}

Schwannoma usually runs a benign course with excellent prognosis, yet there are very few reported cases of Malignant GI schwannoma^{28,29}, which was indicated by the Ki-67 proliferative index, an index that use mitotic activity, tumor size and atypia to indicate malignancy²³.

Benign or malignant, until now the treatment of choice is surgical excision with free margin whether open or laparoscopic depending on the location and size of the tumor, with definitive diagnosis being made after histopathological examination^{30,35,36}. One study concluded that molecular therapy with (imatinib mesylate) might be of a relevant application but yet it has not been implemented^{9,11,31}. New studies looked into endoscopic resection for gastric schwannoma like Li B et al and his group that evaluated five patients diagnosed as gastric schwannoma with average size of the mass of 8 to 25 millimeter. They concluded that endoscopic resection is safe and feasible in small size schwannoma³⁴.

Conclusion:

Due to the rarity of the disease it is difficult to unify the diagnostic method or increase the accuracy of pre-operative detection of schwannomas however these two markers (NES and S100) are promising markers that may help in distinguishing schwannoma pre operatively. Schwannoma usually runs a benign course with excellent prognosis, yet there are very few reported cases of Malignant GI schwannoma. Benign or malignant, until now the treatment of choice is surgical excision with free margin.

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