

# Rare Case of Gastric Schwannoma Simulating Gastrointestinal Stromal Tumour-A Case Report

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## Research Article

**Keywords:** Gastric mass, Gastric Schwannoma, Gastrointestinal Stromal tumour, enhancing gastric mass, S- 100, case report

**Posted Date:** January 10th, 2022

**DOI:** <https://doi.org/10.21203/rs.3.rs-947941/v1>

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# Abstract

Gastric schwannoma (GS) is a rare, benign, slow-growing neoplasm representing 0.2% of all gastric tumors. We report a rare case of Gastric schwannoma in a 56 year old woman who presented with abdominal discomfort and vomiting since few years. Contrast enhanced Computed Tomography showed a well-defined homogeneously attenuating lesion with homogenous enhancement. Wedge gastrectomy was done with a possible diagnosis of Gastrointestinal stromal tumor, but was confirmed to have Gastric schwannoma on histopathology showing spindle cells with peripheral cuff of lymphoid aggregates and S-100 protein positivity on immunohistochemistry.

Gastric schwannoma should be included in the differential diagnosis of a gastric intramural or exophytic mass when Computed Tomography shows a well-defined homogeneously attenuating lesion with progressively increasing homogenous enhancement without hemorrhage, necrosis, and degeneration.

## Introduction:

Schwannomas are benign, slow growing neoplasms arising in any nerve that has a Schwann cell sheath. Gastrointestinal tract (GIT) Schwannomas arise from Schwann cells of the Auerbach's plexus of the GIT wall<sup>11</sup>. Computed Tomography (CT) and upper GI endoscopy are not pathognomonic<sup>9</sup> and correct diagnosis is by histology and Immunohistochemistry(IHC).The key in the immunohistochemical study is S100 protein positivity.<sup>3,4,5</sup> Gastrointestinal stromal tumor (GIST) is its major differential diagnosis clinically, endoscopically, radiologically and histopathologically<sup>9</sup>. GS is not widely recognized by clinicians, making it difficult to distinguish GS from GIST preoperatively<sup>10</sup>.

## Case Presentation:

A 56 year old woman with abdominal discomfort and vomiting since few years ,presented with gradually progressive worsening of symptoms. She had history of hysterectomy and incisional hernia repair. Physical examination revealed mild tenderness in epigastric region. On upper GI endoscopy, submucosal nodular lesion was seen on the lesser curvature of stomach (measuring ~5 x 8cm) with mucosa showing small central ulceration.

Non contrast CT scan showed well-defined, homogeneous, hypodense mass arising from the anterior wall of body of stomach measuring 3.1 x 4.8 x 5.1 cm with a CT attenuation value of 35 HU (Fig. 1a). Contrast-enhanced CT scan of abdomen and pelvis was performed. On the arterial phase, mild homogenous enhancement of both endophytic and exophytic components of the mural mass was seen with a CT value of 45 HU (Fig. 1b). Gradual homogeneous enhancement of the lesion was seen in the portal venous phase (Fig. 1c) with a CT value of 82 HU. Delayed scan showed increase in the CT attenuation value of the mass further to 114 HU. Focal central ulceration was seen along the luminal surface (Fig. 1d). There was no adjacent organ invasion, lymphadenopathy or metastases, indicating a possible GIST. Wedge gastrectomy was performed to relieve the obstruction symptoms.

Macroscopic examination revealed nodular mass which on cut section was greyish white, firm nearly involving full thickness. Microscopic examination (Figure 2a) revealed submucosal neoplasm with spindle cells, intervening stroma containing lymphocytic infiltrates and peripheral cuff of lymphoid aggregates, suggesting benign spindle cell neoplasm (GIST with neural differentiation). Immunohistochemistry (Figure 2b) showed tumor cells were positive for S100 and negative for CD117, Actin and Desmin, favoring GS.

## Discussion:

Gastric submucosal tumors and mesenchymal origin stromal tumors arise in the submucosa or muscularis propria of the gastric wall. They are classified as (i) neurogenic tumors – a) schwannomas b) granular cell tumors c) neurofibromas (ii) GIST and (iii) myogenic tumors- a) leiomyomas b) leiomyosarcomas.<sup>6,11</sup> GI neurogenic tumors are derived from autonomic components of Auerbach's plexus. The most common site for GIT Schwannoma is the stomach (60–70% of cases)<sup>6</sup>, followed by the colon and rectum.<sup>1</sup>

GI schwannomas were initially reported by Daimaru et al<sup>3</sup>. They have excellent prognosis after surgical resection.<sup>1,2,3</sup> Only one malignant case of GS<sup>7</sup> and one related to Von Recklinghausen's disease<sup>13</sup> have been reported. GS are frequently seen in the 5th to 8th decades with female predominance.<sup>1,9,10,12</sup> They can occur in children and, can be malignant rarely.<sup>8,10</sup> They are usually asymptomatic or present with non-specific symptoms.<sup>8,9</sup>

Initial evaluation by upper GIT endoscopy may be normal or show non-specific findings such as extrinsic mass effect or ulceration.<sup>2</sup> Since GS's are submucosal tumours, endoscopic biopsy may not provide definite diagnosis.

CT is the mostly used imaging modality for gastric tumors, providing information for surgical planning by identifying the tumor, location, extent and relationship with neighbouring structures.<sup>8,9</sup> GS appears as well circumscribed, spherical, ovoid or multilobulated solid hypodense mural tumors.<sup>8</sup> Its characteristic CT feature is the homogeneous tumor attenuation due to lack of hemorrhage, cystic change, necrosis or degeneration within the tumors<sup>1,11</sup>. They show mild enhancement in the arterial phase with progressively increasing enhancement in the venous and delayed phases.<sup>1,10</sup> They rarely appear cystic, although large tumors may show cystic degeneration, and calcification is uncommon.<sup>8</sup>

MRI shows low signal intensity on T1 weighted images and high signal intensity on T2-weighted images. Details about its internal features such as hemorrhage, necrosis or cystic changes and its relationship with the surrounding structures can be assessed.<sup>10</sup>

Fluoro-Deoxy Glucose (FDG)- PET is of limited value for the differentiation of GS and GIST, as the FDG uptake in them does not significantly differ. However, it can assess the recurrence or metastasis of malignant tumors.<sup>10</sup>

GS should be differentiated from GIST, which may be malignant or have malignant potential.<sup>1</sup> CT features of GIST depends on its size and aggressiveness. The most important feature for differentiation is the heterogeneous appearance with hypervascularity of GIST with hemorrhage, necrosis, and cystic change resulting in peripheral enhancement (in 92% of case).<sup>1,6</sup> However GS demonstrates homogeneously enhancement.<sup>1,6</sup> Smaller GISTs appear similar to GS on CT making it impossible to distinguish them by imaging alone<sup>12</sup>. However, GS is frequently exophytic or shows mixed growth pattern, homogeneous enhancement, perilesional lymph nodes and grow slower than GIST.<sup>6,11</sup> Final diagnosis is by histopathological and immunohistochemical examination.

Histologically, the pathognomonic feature of GIT schwannomas is a well circumscribed lesion surrounded by a cuff of lymphoid aggregates.<sup>9</sup> They are spindle cell tumors with a microtrabecular pattern, peripheral lymphoid cuffing<sup>2,6,9</sup>, lymphoplasmacytic infiltrate and occasional germinal centers.<sup>1,3,4</sup> Spindle-shaped cells are noted in 70% of all GISTs and 20% contain epithelioid type or they can be pleomorphic. Immunohistochemistry is confirmatory for distinguishing GS and GIST.<sup>6</sup>

Immunohistochemistry shows positive staining for S100 and vimentin and negative staining for smooth muscle actin and CD34 in neurogenic tumour.<sup>9</sup> GISTs are mostly CD117-positive, CD34-positive, actin-positive, and S100-negative.<sup>6</sup> In our case, the tumor cells were positive for S100 and negative for CD117, Actin and Desmin consistent with gastric neurogenic tumor. Lymphoid aggregation and presence of capsule highly suggested schwannoma rather than neurofibroma. Calretinin is a good marker to differentiate schwannomas and neurofibromas.<sup>6</sup>

Other differential diagnosis are primary /secondary lymphomas and adenocarcinomas. Lymphomas show homogeneous CT attenuation before treatment, similar to GS. However, they are commonly associated with lymphadenopathy.<sup>1</sup> Spiculated margins associated with regional adenopathy is seen in adenocarcinomas.<sup>1,11</sup>

The goal of surgery is to achieve R0 resection, including the lesion and any involved adjacent structures. Follow-up is not offered unless there are signs of malignant transformation.<sup>9</sup> The treatment for lesser curvature, middle and distal third gastric neoplasms is subtotal gastrectomy.<sup>6</sup>

## Conclusions

Gastric schwannomas should be considered when a well-defined intramural or exophytic gastric mass with homogeneously attenuation lacking hemorrhage, necrosis, and degeneration, is encountered, helping in its differentiation from GIST. Knowledge of these characteristic findings helps to prevent unnecessary surgery and improve planning for minimally invasive surgery.<sup>11</sup>

## Declarations

**Funding:** Nil.

**Conflicting of interests /Competing interests:** The authors declare that they have no competing interests

**Ethics approval:** Ethics has approved this study.

**Consent to participate:** The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal

**Written Consent for Publication:** The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Availability of data and materials:** Provided in this case report.

**Code availability:** Not applicable

**Authors' contributions:** All authors had substantial contributions to the conception of the case report. All authors were active participants in the drafting and revising of the case report. All authors approved the final version of manuscript. All authors agree to be accountable for all aspects of the work. All authors were directly involved in this patient's care and contributed to the writing and editing of this manuscript. SK, RM researched and wrote the patient background and pulmonary history. HV, SS researched and wrote the abstract and conclusion of the manuscript, captured and arranged the images. SP conceptualized the process; researched, compiled, and wrote the procedure, conclusion, and introduction; and served as the overall editor.

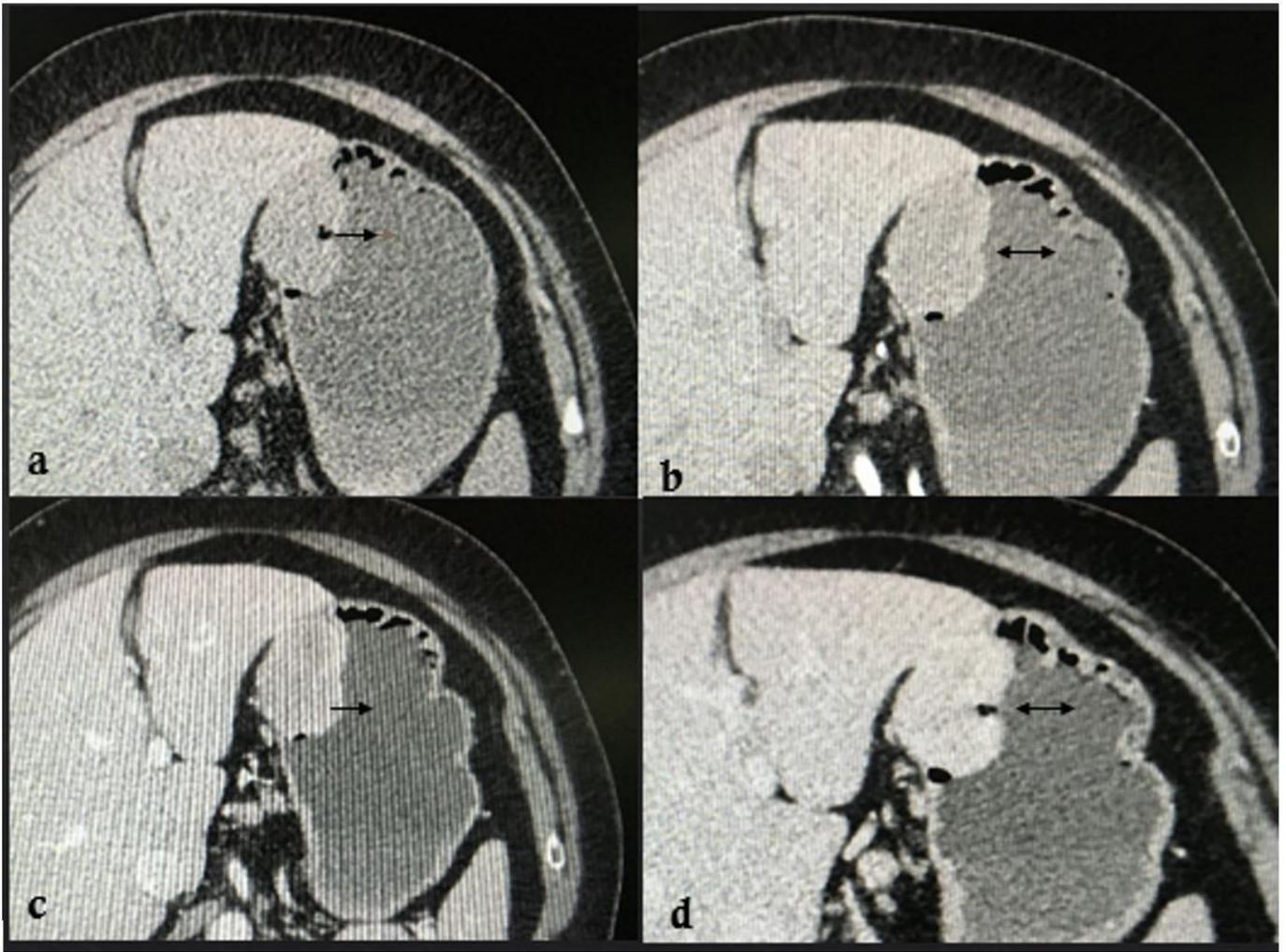
**Acknowledgements:** Nil

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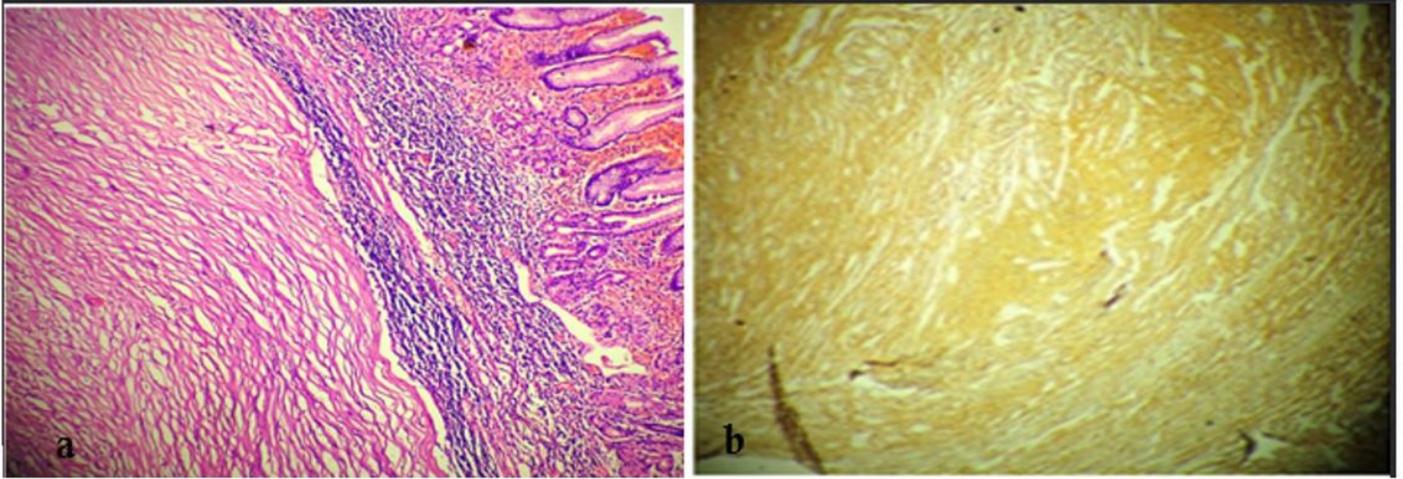
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## Figures



**Figure 1**

(a) Non contrast CT scan shows well-defined, homogeneous, low-attenuation mass arising from the anterior wall of gastric body( → ). The CT value of the mass was about 35 HU before the injection of contrast medium. IV contrast-enhanced CT scan (b, c, d) showed (b) homogeneously enhancing mural mass with mild homogeneous enhancement with a CT value of 45 HU on arterial-phase ( ↔ ). (c) Lesion shows gradual homogeneous enhancement in the portal venous phase ( → ) with CT value of 82 HU. (d)The CT value of the mass further increased to 114 HU on delayed phase after a delay of 3 minutes. Surface ulceration noted at the endoluminal surface of the lesion ( ↔ ). After wedge resection, the lesion was confirmed as a gastric schwannoma.



**Figure 2**

(a) Microscopic examination shows spindle cells with intervening stroma containing lymphocytic infiltrates. Peripheral cuff of lymphoid aggregates seen. (b) Immunohistochemistry shows tumor cells positive for S100.

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