

# Gastric Schwannoma: Incidentally Found Rare Gastric Tumor Presented with Nonspecific GI Symptom

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## Abstract

Gastric schwannomas (GSs) are generally slow-growing, asymptomatic encapsulated tumors of mesenchymal origin arising from Schwann cells of nerve plexus within the gastric wall. Schwannomas are rare in the stomach, and account only 0.2% of all gastric tumors. Among mesenchymal tumors in the gastrointestinal (GI) tract, gastrointestinal stromal tumors (GISTs) are the most common, and 60–70% of them occur in the stomach. Under endoscopic examination, GS/GSs and GIST/GISTs appear grossly similar as firm submucosal neoplasm. The most common presenting symptom is upper gastrointestinal bleeding, followed by abdominal pain. Here, we report a 77-year-old lady presented with progressive weakness, and received prompt esophagogastroduodenoscopy (EGD) due to anemia. The EGD revealed a submucosal tumor with hemorrhagic ulceration at the middle to lower body of the stomach. After contrast-enhanced computed tomography (CT), the patient received partial gastrectomy with initial impression of GIST/GISTs, but confirmed as schwannoma postoperatively. This case highlights the importance of considering schwannomas as a differential diagnosis when preoperative imaging studies revealed a submucosal, exophytic gastric mass. The case also underscores the value of prompt EGD survey even for normocytic anemia with negative stool occult blood and nonspecific GI symptoms, especially when it comes to the elderly. (J Intern Med Taiwan 2014; 25: 350-356)

**Key Words:** Schwannoma, Stomach, Submucosal tumor, Neurogenic gastrointestinal tumor, Immunochemical stain

## Introduction

Gastric schwannomas (GSs) are generally slow-growing, asymptomatic encapsulated tumors of mesenchymal origin arising from Schwann cells

of nerve plexus within the gastric wall. Schwannomas are responsible for myelin sheath formation and trophic support for peripheral nerve system. They are rare in the stomach, and account only 0.2% of all gastric tumors. Among mesenchymal tumors

in the GI tract, gastrointestinal stromal tumors (GISTs) are the most common, and 60–70% of them occur in the stomach. Under modern imaging studies as esophagogastroduodenoscopy (EGD) or computed tomography (CT), GS/GSSs and GIST/GISTs appear grossly similar as round submucosal neoplasm. Here, we report a 77-year-old lady who suffered from progressive weakness, related to normocytic anemia. She undertook a prompt EGD, which revealed a round submucosal tumor with a central ulceration at the middle to lower body of the stomach. Pigmented hematin on the ulcer base implied recent bleeding event. The patient received partial gastrectomy under the impression of GIST, but the pathology confirmed as gastric schwannoma.

## Case report

A 77-year-old female presented to neurology outpatient clinic with progressive weakness and dizziness for months. Her medical history was significant for hypertension, old cerebrovascular accident, peptic ulcer disease, and cholelithiasis related cholecystitis, which was treated with cholecystectomy 20 years prior. Physical examination revealed stable vital sign and general weakness. Serial neurological examinations yielded no definite diagnosis, and laboratory test revealed only normocytic anemia (hemoglobin = 11.1 g/dL, normal range: 12.3~15.3g/dL; hematocrit = 34.0%, normal range: 35.9~44.6%; mean corpuscular volume = 93 femtoliters, normal range: 80.0~96.1 femtoliters). Iron profile showed no evident iron deficiency (Ferritin = 70.7 ng/mL, normal range: 11~307 ng/mL; Serum iron = 31 ug/dL, normal range: 28~170 ug/dL; TIBC = 299.6 ug/dL, normal range: 250~450 ug/dL). For anemia and history of peptic ulcer disease, she was referred to GI outpatient department. The patient denied abdominal pain, nausea/vomiting, diarrhea, or constipation. No definite episode of melena, hematochezia or hematemesis was found. No significant body weight loss was observed by the family.

Her appetite was not decreased, but she noticed postprandial abdominal fullness recently. Though repeated stool occult blood examination reported negative result, EGD was arranged for anemia workup considering history of peptic ulcer disease. The endoscopy reported a huge, round submucosal tumor with central ulceration noted on the lesser curvature side, middle to lower body (Fig. 1a). The overlying mucosa was intact except for the central ulcer, and the quality was consistent with submucosal tumor and revealed negative pillow sign when probed. Flat pigmented hematin on the ulcer base was noted, indicating recent bleeding event (Fig. 1b). Biopsy was performed during EGD examination, and pathology examination reported chronic active gastritis and ulcer. Subsequent abdominal contrast-enhanced CT scan showed a submucosal and exophytic growing mass lesion, about 7.7 x 5.2 x 5.5 cm at the anterior aspect of gastric body, with a central ulcer-like defect (Fig. 2). Overlying mucosa was preserved except in the defect. There was no evidence of any other abnormalities. The initial impression of EGD and CT study was GIST with differential diagnosis of lymphoma. Further laboratory tests revealed persisting normocytic anemia, mild hypoalbuminemia and tumor markers of CA 125, CA 19-9, AFP and CEA were all within normal limit.

After consulting with general surgery, surgical intervention was recommended. The patient received partial gastrectomy with pyloroplasty. During operation, a round, exophytic submucosal tumor located at the middle to lower body of the stomach was seen (Fig. 3a). On gross inspection, the tumor was a solid mass with a smooth surface and a deep central ulceration, measuring 7.2 x 6.5 x 5.2 cm in size (Fig. 3b).

The pathological examination revealed a picture of bland spindle cells proliferation with nuclear palisades and whorls, consistent with schwannoma (Fig. 4). The picture of H&E stain

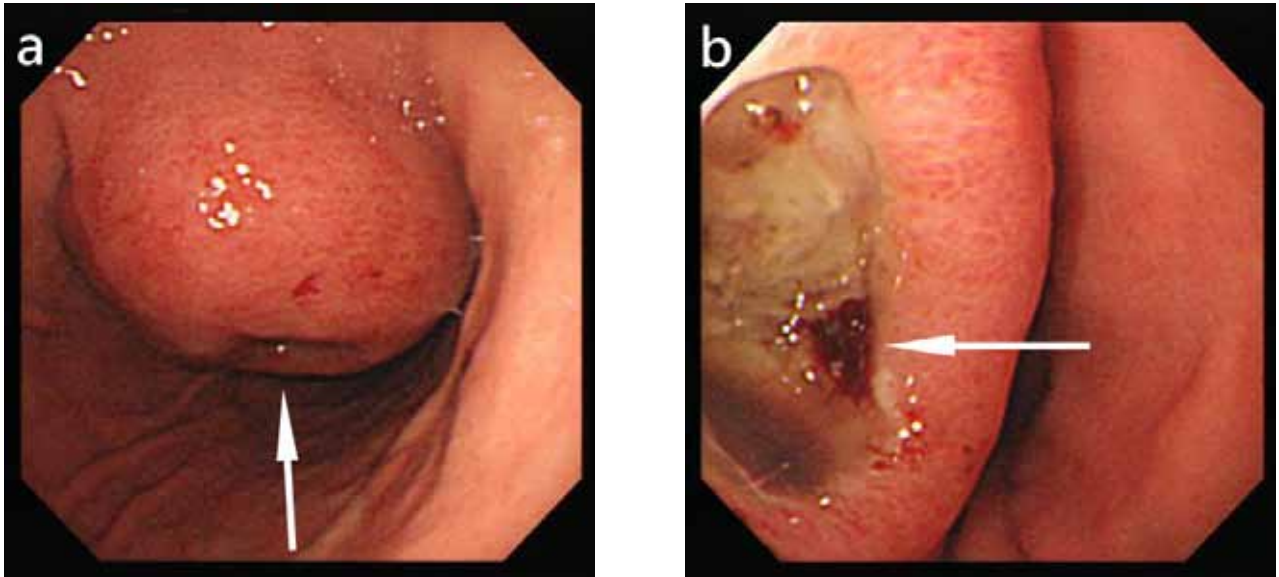


Figure 1. (a) The esophagogastroduodenoscopy reported a huge submucosal tumor with central ulceration (arrow) on the lesser curvature side, middle to lower body. (b) Note the flat pigmented hematin on the ulcer base (arrow) indicating recent bleeding event. The quality was firm with negative pillow sign when probed.

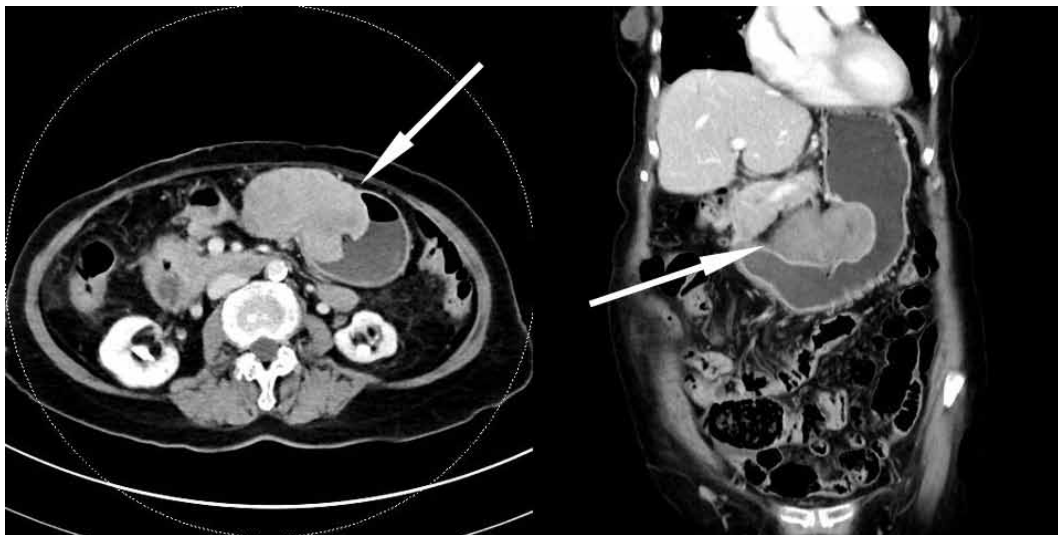


Figure 2. Subsequent abdominal contrast-enhanced CT scan showed a submucosal, exophytic growing mass lesion (arrows), about 7.7 x 5.2 x 5.5 cm at anterior aspect of gastric body with a central ulcer.

revealed typical lymphocytic cuffing at periphery to the tumor (Fig. 5). Immunohistochemical studies revealed strongly positive for S-100 protein (Fig. 6), and non-reactive for CD117, CD34, DOG-1, desmin and actin.

The postoperative course was smooth and the patient was discharged under stable condition. Six months later, EGD follow up showed scarring

convergence of gastric wall with normal mucosal pattern, laboratory test of blood revealed normal complete blood count. The patient was free of symptoms as weakness or dizziness.

## Discussion

Schwannoma are encapsulated nerve-sheath tumors arising from Schwann cells, which form the

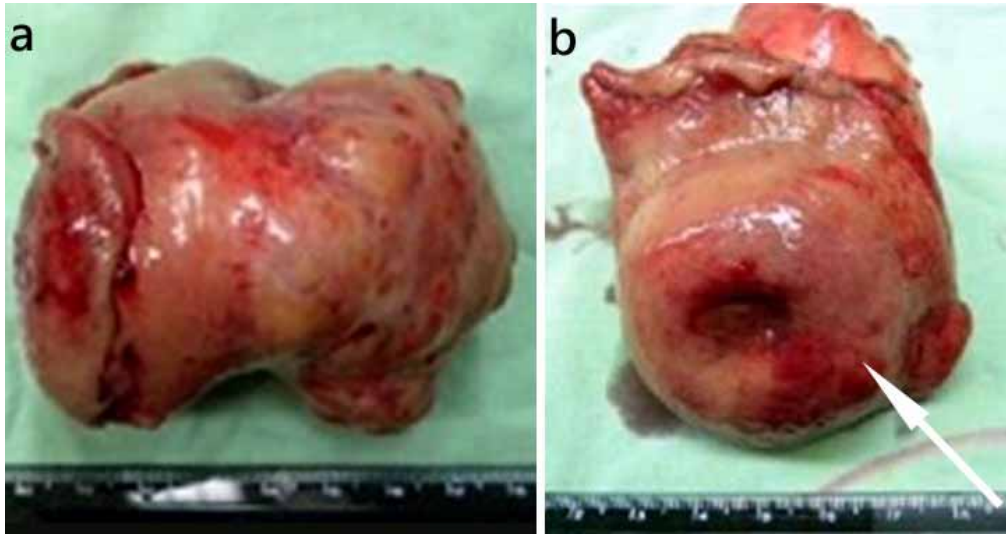


Figure 3. (a) A round tumor measuring 7.2 x 6.5 x 5.2 cm in size, located at the middle to lower body of the stomach was seen (A). (b) A deep central ulceration (arrow) on the tumor was observed (B).

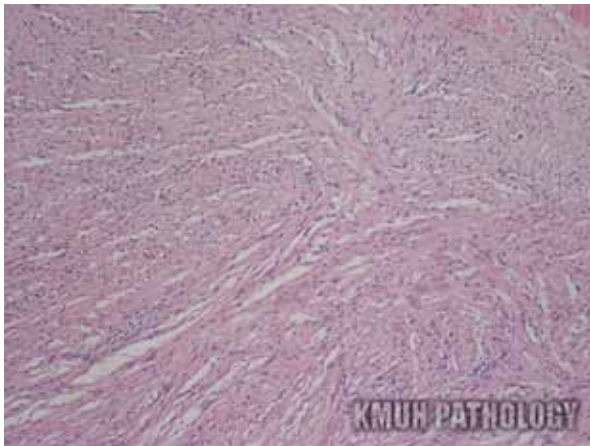


Figure 4. A picture of bland spindle cells proliferation with nuclear palisades and whorls, consistent with schwannoma (hematoxylin and eosin stain,  $\times 100$ ).

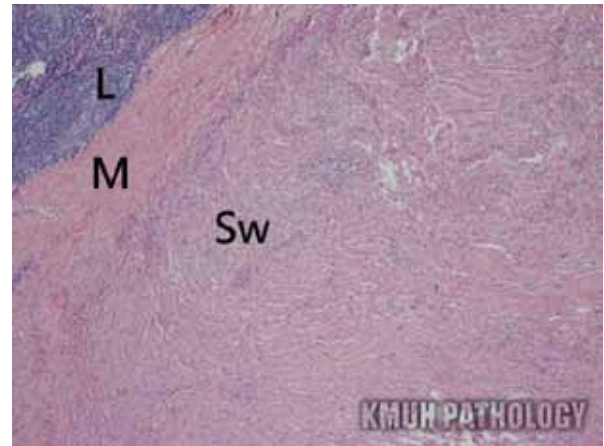


Figure 5. A cuff-like lymphoid aggregate was recognized peripheral to the tumor (L: lymphoid cuff; M: muscular layer; Sw: tumor) (hematoxylin and eosin stain,  $\times 40$ ).

myelin sheath and play a pivotal role in the maintenance and regeneration of axons of the neurons in the peripheral nervous system. It is usually solitary and most commonly involves in the acoustic nerve. In rare cases, it occurs in the GI tract, most often in the stomach, particularly gastric body<sup>1</sup>. In the gastric wall, it originate from the nerve sheath of Auerbach plexus or, less commonly, Meissner plexus<sup>2</sup>. It was not considered a primary GI tumor till 1988, Daimaru et al. successfully distinguished it from

GIST based on the positive S-100 stain<sup>2</sup>. Before the recognition of S-100 antigen and c-kit (also known as CD117) antigen in gastric schwannomas (GSs) and in GIST/GISTs, respectively, these neoplasms were categorized as gastrointestinal mesenchymal tumors (GIMTs) including myogenic tumors, neurogenic tumors and gastrointestinal stromal tumors. In this case, the tumor revealed spindle cells, strongly positive for S-100 stain, and non-reactive for CD117, CD34, DOG-1, desmin and actin, which indicated



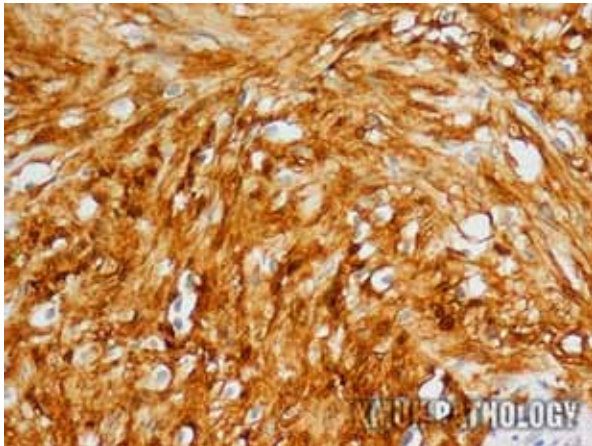


Figure 6. Immunohistochemical studies revealed strongly positive for S-100 protein.

the diagnosis of schwannoma.

Gastric schwannomas occur predominantly in the fifth to sixth decade of life and commonly in female patients. They are usually asymptomatic or associated with non-specific abdominal discomfort. Because of their indolent growth pattern, as with our case, these tumors are often discovered incidentally via endoscopy, radiography or computed tomography. The most common presenting symptom is upper gastrointestinal bleeding, followed by abdominal pain.

The patient was found of mild normocytic anemia (hematocrit = 34.0%, normal range: 35.9~44.6%) with negative stool occult blood. No episode of overt GI bleeding or significant body weight loss was told. Iron profile showed no evident iron deficiency. If iron deficiency anemia is established, gastrointestinal endoscopy evaluation is always indicated to exclude gastrointestinal malignancy<sup>3</sup>, or other associate disease as atrophic gastritis or celiac disease. However, for a patient presented with normocytic anemia, not categorized as iron deficiency anemia or occult GI bleeding, a prompt EGD survey, as with this case, disclosed significant finding. The fecal occult blood tests, guaiac based, were ordered twice by neurologist and GI specialist, and both tests revealed negative results. As the

endoscopic survey confirmed recent UGI bleeding, the negative fecal occult blood test results were considered due to no bleeding events during tests or inadequate diet preparation. The typical endoscopic description of GS/GSs is a round protruding submucosal mass with overlying ulcerated mucosa. Endoscopic biopsies always yield false-negative results in terms of submucosal lesion<sup>4</sup>, and deep biopsy had been advised<sup>5</sup>.

Under endoscopy ultrasound (EUS) evaluation they were tend to be hypoechoic, with a connection between the tumor and the muscularis propria. Compared with GIST/GISTs, GS/GSs were found to be more exophytic in growth pattern, may be characterized by marginal halo resulted from lymphoid cuff, and less feature in lobulation, cyst or calcification<sup>4</sup>. Computed tomography also helps evaluate submucosal tumors, especially for preoperative evaluation. Sharing some of the features of EUS, GS/GSs under CT more frequently show an exophytic growth pattern, homogeneous enhancement pattern, perilesional lymphnodes and grow slower than GIST/GISTs<sup>6</sup>. Fluorodeoxyglucose (FDG) positron emission tomography (PET) has also been used to evaluate GS/GSs, and increased fluorodeoxyglucose uptake has been reported<sup>7</sup>. To distinguish GS/GSs from malignant submucosal tumors via FDG PET is therefore considered less practical.

Despite the features of GS/GSs aforementioned diagnostic modalities provide, preoperative diagnosis of GS/GSs remains difficult. Endoscopic deep biopsy or prompt surgical intervention should therefore be considered to reach a conclusive diagnosis<sup>5,8,9</sup>. Complete resection of the tumors has been considered curative, and the prognosis is excellent<sup>10</sup>.

In summary, this case highlights the importance of considering schwannomas as a differential diagnosis when preoperative imaging studies revealed a submucosal, exophytic gastric mass. For an elderly with nonspecific symptom, this case also underscores the value of prompt EGD survey for

normocytic anemia with normal iron profile and negative stool occult blood. Prompt surgical resection not only provides the diagnosis of GS/GSs, but also treats gastrointestinal bleeding or other abdominal symptoms. More importantly, complete excision of the tumor, as in this case, leads to promising prognosis.

## Disclosures

The authors declare that they have no conflict of interest.

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The authors had full access to all of the clinical records in the case report and take responsibility for the integrity of the clinical records.

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# 胃部許旺氏細胞瘤： 以非特異腸胃道徵狀表現的罕見黏膜下腫瘤

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## 摘 要

許旺氏細胞瘤(schwannoma)通常是生長緩慢，症狀不明顯的間質細胞腫瘤，腫瘤由鞘膜包覆，源自於周邊神經叢的許旺氏細胞(Schwann cell)。此類腫瘤好發於聽神經，即所謂聽神經瘤(acoustic neuroma)，在胃部發生較為罕見。胃部的許旺氏細胞瘤生長自胃壁的神經叢，佔所有胃部腫瘤的百分之零點二。然而，在胃腸道的間質細胞瘤當中，腸胃道基質瘤(gastrointestinal stromal tumors, GISTs)最為常見，其中胃部的腸胃道基質瘤佔了六至七成。在內視鏡檢查下，胃部的許旺氏細胞瘤(gastric schwannoma)與腸胃道基質瘤皆以黏膜下腫瘤表現，兩者極為相似。胃部的許旺氏細胞瘤常見的症狀表現以出血為主，腹部的疼痛與不適感次之。在此案例報告中，我們提出一位七十七歲婦女，因為逐漸虛弱無力於神經科求診，實驗室檢查發現貧血後，於胃腸內科安排胃鏡而發現胃體部黏膜下腫瘤，腫瘤中央出現潰瘍與出血後徵象。經腹部電腦斷層檢查後，初步臆診為腸胃道基質瘤並進行部分胃切除術。然而，術後病理報告顯示該腫瘤為胃部許旺氏細胞瘤。此案例提醒臨床醫師在內視鏡、影像學檢查發現黏膜下、向外生長的胃部腫瘤時，應考慮許旺氏細胞瘤之可能。同時，此案例也強調單純的貧血、未合併明確腹部症狀，儘管無明確胃腸道出血證據時，盡早進行內視鏡檢查的價值。最後，六個月後本案例的內視鏡與實驗室檢查，可發現病人胃部無腫瘤復發跡象，血色素回復正常範圍。