Gastric Glomus Tumor: A Case Report and Review of the Literature

Chuang-Chi Huang,[1] Fang-Jung Yu,[2] Chang-Ming Jan,[2,3] Sheau-Fang Yang,[4,5] Yu-Ting Kuo,[6] Jan-Sing Hsieh,[1,7] and Jaw-Yuan Wang[1,7]

Departments of [1]Surgery, [2]Internal Medicine, [5]Pathology and [6]Radiology, Kaohsiung Medical University Hospital, and Departments of [3]Internal Medicine, [4]Pathology, and [7]Surgery, Faculty of Medicine, College of Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan.

Glomus tumors are usually thought of as benign tumors although some malignant cases have been reported. These tumors arise from the glomus body and are commonly observed in the dermis or subcutis, but rarely in visceral organs. Here, we report a 37-year-old female who initially presented with epigastric discomfort. The preoperative diagnosis was a gastrointestinal stromal tumor. A minilaparotomy was done with an incision length of 4 cm followed by wedge resection. The final pathologic diagnosis was a gastric glomus tumor. We have reviewed the only five cases of gastric glomus tumors that have been reported to date in Taiwan, including the present case, and compare these cases with those reported in other countries. The age of onset ranged from 35 to 69 years (median, 41 years) with female dominance (4 females and 1 male). Two of the five cases presented with gastrointestinal bleeding with an ulcerative tumor, and the others only had epigastric discomfort. The tumors were located around the prepyloric antrum of the stomach. No definite diagnosis was reached before surgery in any of the five cases, and all of the tumors were considered likely to be benign lesions. Clinicians who treat such patients should be aware of this problem because of the difficulty in accurate preoperative diagnosis.

Key Words: endoscopic ultrasonography, glomus tumor, minilaparotomy, stomach
(*Kaohsiung J Med Sci* 2010;26:321–6)

Glomus tumors are rare benign tumors arising from the glomus body, which plays a role in regulating arterial blood flow, and is sensitive to temperature variation. Glomus bodies are commonly observed wherever arteriovenous anastomoses are present. The first clinical and pathological description of a glomus tumor was described by Barre and Masson in 1924 [1].

Received: Sep 23, 2009 Accepted: Oct 28, 2009
Address correspondence and reprint requests to: Professor Jaw-Yuan Wang, Department of Surgery, Kaohsiung Medical University Hospital, and Department of Surgery, Faculty of Medicine, College of Medicine, Kaohsiung Medical University, 100 Tzyou 1st Road, Kaohsiung 807, Taiwan. E-mail: cy614112@ms14.hinet.net

Glomus tumors are commonly noted in the extremities and are rarely found in visceral organs, although tumors in the mediastinum, trachea, kidney, uterus, vagina and stomach have been described. Since Kay et al first reported three glomus tumors of the stomach in 1951 [2], fewer than 200 cases have been published in the literature in any language, and less than 100 cases have been published in English-language journals. The incidence of gastric glomus tumors is much less common than that of gastrointestinal stromal tumors (GISTs), with only 1 in 100 GISTs being gastric glomus tumors [3]. Because of the limited number of cases experienced at a single institute, we have presented our case and reviewed all five cases reported in Taiwan to date, and compare these cases with those of other countries.

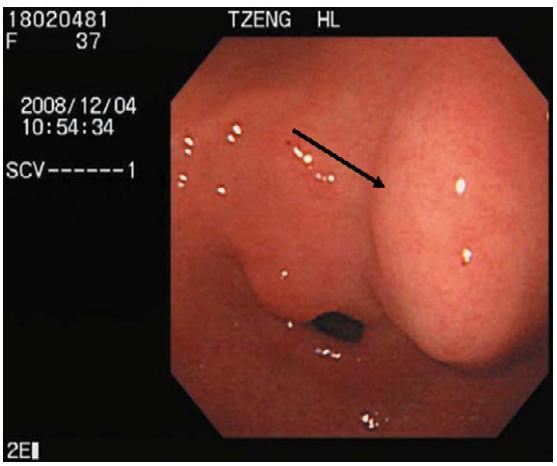

Figure 1. *A submucosal tumor was found near the prepyloric antrum by panendoscopy (arrow).*

Case Presentation

A 37-year-old female visited our institution for intermittent epigastric discomfort over a 2-month period. No nausea or vomiting was noted and her physical examination was normal. Panendoscopy revealed an antral submucosal tumor with an intact surface (Figure 1). Abdominal computerized tomography (CT) revealed a hypervascular submucosal tumor at the posterior, medial and inferior wall of the antrum, near the pylorus ring (Figure 2). Endoscopic ultrasonography (EUS) showed that the tumor was within the muscularis propria layer and had a heterogeneous appearance (Figure 3). Initial laboratory tests revealed the following: serum aspartate aminotransferase level of 55 IU/L, serum alanine aminotransferase level of 67 IU/L, and serum hemoglobin level of 11.9 g/dL. Because we initially suspected antral GIST, a minilaparotomy was done with an incision length of 4 cm. At operation, a submucosal tumor of about 2.0 × 1.5 cm in size was found at the posterior wall of the antrum, and a wedge resection of the tumor was done. She was discharged 5 days later with an uneventful postoperative course. The pathology revealed that the tumor cells were surrounded by hyperplastic smooth muscle fibers of the muscularis propria. The tumor had sheets of round or epithelioid tumor cells with small, uniform nuclei and sharp cell borders surrounding dilated thin-walled blood vessels (Figure 4). Immunohistochemistry revealed the tumor was positive for smooth muscle actin, but negative for cytokeratin and CD56 (Figure 5). Thus, the final diagnosis was gastric glomus tumor.

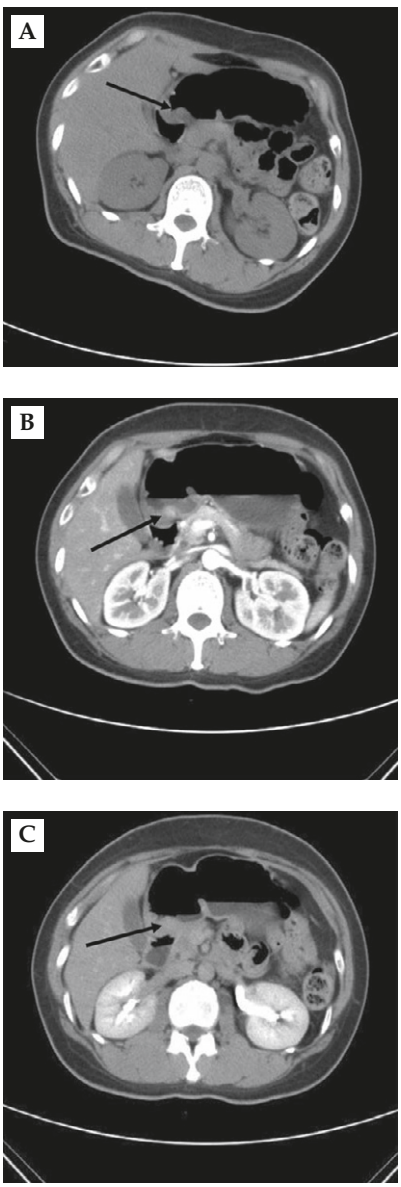

Figure 2. *Abdominal computed tomography revealed a submucosal tumor at the posterior, medial and inferior wall of the antrum near the pylorus ring (arrow). (A) Unenhanced; (B) arterial phase; and (C) delayed phase.*

Discussion

We searched Medline for all cases of gastric glomus tumors reported up to May 2009, and found no more than 200 cases in all languages and less than 100 cases in English-language journals. To date, only five cases have been reported in Taiwan, including the present case. The clinical presentations, diagnostic tools, surgical techniques, preoperative tentative diagnosis, and the characteristics of the specimens are listed in the Table [4–7]. The clinical presentations showed that

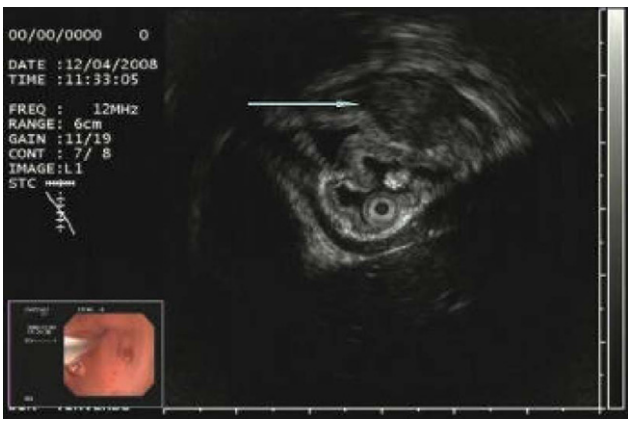

Figure 3. *The submucosal tumor was noted in the muscularis propria layer and had a heterogeneous appearance under endoscopic ultrasound (arrow).*

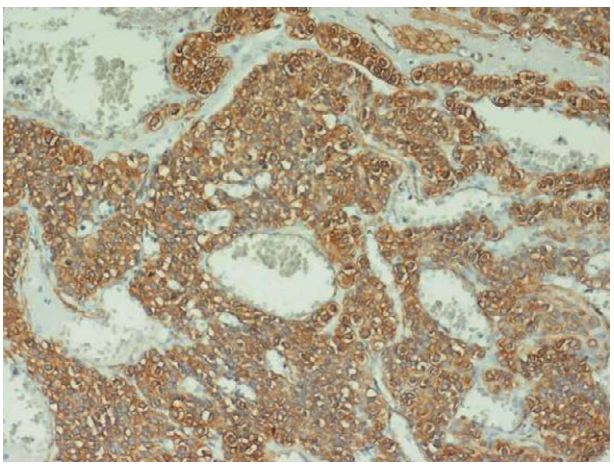

Figure 5. *Immunohistochemistry confirmed the tumor was positive for smooth muscle actin (original magnification, 100×).*

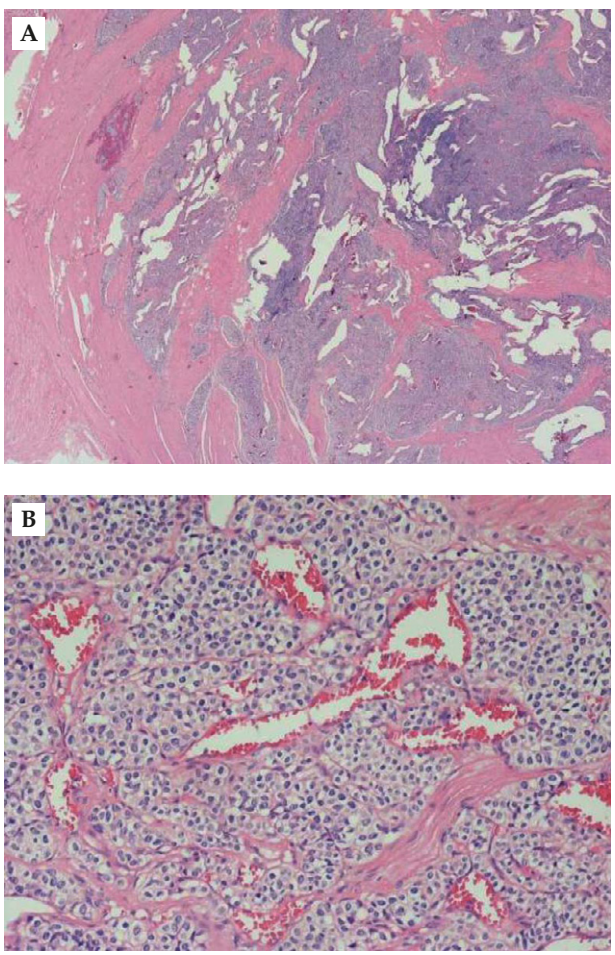

Figure 4. *(A and B) The tumor was surrounded by hyperplastic smooth muscle fibers of the muscularis propria. The tumor has sheets of round or epithelioid tumor cells with small, uniform nuclei and sharp cell borders around the dilated thin-walled blood vessels. (Hematoxylin and eosin; original magnification, (A) 20× and (B) 100×).*

these gastric glomus tumors in Taiwan have a female predominance (4/5 cases, 80%), which is similar to the incidence in other countries: 76.9% in Korea [3] and 60% in Canada [8]. The age range (35–69 years) of the cases in Taiwan was also similar to that in Korea and Canada, although a wider age range was reported in Canada [8]. Gastrointestinal (GI) bleeding with an ulcerative tumor was observed in two cases, while three patients with a non-ulcerative tumor presented with epigastric discomfort. Consequently, GI bleeding with hematemesis/melena and epigastric discomfort are the most two common initial symptoms/signs, and GI bleeding can be life-threatening or lead to chronic anemia. No incidental case have been reported in Taiwan, which is similar to that in Korea [3], although five of 31 cases were incidental in Canada [8].

In terms of preoperative evaluations, EUS was performed in three of the five cases, and fine needle aspiration (FNA) was done in one case. However, a definitive diagnosed was not reached before surgery in any case, even in the case that underwent FNA. Until now, only three articles have reported a preoperative definite diagnosis with FNA [9–11]. The findings of EUS were usually heterogeneous tumors between the submucosal and muscularis propria layer and these findings may be confused with malignant GIST or leiomyosarcoma, which is also represented by a heterogeneous tumor on EUS [12,13]. Our case also had a heterogeneous submucosal tumor within the fourth layer. One case underwent subtotal gastrectomy with Billroth I anastomosis because the EUS findings suggested malignant GIST. Abdominal CT

Table. Demographic and clinical description of the five gastric glomus tumors in Taiwan

	Case 1 [4]	Case 2 [5]	Case 3 [6]	Case 4 [7]	Present case
Year of publication	1993	1999	2005	2007	2009
Sex	F	F	M	F	F
Age (yr)	41	35	60	69	37
Location	Antrum	Lower body	Antrum (greater curvature)	Antrum (less curvature)	Antrum (less curvature)
Ulceration	Yes	Yes	No	No	No
Symptom/sign	Melana	Hematemesis, melana	Epigastric discomfort	Intermittent epigastric discomfort	Epigastric discomfort
Size (cm)	3.0×2.5×2.5	2.5×3.0	2.0	2.0	2.0×1.5
EUS	ND	Heterogeneous with hyperechoic and anechoic	ND	Heterogeneous, hypoechoic mass with internal hyperechoic spots	Heterogeneous
FNA	ND	ND	ND	Nondiagnostic	ND
Operative procedure	Distal hemigastrectomy and Billroth I anastomosis	Subtotal gastrectomy and Billroth I anastomosis	Wedge resection	Wedge resection	Wedge resection
Tentative diagnosis	Carcinoid tumor by endoscopic biopsy	Leiomyosarcoma	GIST, carcinoid, glomus tumor	Not diagnostic	GIST
Immunoreactive stains	SMA, vimentin	ND	SMA	SMA	SMA
CT findings	ND	ND	Well-defined hypervascular tumor	Homogenous enhancement	Significant enhancement during arterial phase
Other examination	No	No	MRI: T1-slightly hypointense T2-slightly hyperintense	Power Doppler sonogram: prominent vascular signals within the mass	No

EUS = Endoscopic ultrasonography; FNA = fine needle aspiration; CT = computerized tomography; MRI = magnetic resonance imaging; ND = not done; SMA = smooth muscle actin; GIST = gastrointestinal stromal tumor.

was performed in three of the five cases and all showed strong enhancement in the arterial phase. One case [6] also underwent magnetic resonance imaging, but the imaging was unable to differentiate between GIST and a carcinoid tumor. The tumors were located around the prepyloric antrum in all five cases, and the size of the tumors was approximately 2–3 cm.

Gastric glomus tumors are commonly benign lesions but are difficult to distinguish from GISTs or carcinoid tumors by radiography, without specific immunohistochemical staining of FNA samples. In the National Institute of Health classification, tumor size and mitotic count are the major criteria indicating the malignant potential of GISTs. Shah et al proposed that EUS characteristics, including tumor size, extraluminal border, depth and heterogeneity, could be used to predict the malignant potential of GISTs [10]. Thus, definite diagnosis before operation may be necessary and repeated FNA must be considered if the initial FNA does not provide definitive diagnosis. Debol et al reported a gastric glomus with definite preoperative diagnosis through repeated FNA, and the tumor was resected by partial gastrectomy [9]. In our case, EUS revealed a heterogeneous submucosal tumor mimicking a GIST and a partial gastrectomy was subsequently performed because of very low malignant potential. Laparoscopic resection and endoscopic enucleation of gastric glomus tumors have been reported and may offer an alternative surgical procedure [14,15].

Most gastric glomus tumors have benign characteristics, but malignant cases have been reported. Folpe et al proposed a classification scheme with criteria for malignant glomus tumors, including deep location, tumor size more than 2 cm, or the presence of atypical mitotic cells, but it is unclear whether these criteria are suitable for gastric glomus tumors [16]. Furthermore, no long-term follow-up data were available for these cases, meaning criteria for the malignant potential of gastric glomus tumors remain poorly defined.

Here, we report our clinical experience of the fifth gastric glomus tumor in Taiwan, and also review the demographic and clinical presentation of this and four previous cases. The clinical characteristics of gastric glomus tumors, including female dominance, age range, tumor size, and EUS and CT findings were similar to those reported in other countries. It is difficult to differentiate between glomus tumors, GIST and carcinoid tumors by CT, magnetic resonance imaging and EUS. Therefore, invasive and repeated FNA may be needed to avoid extensive surgical resection, particularly in larger tumors.

References

1. Barre JA, Masson P. Etude anatoclinique de certaines tumeurs sousungueales douloureuses (Tumeurs du glomus neuromyo-arterial des extremities L). *Bull De Soc Francaise de Dermologie et Sypheligraphie* 1924;31:148. [In French]
2. Kay S, Callahan WP, Murray MR, et al. Glomus tumors of the stomach. *Cancer* 1951;4:726–36.
3. Lee HW, Lee JJ, Yang DH, et al. A clinicopathologic study of glomus tumor of the stomach. *J Clin Gastroenterol* 2006;40:717–20.
4. Chang SM, Ho WL. Glomus tumor of the stomach: a case report. *Zhonghua Yi Xue Za Zhi (Taipei)*. 1993;52:276–9.
5. Tsai TL, Hsiaw CM, Changchien CS. Hemorrhagic gastric glomus tumor mimics a leiomyosarcoma on both transabdominal and endoscopic ultrasonography: case report. *Changgeng Yi Xue Za Zhi* 1999;22:687–91.
6. Liu KL, Wang HP, Tseng WY, et al. Glomus tumor of the stomach: MRI findings. *AJR Am J Roentgenol* 2005; 185:1190–2.
7. Yan SL, Yeh YH, Chen CH, et al. Gastric glomus tumor: a hypervascular submucosal tumor on power Doppler endosonography. *J Clin Ultrasound* 2007;35:164–8.
8. Miettinen M, Paal E, Lasota J, et al. Gastrointestinal glomus tumors: a clinicopathologic, immunohistochemical, and molecular genetic study of 32 cases. *Am J Surg Pathol* 2002;26:301–11.
9. Debol SM, Stanley MW, Mallery S, et al. Glomus tumor of the stomach: cytologic diagnosis by endoscopic ultrasound-guided fine-needle aspiration. *Diagn Cytopathol* 2003;28:316–21.
10. Vinette-Leduc D, Yazdi HM. Fine-needle aspiration biopsy of a glomus tumor of the stomach. *Diagn Cytopathol* 2001;24:340–2.
11. Gu M, Nguyen PT, Cao S, Lin F. Diagnosis of gastric glomus tumor by endoscopic ultrasound-guided fine needle aspiration biopsy: a case report with cytologic, histologic, and immunohistochemical studies. *Acta Cytol* 2002;46:560–6.
12. Shah P, Gao F, Edmundowicz SA, et al. Predicting malignant potential of gastrointestinal stromal tumors using endoscopic ultrasound. *Dig Dis Sci* 2009;54:1265–9.
13. Tsai TL, Changchien CS, Hu TH, et al. Demonstration of gastric submucosal lesions by high-resolution transabdominal sonography. *J Clin Ultrasound* 2000;28:125–32.
14. Vanwijnsberghe S, Rubay R, Descamps C, et al. A glomic tumour of the stomach treated by laparoscopy. *Acta Chir Belg* 2006;106:613–5.
15. Park YS, Park SW, Kim TI, et al. Endoscopic enucleation of upper-GI submucosal tumors by using an insulated-tip electrosurgical knife. *Gastrointest Endosc* 2004;59:409–15.
16. Folpe AL, Fanburg-Smith JC, Miettinen M, et al. Atypical and malignant glomus tumors: analysis of 52 cases, with a proposal for the reclassification of glomus tumors. *Am J Surg Pathol* 2001;25:1–12.

胃脈絡球腫瘤－病例報告及文獻回顧

黃仲琦[1] 余方榮[2] 詹昌明[2,3] 楊曉芳[4,5] 郭禹廷[6] 謝建勳[1,7] 王照元[1,7]

高雄醫學大學附設醫院 [1]外科 [2]內科 [5]病理科 [6]放射線科

高雄醫學大學 醫學院醫學系 [3]內科 [4]病理科 [7]外科

Glomus tumors 通常發生在四肢的甲床下，很少會發生在臟器，我們報告在本院發現的第一個胃部的 **glomus tumor**。這位病患是 **37** 歲的女性因上腹部間斷性疼痛而至本院胃腸內科接受內視鏡檢查，因而發現在胃竇有一黏膜下的腫瘤，經內視鏡超音波及電腦斷層檢查，發現這是一個血流豐富的腫瘤，但手術前無法確定腫瘤類別。經由外科以約 **4** 公分的手術傷口施行迷你剖腹手術，並施行局部胃腫瘤切除手術，最後病理報告的免疫染色呈現 **SMA**（**Smooth Muscle Actin**）陽性，確定是發生在胃的 **glomus tumor**。同時我們回顧過去曾在臺灣報告過的 **4** 個病例，並與其他國家的胃部 **glomus tumor** 做比較並回顧文獻。

關鍵詞：內視鏡超音波，脈絡球腫瘤，迷你剖腹手術，胃

（高雄醫誌 **2010;26:321–6**）

收文日期：**98** 年 **9** 月 **23** 日

接受刊載：**98** 年 **10** 月 **28** 日

通訊作者：王照元教授

高雄醫學大學附設醫院外科

高雄市三民區自由一路 **100** 號