Case Report

Gastric Schwannoma Presenting with Perforation and Abscess Formation: A Case Report and Literature Review

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Gastric Schwannoma represent only 0.2% of all gastric tumors and 4% of all benign gastric neoplasms. They are usually asymptomatic but can present with variable symptoms. The authors report a case of a 29-year-old male patient who presented with fever and abdominal pain with epigastric mass. Pre-operative diagnosis was gastric lymphoma with perforation and an abscess formation. Hemigastrectomy with Billroth II anastomosis was performed. The pathologic examination and immunohistochemical studies confirmed gastric Schawannoma as the diagnosis. The post-operative course was uneventful. Gastric Schwannoma are difficult if not impossible to diagnose preoperatively as endoscopic and radiologic findings are nonspecific. The treatment of choice is complete surgical resection because of diagnostic uncertainty and the long-term outcome is excellent. This is the first report of gastric Schwannoma presenting with concealed perforation and an abscess formation. The literature was reviewed.

Keywords: *Gastric Schwannoma, Perforation, Intrabdominal abscess*

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Schwannomas, also known as neurinomas or neurilemmomas, are neurogenic tumors originating from any nerve that has a Schwann cell sheath[1]. Schwannomas most commonly occur in the soft tissue of the head and neck, as well as on the flexor surfaces of the extremities[2]. The occurrence of GI tract Schwannomas are extremely rare[1-3] and the most common site is the stomach[4-10]. Gastric Schwannomas constituting only 0.2% of all gastric tumors and 4% of all benign gastric neoplasms[1,11-14].

GI tract Schwannoma was first reported by Daimaru et al in 1988 from Japan[7]. They reported that 24 out of 306 diagnosed GI spindle cell tumor of GI tract were determined to be GI tract Schwannoma by immunohistochemical method. Since then, only a few series have been reported in the pathological literature[3,4,8-10]. Reports of this condition outside the pathological literature have been limited to case reports[5,14]. Loffeld et al in 1998 reported a case of UGI bleeding due to gastric Schwannoma[6].

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Sarlomo-Rikala and Miettinen in 1995 reported six gastric Schwannoma found among 150 mesenchymal tumors of the GI tract (4%)[8]. Patients with gastric Schwannoma are usually asymptomatic and usually discovered incidentally during UGI examination, however, they may present with the symptoms of abdominal pain or discomfort, occult or overt UGI bleeding from the ulceration of the overlying mucosa[4,8-10,15]. A palpable mass may be present when the tumors is larger and predominantly exophytic[1,4,7,8,14].

Few cases of gastric submucosal tumors accompanied with an abscess formation have been reported. There were two reports from gastrointestinal stromal tumor (GIST)[16,17], one report from leiomyosarcoma[18] and one report from gastrointestinal autonomic nerve tumor[19], respectively, but none from gastric Schwannoma. To the authors' knowledge, this is the first reported case of gastric Schwannoma presenting with perforation and an abscess formation.

The present study was conducted with the approval of the institutional ethics boards of Ramathibodi Hospital.

Case Report

A 29-year-old man was referred to Ramathibodi Hospital with a history of fever with abdominal pain

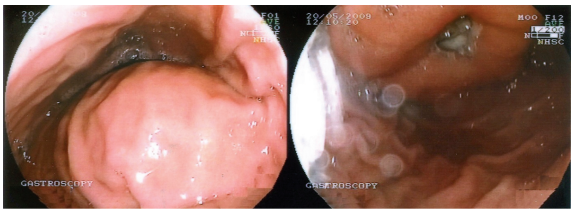

Fig. 1 Gastroscopic view of a submucosal mass with ulceration

for two weeks. He had been well until two weeks earlier, when fever and abdominal pain with epigastric mass developed. He had no special medical history. On physical examination, he was found to have a painful, ill-defined palpable mass in the epigastric area with tenderness, size 6 to 8 cm. He had a high fever (39°C). Laboratory tests showed only leukocytosis. Abdominal ultrasonography showed thickening stomach wall with a fluid collection at the midline, below the abdominal wall. Perforation with localized fluid collection was suspected. Gastroscopy revealed a submucosal mass at the anterior aspect of the body of stomach. The elevated lesion had two ulcerations of 5 mm in diameter, one in the center and one at the edge of the tumor (Fig. 1). Biopsy specimens obtained from the ulcer resembled only fibrinonecrotic tissue without evidence of malignancy, consistent with acute ulcer. CT scan showed a large exophytic solid mass, size 6.2 x 6.2 x 4.7 cm, arising from anterior wall of gastric body with ulcerated lesion and perforation, likely to be gastric lymphoma (Fig. 2). A large multiloculated mixed solid and cystic mass, size 10.9 x 7.1 x 13.9 cm, adjacent to the exophytic solid mass of the stomach, could be abscess formation. There was no evidence of distant metastasis. The patient was treated with IV antibiotics. Subsequent to improvement in his clinical condition, surgery was performed. Operative findings showed a large exophytic mass at anterior gastric antrum with two ulcerations (one central ulceration and one perforated ulcer with pus collection about 100 ml, wall off by anterior abdominal wall, left lobe of liver and transverse colon). No gross intraabdominal lymphadenopathy or metastasis was found. Hemi-gastrectomy with Billroth II anastomosis was performed. Macroscopic examination revealed a 6 x 7 x 7 cm, well circumscribed exophytic, intramural tumor protruding from the serosal surface of gastric antrum (Fig. 3, 4). The cut surface was yellow-tan with central ulcerations and an abscess formation from perforation of an ulcer at the edge of the tumor (Fig. 5). Microscopic examination revealed a spindle cell tumor compatible with Schwannoma. Mitotic activity was 0-2 mitoses/50HPF. The surgical margins were free of tumor. Immunohistochemically, the tumor was positive for S-100 protein and negative for CD117 and smooth muscle actin (SMA). The histologic and immunohistochemical features were consistent with Schwannoma. Two months after the operation, the patient was well with no recurrence.

Discussion

Gastric Schwannomas are rare tumors that belong to the family of gastrointestinal mesenchymal

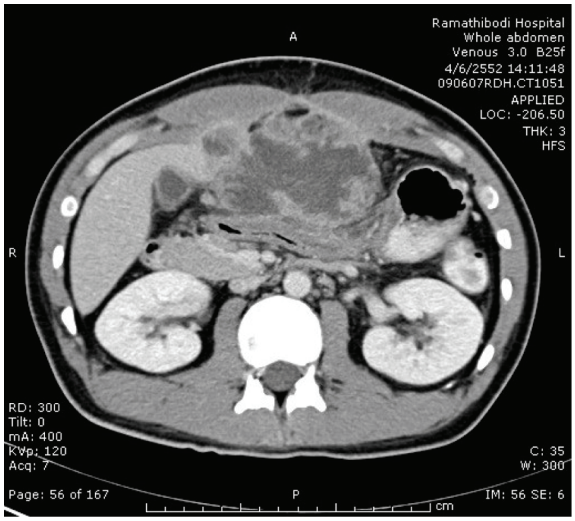

Fig. 2 CT scan showing a large exophytic solid mass, size 6.2 x 6.2 x 4.7 cm, arising from anterior wall of gastric body with perforation

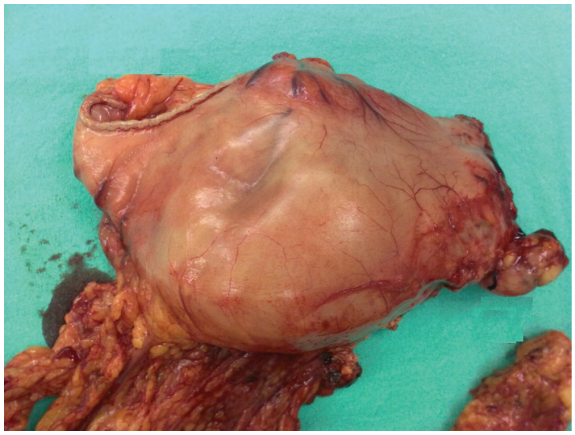

Fig. 3 The resected specimen – a hemigastrectomy

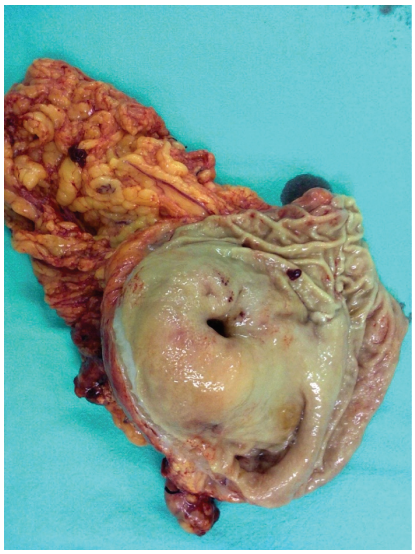

Fig. 4 Resected specimen opened to show a 6 x 7 x 7 cm, well circumscribed exophytic, intramural tumor protruding from the serosal surface of gastric antrum

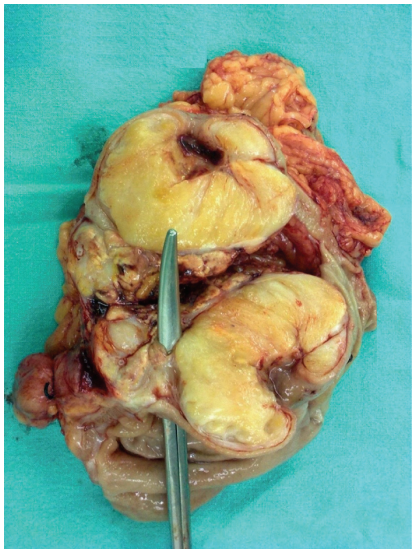

Fig. 5 The cut surface of the specimen revealed a yellow-tan mass with central ulcerations

tumors of which the most common (about 80%) are GIST[3,13,20]. Schwannomas have been reported to represent only 3%-12% of all GI mesenchymal tumors[3,7-10,21,22]. The tumors were most frequently thought to be GIST before surgery[3]. None had a correct definitive preoperative diagnosis[3]. These tumors occur in patients with an average age of 50-60 years[9,10]. Most series report a female preponderance[4,7,9].

GI tract Schwannoma differed from conventional soft tissue Schwannoma in that they had peripheral lymphoid cuffs, lacked fibrous capsules and rarely showed degenerative change such as necrosis, hemorrhage and cytic change[9-11]. GI Schwannoma are not encapsulated, although most are well circumscribed with pseudocapsule[9]. Macroscopically, these are homogeneous, rubbery to firm. The cut surface appeared whitish-yellow and glistening[2]. Gastric Schwannomas are usually detected preoperatively via endoscopy or cross-sectional imaging. However, preoperative diagnosis is difficult as none of these modalities have shown any pathognomonic features unique to these tumors[3]. Endoscopic evaluation are usually nonspecific as these tumors appear grossly as submucosal or exophytic mass, which are indistinguishable from other submucosal lesions. Furthermore, as in the presented case, endoscopic biopsies are usually not representative of the deeper submucosal tissue. Even when the endoscopist succeeded in obtaining samples from fine-needle aspiration, those usually demonstrate non-specific spindle cells, and there is usually insufficient tissue for the pathologist to obtain a definite diagnosis[3].

Endoscopic sonography (EUS) has been considered the best modality in diagnosis of a gastric submucosal tumors[23]. EUS can assess the size and origin of a gastric submucosal tumor and define the location and extent of the tumor with displacement of surrounding organs or vessels. From EUS, gastric Schwannoma had homogeneous hypoechoic mass compared to the normal surrounding muscle layers that is contiguous with the fourth hypoechoic layers of the normal gastric wall, with the presence of a marginal halo, which histopathologically corresponded to the peripheral lymphoid cuff[23]. Fujii et al in 2004 showed the first case reported of sonographic findings of gastric Schwannoma[24]. Sonography showed a homogeneously hypoechoic mass. The reduction of acoustic impedance due to the dense composition of spindle cells seems to cause the lower echogenicity than that of the normal muscularis propria[24]. CT scan and MRI are useful in the detection and characterization of large gastric submucosal or exophytic tumor and its relation with surrounding organs. Gastric Schwannomas have benign features, *i.e.*, discrete margin, homogeneous enhancement and no lymphadenopathy[2,3,11,25].

Calcification is uncommon[25]. Karabulut et al in 2007 reported the first time of gastric Schwannoma with MRI documentation[26].

Definite diagnosis of gastric Schwannoma is made by microscopic examination and immunohistochemical study[4]. Ultrastructural confirmation with electromicroscopy is not necessary[4]. Histologically, the tumor is composed of spindle cells with mild nuclear atypia and rare mitoses[2]. The tumors were located in the muscle layer. There was a peripheral cuff-like lymphocytic infiltration[2,8]. Schwannoma can be distinguished from other gastric mesenchymal tumors based on immunohistochemical findings. Gastric Schwannomas express S-100 protein, vimentin, and glial fibrillary acidic protein (GFAP) but CD117, CD34, smooth muscle actin (SMA), and desmin are uniformly negative[7,9]. Hence, immunohistochemical study is extremely useful in distinguishing Schwannoma from GIST, which express CD117 (usually) and CD34 (frequently) and true smooth muscle tumors, which express SMA and desmin[3,8,27]. Gastrointestinal autonomic nerve tumors (GANTs) constitute a distinct subcategory of GIST that are distinguishable from gastrointestinal tumor of peripheral nerve sheath origin (Schwannoma)[19]. If we compare Schwannoma and GANTs, some differences are noticeable. Schwannoma are spindle cell tumor without any epithelioid features and always have a peripheral cuff of lymphoid aggregates; these features are only occasional in GANTs. GANTs also have a variable immunohistochemical profile like GIST[4]. The separation of these two entities is important for practical purpose. GANTs pursue an aggressive course in more than 50% of cases[4,28,29], whereas gastric Schwannoma behave in a benign fashion[2,3,7-11]. Malignant transformation is very rare, and only two cases have been reported in the literatures by Bee et al in 1997[30] and Loffeld et al in 1998[8].

In clinical practice, preoperative differentiation between GI mesenchymal tumors according to histogenesis is usually difficult. Although most of the gastric Schwannoma are uniformly benign and have an excellent prognosis, the treatment of choice of gastric Schwannoma is complete surgical resection in fit, healthy patients, as it is impossible to distinguish those tumors from GIST and smooth muscle tumor, which have a high risk of aggressive behavior[1,3,14,15].

Conclusion

Here the authors present a young man with a gastric Schwannoma manifesting as a painful palpable abdominal mass with concealed perforation and an abscess formation. This is the first reported case of this condition. Although gastric Schwannoma are rare tumors, they should be included in the differential diagnosis of mesenchymal tumors arising in the stomach.

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Potential conflicts of interest

None.

References

1. Yoon HY, Kim CB, Lee YH, Kim HG. Gastric schwannoma. Yonsei Med J 2008; 49: 1052-4.
2. Hong HS, Ha HK, Won HJ, Byun JH, Shin YM, Kim AY, et al. Gastric schwannomas: radiological features with endoscopic and pathological correlation. Clin Radiol 2008; 63: 536-42.
3. Goh BK, Chow PK, Kesavan S, Yap WM, Ong HS, Song IC, et al. Intraabdominal schwannomas: a single institution experience. J Gastrointest Surg 2008; 12: 756-60.
4. Prevot S, Bienvenu L, Vaillant JC, Saint-Maur PP. Benign schwannoma of the digestive tract: a clinicopathologic and immunohistochemical study of five cases, including a case of esophageal tumor. Am J Surg Pathol 1999; 23: 431-6.
5. Khan AA, Schizas AM, Cresswell AB, Khan MK, Khawaja HT. Digestive tract schwannoma. Dig Surg 2006; 23: 265-9.
6. Loffeld RJ, Balk TG, Oomen JL, van der Putten AB. Upper gastrointestinal bleeding due to a malignant Schwannoma of the stomach. Eur J Gastroenterol Hepatol 1998; 10: 159-62.
7. Daimaru Y, Kido H, Hashimoto H, Enjoji M. Benign schwannoma of the gastrointestinal tract: a clinicopathologic and immunohistochemical study. Hum Pathol 1988; 19: 257-64.
8. Sarlomo-Rikala M, Miettinen M. Gastric schwannoma - a clinicopathological analysis of six cases. Histopathology 1995; 27: 355-60.
9. Kwon MS, Lee SS, Ahn GH. Schwannomas of the gastrointestinal tract: clinicopathological features of 12 cases including a case of esophageal tumor compared with those of gastrointestinal stromal tumors and leiomyomas of the gastrointestinal tract. Pathol Res Pract 2002; 198: 605-13.

10. Hou YY, Tan YS, Xu JF, Wang XN, Lu SH, Ji Y, et al. Schwannoma of the gastrointestinal tract: a clinicopathological, immunohistochemical and ultrastructural study of 33 cases. Histopathology 2006; 48: 536-45.
11. Levy AD, Quiles AM, Miettinen M, Sobin LH. Gastrointestinal schwannomas: CT features with clinicopathologic correlation. AJR Am J Roentgenol 2005; 184: 797-802.
12. Chaudry NU, Zafar S, Israr uH. Benign nerve sheath tumor of stomach. J Coll Physicians Surg Pak 2007; 17: 277-9.
13. Lin CS, Hsu HS, Tsai CH, Li WY, Huang MH. Gastric schwannoma. J Chin Med Assoc 2004; 67: 583-6.
14. Melvin WS, Wilkinson MG. Gastric schwannoma. Clinical and pathologic considerations. Am Surg 1993; 59: 293-6.
15. Kim JY, An JY, Choi MG, Noh JH, Sohn TS, Kim S. A case of giant gastric schwannoma accompanied with bleeding. J Korean Gastric Cancer Assoc 2006; 6: 47-51.
16. Osada T, Nagahara A, Kodani T, Namihisa A, Kawabe M, Yoshizawa T, et al. Gastrointestinal stromal tumor of the stomach with a giant abscess penetrating the gastric lumen. World J Gastroenterol 2007; 13: 2385-7.
17. Nozawa S, Bando T, Nagata T, Tsukada K. Abscess formation in a giant gastrointestinal stromal tumor of the stomach following endoscopic biopsy. Endoscopy 2006; 38: 955.
18. Seidel RH Jr, Burdick JS. Gastric leiomyosarcoma presenting as a gastric wall abscess. Am J Gastroenterol 1998; 93: 2241-4.
19. Honda K, Mikami T, Ohkusa T, Takashimizu I, Fujiki K, Araki A, et al. Gastrointestinal autonomic nerve tumor with giant abscess. A case report and literature review. J Clin Gastroenterol 1997; 24: 280-5.
20. Snyder RA, Harris E, Hansen EN, Merchant NB, Parikh AA. Gastric schwannoma. Am Surg 2008; 74: 753-6.
21. Miettinen M, Sarlomo-Rikala M, Lasota J. Gastrointestinal stromal tumors: recent advances in understanding of their biology. Hum Pathol 1999; 30: 1213-20.
22. Miettinen M, Virolainen M, Maarit SR. Gastrointestinal stromal tumors—value of CD34 antigen in their identification and separation from true leiomyomas and schwannomas. Am J Surg Pathol 1995; 19: 207-16.
23. Jung MK, Jeon SW, Cho CM, Tak WY, Kweon YO, Kim SK, et al. Gastric schwannomas: endosonographic characteristics. Abdom Imaging 2008; 33: 388-90.
24. Fujii Y, Taniguchi N, Hosoya Y, Yoshizawa K, Yasuda Y, Nagai H, et al. Gastric schwannoma: sonographic findings. J Ultrasound Med 2004; 23: 1527-30.
25. Levy AD, Remotti HE, Thompson WM, Sobin LH, Miettinen M. Gastrointestinal stromal tumors: radiologic features with pathologic correlation. Radiographics 2003; 23: 283-304.
26. Karabulut N, Martin DR, Yang M. Case report: gastric schwannoma: MRI findings. Br J Radiol 2002; 75: 624-6.
27. Fletcher CD, Berman JJ, Corless C, Gorstein F, Lasota J, Longley BJ, et al. Diagnosis of gastrointestinal stromal tumors: A consensus approach. Hum Pathol 2002; 33: 459-65.
28. Herrera GA, Cerezo L, Jones JE, Sack J, Grizzle WE, Pollack WJ, et al. Gastrointestinal autonomic nerve tumors. 'Plexosarcomas'. Arch Pathol Lab Med 1989; 113: 846-53.
29. Lauwers GY, Erlandson RA, Casper ES, Brennan MF, Woodruff JM. Gastrointestinal autonomic nerve tumors. A clinicopathological, immunohistochemical, and ultrastructural study of 12 cases. Am J Surg Pathol 1993; 17: 887-97.
30. Bees NR, Ng CS, Dicks-Mireaux C, Kiely EM. Gastric malignant schwannoma in a child. Br J Radiol 1997; 70: 952-5.

รายงานผู้ป่วยชวานโนมา (Schwannoma) ของกระเพาะอาหารที่มาด้วยเนื้องอกทะลุและเกิดโพรงหนองพร้อมทบทวนวรรณกรรม

จักรพันธ์ เอื้อนรเศรษฐ์, วีรพัฒน์ สุวรรณธรรมา

เนื้องอกชวานโนมาของกระเพาะอาหารพบได้เพียง 0.2% ของเนื้องอกกระเพาะอาหารทั้งหมด หรือ 4% ของเนื้องอกชนิดไม่ร้ายแรงของกระเพาะอาหาร เนื้องอกชนิดนี้ ส่วนใหญ่มักไม่มีอาการ ถ้ามีอาการจะมีอาการได้หลายแบบไม่จำเพาะ ผู้นิพนธ์ได้รายงานผู้ป่วยชาย อายุ 29 ปี มาด้วยอาการ ไข้ ปวดท้อง และมีก้อนที่ท้องบริเวณลิ้นปี่ การวินิจฉัยโรคก่อนผ่าตัด สงสัยเป็นเนื้องอกของกระเพาะอาหาร ชนิดเม็ดเลือดขาวที่แตกทะลุและมีโพรงหนอง การรักษาได้ทำผ่าตัดกระเพาะอาหารออกครึ่งหนึ่ง ผลการตรวจชิ้นเนื้อทางพยาธิ และการย้อมอิมมูโนฮีสโตเคมีแสดงว่าเป็นเนื้องอกชวาลโนมาของกระเพาะอาหาร หลังผ่าตัดไม่มีภาวะแทรกซ้อน โรคเนื้องอกชวานโนมาของกระเพาะอาหาร ทำการวินิจฉัยก่อนผ่าตัดได้ยาก เพราะการตรวจด้านการส่องกล้องกระเพาะอาหารและรังสีวินิจฉัยไม่มีลักษณะจำเฉพาะ การรักษาคือการผ่าตัดเอาก้อนออกหมด เพราะว่ามักไม่ได้การวินิจฉัยก่อนผ่าตัด และผลการรักษาผ่าตัดได้ผลดีมาก ผู้นิพนธ์ได้ทบทวนรายงานวารสารทางการแพทย์เกี่ยวกับเรื่องนี้ รายงานนี้เป็นรายงานแรกของประเทศไทยของเนื้องอกชวานโนมาของกระเพาะอาหาร ที่มาด้วยเนื้องอกทะลุและเกิดโพรงหนอง