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A Gastric Schwannoma Misdiagnosed as B Cell Lymphoma

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Abstract

Schwannomas are benign, slow-glowing tumor, and they are very rare. Schwannomas are only 0.2% of all gastric neoplasm, and 4% of benign gastric neoplasm. Schwannomas are histologically composed of mostly spindle cells and peripheral cuff of lymphoid cells aggregations. Peripheral lymphoid cuff of schwannomas is usually focal and thin, but it may rarely have thick band-like lymphoid tissue. Here we report a 53-year-old male, who was initially suspicious of MALT lymphoma in endoscopic biopsy due to thick lymphoid tissue, and diagnosed gastric schwannoma by partial gastrectomy later for the diagnostic confirmation. This case report is written to report a case of misdiagnosis and to tell the reason why repeated biopsy is important.

Key Words : Lymphoid cells, Schwannoma, Stomach

Introduction

Schwannomas, arising from Schwann cells, are benign tumors which rarely develop in the gastrointestinal tract. And among the GI tract, stomach is the most commonly occurring place [1]. Schwannomas are benign, slow-glowing tumor, and they are very rare. Schwannomas are only 0.2% of all gastric neoplasms, and 4% of benign gastric neoplasm [2]. They mainly occur in mid-aged, age between 40 to 60, females [2]. They can also occur in children. And in rare cases schwannomas transform into malignant tumors [3].

Schwannomas are mostly asymptomatic, but in some cases, symptomatic patients present with ulcerations and bleeding. Gastrointestinal endoscopy is the principal diagnostic tool. However, to differentiate Schwann cell proliferation from other submucosal tumors is very difficult on pre-operative examination. Schwannomas have a

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characteristic cuff of lymphoid aggregates around their periphery. The nuclear palisading, verocay bodies, and hyalinized vessels seen in schwannomas elsewhere in the body are less common or are absent in the gut [4].

In this case, lymphoid cuff was harvested by biopsy. And it was look like B cell lymphoma. Although the mass was a gastric schwannoma, it was misdiagnosed as B cell lymphoma. And partial gastrectomy was practiced. This case report is written to report a case of misdiagnosis and to tell the reason why repeated biopsy is important.

Case Report

A-53-year-old man was referred because of abnormal finding in the biopsy specimen obtained by esophgo-gastro-duodenoscopy in local clinic.

The patient had typhoid fever, Tb pleuritis, and *H. pylori* gastritis previously. In the routine esophago-gastro-duodenoscopy, a small mass was detected in the stomach without any other gastrointestinal symptom. After follow up for a year, the gastric mass grows slowly. So, in a suspicious of GIST, a deep biopsy using esophago-gastro-duodenoscopy was done in local clinic. Histologically, numerous lymphoid tissues with cautery artifact were aggregated. Patient referred to our hospital and pathologists review the tissue again.

The specimen show well differentiated, nodular, lymphoid cells under the microscope. Immunohistochemically these lymphoid cells show focal positivity for CD5 and a little more diffuse positivity reaction for CD20. But there was no reaction for CD23 and cyclin D1 (Fig. 1).

CT-scan of his abdomen and pelvis showed a 25 mm-sized round enhancing exophytic submucosal tumor in gastric antrum with overlying intact mucosal layer (Fig. 2). And two lymph nodes in gastroepiploic area seemed suspicious.

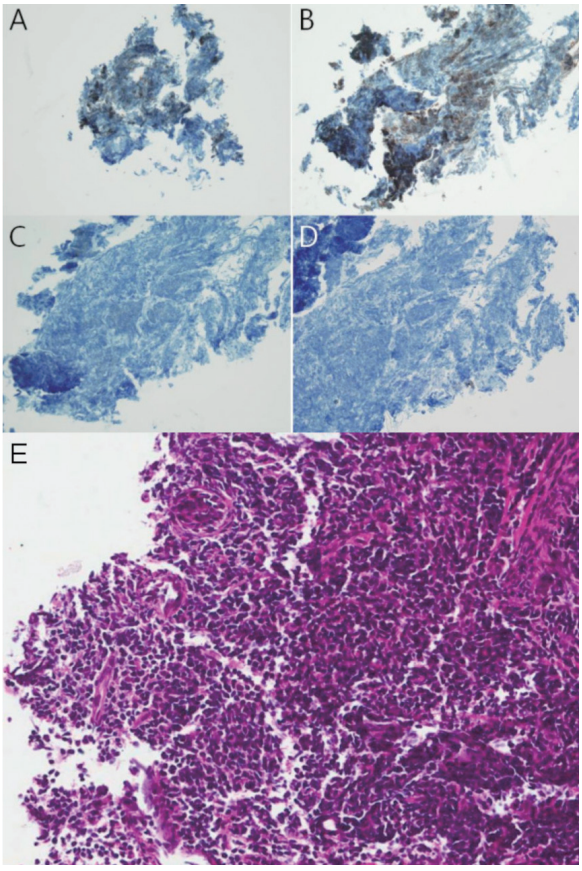

Fig. 1. (A) Lymphoid aggregates with cautery artifact shows focal positivity for CD5. (B) A Little more diffuse positivity reaction for CD20. (C) But no reaction for CD23. (D) Cyclin D1 is seen. (E) Numerous small-sized lymphoid cells are aggregates with severe cautery artifact.

Radionuclide PET scan reveals a round exophyting mass (SUVmax: 3.9, 25×21 mm) at anterior wall and greater curvature of the gastric antrum with eccentric mild FDG uptake (Fig. 3).

According to the past history of *H. pylori* gastritis [5] and histologic finding such as lymphoid tissue aggregates, MALT lymphoma was suspected. But the distinct mass formation and only lymphoid

tissue with severe cautery artifact were not proper to the diagnosis of MALT lymphoma. So, for the diagnostic confirmation and removal of the mass and suspicious lymph nodes distal gastrectomy and D2 lymph node dissection was done.

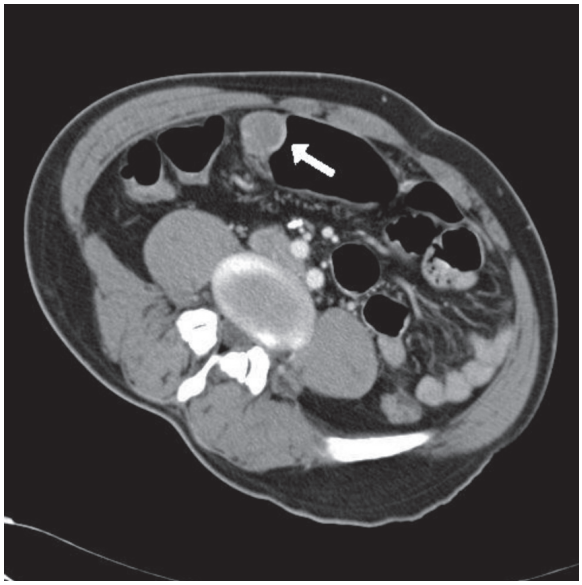

Fig. 2. A round exophytic submucosal tumor of gastric antrum was enhanced in CT scan.

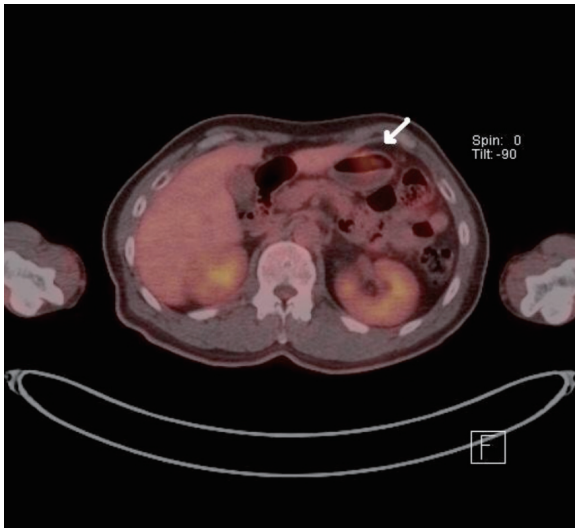

Fig. 3. PET scan reveals a round exophyting mass (SUVmax: 3.9, 25×21 mm) at anterior wall and greater curvature of gastric antrum with eccentric mild FDG uptake.

There was a gastric submucosal mass with overlying intact gastric mucosa, measuring 3.0 cm in maximum diameter. Cut section showed a relatively well-defined, round, pale tan, solid lesion, burging out subserosal layer (Fig. 4).

Beneath the collapsed submucosa, the gastric mass was composed of mostly spindle cell bundles and peripheral thick lymphoid cuff. Verocay bodies with linear arrangement of elongated tumor nuclei, are seen. The tumor cells showed strongly positive for S-100 protein, and no reactivity for CD34, CD117, desmin and smooth muscle actin (Fig. 5).

The patient recovered well after surgery and

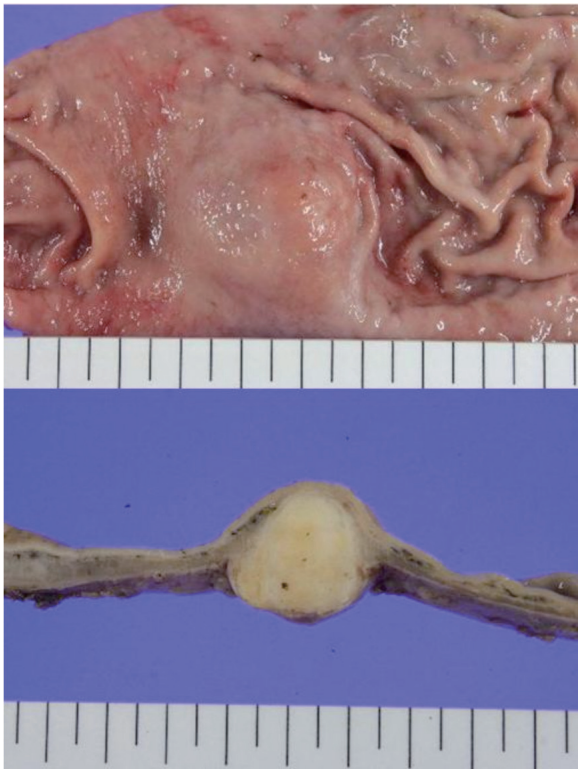

Fig. 4. The gastric mucosa are elevated and expanded by a submucosal mass (upper). Cut section of gastric tumor mass shows a relatively well-defined, round, pale tan, solid lesion, measuring 3.0 cm in maximum diameter (lower).

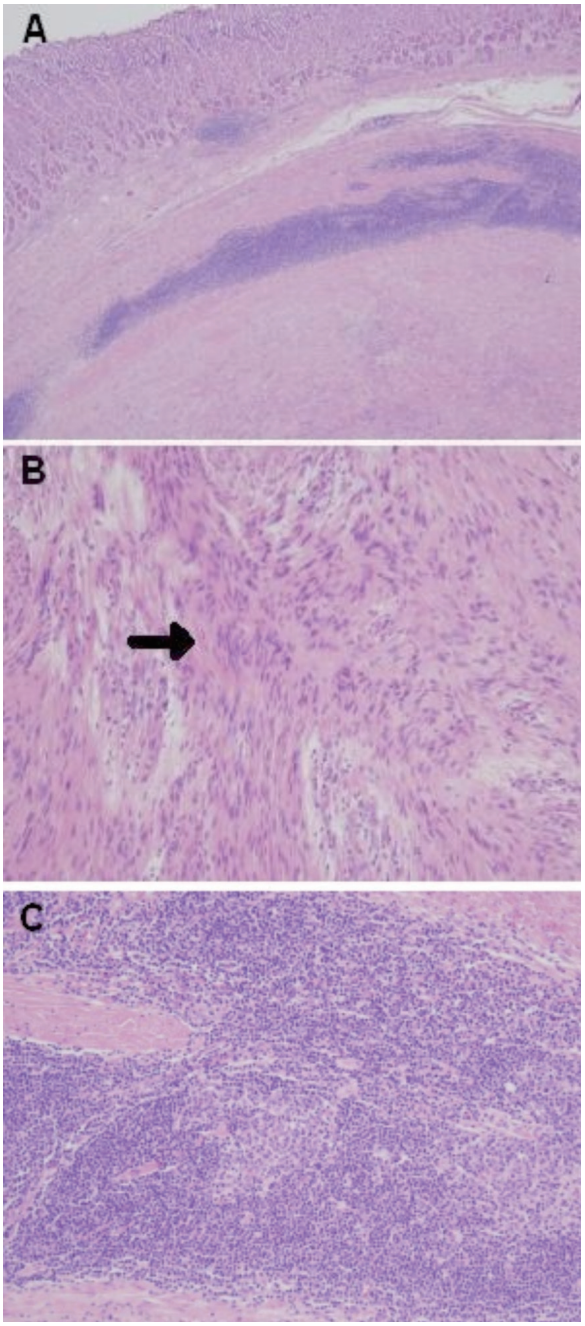

Fig. 5. (A) Beneath the collapsed submucosa, the gastric mass is composed of mostly spindle cell bundles and peripheral thick lymphoid cuff (H&E, ×40). (B) Verocay bodies (arrow), characterized by linear arrangements of elongated tumor nuclei within the gastric mass were seen (H&E, ×200). (C) Lymphoid cuff of schwannoma look similar to a lymphoma (H&E, ×200).

discharged from the hospital 10 days later.

Discussion

Digestive schwannomas are rare, benign, mostly asymptomatic, and non-recurring tumor. Gastric schwannomas are spindle cell tumors without any epithelioid features and usually have a peripheral cuff of lymphoid aggregates [6]. Lymphoid cuff are found in 96% of schwannoma cases [7].

If the lymphoid cuff is especially thick and cautery artifact is severe in the limited biopsy material, the possibility of low grade lymphoma such as MALT lymphoma may not be excluded. In addition, MALT lymphoma is composed of heterogeneous B cells, including small lymphocytes with round nuclei and clumped chromatin, monocytoid cells, and plasmacytoid cells. The tumor cells are positive for CD20, CD79a, CD21, and CD35, and negative for CD5, CD23, CD10, and cyclinD1 [8]. There is no specific immunohistochemical marker for MALT lymphoma yet. Also the lymphoid cuff of schwannoma may reveal positivity for CD20, B cell markers [9].

In this case, the first deep biopsy was targeted to the periphery portion of the gastric mass. So, the biopsy specimen mainly contains peripheral lymphoid cuff, missing the main tumor component. Also, this biopsy material was not proper to the immunohistochemical studies, because the cautery and crushing artifact were severe and the obtained tissue was too small. More biopsies or the surgery for the diagnostic clue were required to identify the type of the gastric mass.

Gastrointestinal stromal tumor (GIST), leiomyoma and schwannoma are gastric mesenchymal tumors. These gastric mesenchymal tumors are composed of spindle shaped cell and look similar under light microscopic examinations

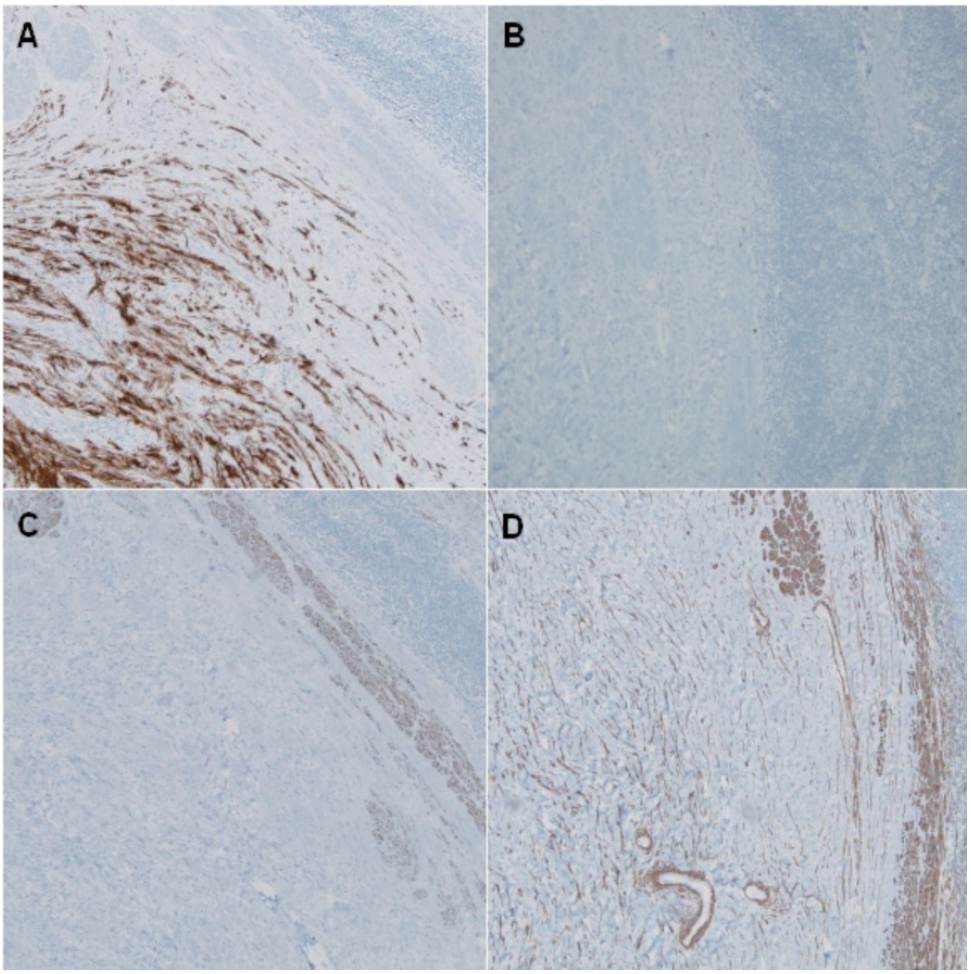

Fig. 6. (A) The tumor cells shows strong positivity for only S-100 protein (×100). (B) Negative reaction for CD117 (×100). (C) Negative reaction desmin (×100). (D) Negative reaction smooth muscle specific actin (×100).

[10,11]. In schwannoma, verocay body and strong S-100 protein positivity in tumor cells are characteristics [12,13]. But in many schwannomas without verocay body, to differentiate it from GIST only histologically, is not easy [14]. At this time, the immunohistochemical staining is needed. GISTs are positive for CD117, CD34 and vimetin. Schwannomas are positive for S-100, and negative for CD34, CD117, desmin and actin (Fig. 6).

This misdiagnosis was able to be avoided, if biopsy was repeated. Submucosal tumors are hard to be diagnosed in initial biopsy. So repetition of biopsy is needed to confirm a submucosal tumor. But repeated biopsies are often not done, due to the discomfort of the patient. Although the techniques of diagnostic radiology made rapid progress, immunohistochemistry is the best way to confirm the character of a tissue. If you find a mass with lymphoid cells but not showing characteristics of any know lymphoma, suspect lymphoid cuff and do biopsy again.

Summary

Gastric schwannomas should be differentiated from low grade lymphoma like this biopsy material, as well as other mesenchymal tumors. For this, the histologic characteristics such as spindle cell growth, verocay body, and the positivity for S100-protein in proper tissue, are needed. If the biopsy specimen of mass-forming lesion contains only lymphoid tissue, the possibility of peripheral lymphoid cuff of schwannoma and missing main tumor cells should be considered. Accurate diagnosis in the proper representative tissue is important to avoid unnecessary surgery.

References

1. Yoon HY, Kim CB, Lee YH, Kim HG. Gastric schwannoma. *Yonsei Med J* 2008;**49**:1052-4.
2. Melvin WS, Wilkinson MG. Gastric schwannoma. Clinical and pathologic considerations. *Am Surg* 1993;**59**:293-6.
3. Bees NR, Ng CS, Dicks-Mireaux C, Kiely EM. Gastric malignant schwannoma in a child. *Br J Radiol* 1997;**70**:952-5.
4. Greenson JK. Gastrointestinal stromal tumors and other mesenchymal lesions of the gut. *Mod Pathol* 2003;**16**:366-75.
5. Hsi ED, Singleton TP, Swinnen L, Dunphy CH, Alkan S. Mucosa-associated lymphoid tissue-type lymphomas occurring in post-transplantation patients. *Am J Surg Pathol* 2000;**24**:100-6.
6. 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.
7. Voltaggio L, Murray R, Lasota J, Miettinen M. Gastric schwannoma: a clinicopathologic study of 51 cases and critical review of the literature. *Hum Pathol* 2012;**43**:650-9.
8. Bertoni F, Coiffier B, Salles G, Stathis A, Traverse-Glehen A, Thieblemont C, *et al*. MALT lymphomas: pathogenesis can drive treatment. *Oncology (Williston Park)* 2011;**25**:1134-42, 47.
9. Terada T, Inatsuchi H. Small juxtadrenal cellular schwannoma. *Virchows Arch* 2004;**444**:95-7.
10. Lin CS, Hsu HS, Tsai CH, Li WY, Huang MH. Gastric schwannoma. *J Chin Med Assoc* 2004;**67**:583-6.
11. Abraham SC. Distinguishing gastrointestinal stromal tumors from their mimics: an update. *Adv Anat Pathol* 2007;**14**:178-88.
12. Mohammadi A, Rosa M, Rhatigan R. Verocay body prominent schwannoma of penis: an unusual localization for this lesion. *J Cutan Pathol* 2008;**35**:1160-2.
13. Sarlomo-Rikala M, Miettinen M. Gastric schwannoma--a clinicopathological analysis of six cases. *Histopathology* 1995;**27**:355-60.
14. Guthrie G, Mullen R, Moses A. Gastric Schwannoma or GIST: accuracy of preoperative diagnosis? *Scott Med J* 2011;**56**:236.