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CASE REPORT

Gastric Schwannoma in an elderly man: A case report

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Abstract

Schwannomas are mesenchymal tumors that are characteristically benign and slow growing, which originate from any nerve with Schwann cell sheath. Gastrointestinal schwannomas are rare with distinct morphologic features as compared to schwannomas of soft tissue or central nervous system. A 77-year-old male patient was diagnosed with gastrointestinal stromal tumor based on radiological findings and clinical impression when he presented with worsening abdominal discomfort and pain. He underwent distal gastrectomy however histopathological examination of the tumour revealed schwannoma. This case report presents a rare case of a symptomatic gastric schwannoma, whose definitive diagnosis was established by histopathological and immunohistochemical findings postoperatively.

Keywords: Schwannoma, stomach schwannoma, gastrointestinal schwannoma

INTRODUCTION

Gastrointestinal tract mesenchymal tumours comprised of non-epithelial spindle cell tumours which include GIST, leiomyoma, leiomyosarcoma and schwannoma.[1] Among these neoplasms, the most common primary mesenchymal tumour of the stomach is gastrointestinal stromal tumour (GIST).[2] Schwannomas however, rarely occur in the GI tract. It accounts for 0.2 % of all gastric tumours and 4% of all benign gastric neoplasms.[3]

Gastric schwannoma originates from Schwann cells, which form the neural sheath of the nervous system. The incidence rates of schwannoma are identical for both genders, and the median age is between 60 to 65 years.[4] These tumours have no recurrence, metastasis or tumour-related mortality. Endoscopic and radiological investigations are non-specific making the diagnosis challenging preoperatively. The aim of this article is to highlight the importance of immunohistochemical studies to make a definitive diagnosis, which can only be performed on tissue biopsy or resected specimen.

CASE REPORT

A 77-year-old man with underlying Parkinson's disease, hypertension and diabetes mellitus which were under medical treatment presented with worsening abdominal discomfort and early satiety. There was no weight loss, body weakness or anorexia. Computed tomography (CT) of the abdomen done at a private center showed a large stomach tumour. With this finding, a diagnosis of GIST was entertained, and the patient was then referred to our center for further management.

Oesophago-duodenoscopy was performed at our center and noted there was no overlying ulcer over the tumour. No biopsies were taken at this point of time. After further discussion with the patient, surgery was determined to be the best treatment option. He underwent a partial gastrectomy with roux-en-y bypass procedure. The specimen was sent to histopathology laboratory for further evaluation.

Macroscopic examination of the stomach revealed a solid, thinly encapsulated and well circumscribed submucosal tumour with a rubbery and yellow cut surface that measured 11 x 10 × 9.9cm, located at the posterior surface of the stomach (FIG. 1). There was no gross mucosal infiltration seen.

Microscopically, the tumour was composed of bland spindle cells arranged in fascicles and

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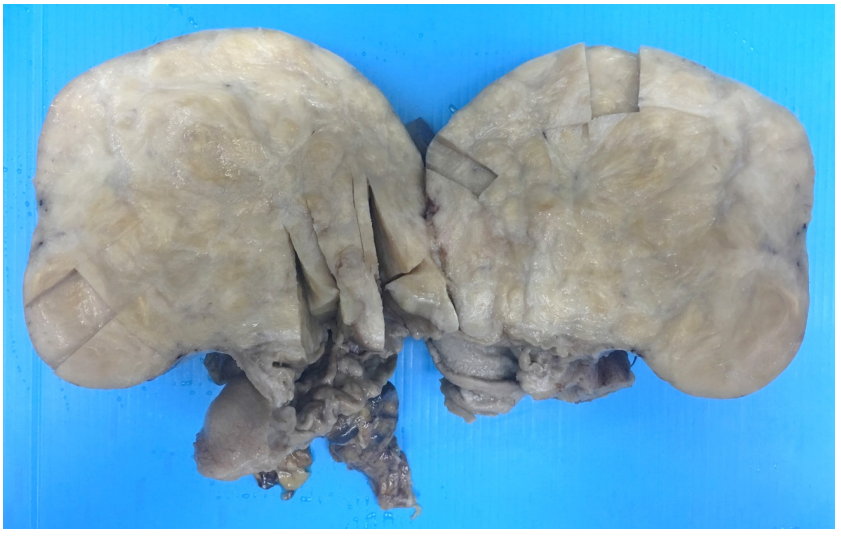

FIG. 1: Macroscopic view of the resected distal gastrectomy specimen showing a well-circumscribed exophytic tumour arising from the posterior surface of the stomach with pale yellow cut surface.

surrounded by rim of lymphoid aggregates (FIG. 2 & 3). No epithelioid cells seen. There was no obvious alternating hypercellular (Antoni A) and hypocellular (Antoni B) area. A palisading Verocay body was also not seen. Focal collections of xanthoma cells and scattered lymphoid aggregates were noted within the tumour. Immunohistochemical studies showed the tumour was diffusely positive to S-100 (FIG. 4). The markers for GIST (CD117 and DOG1) were negative. The tumour cells were also immunonegative to SMA, EMA, CD 34, BCL2, desmin and CK AE1/AE3. The morphological and immunohistochemical features are consistent with schwannoma.

The surgery was uneventful. The patient did not develop any complications postoperatively and was discharged a week later. He was followed up a month after surgery and was noted to be well.

DISCUSSION

Differential diagnosis of primary spindle-shaped mesenchymal tumours are many, the most common of which is GIST. Schwannoma,

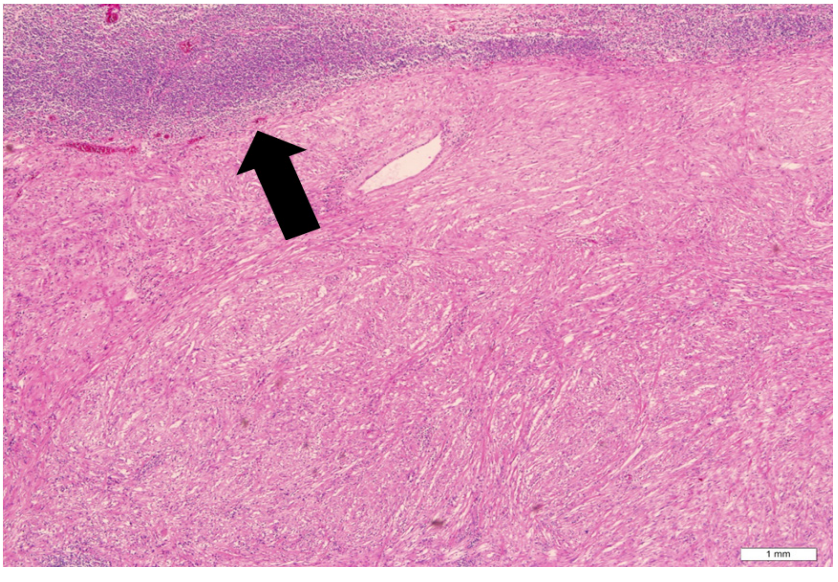

FIG. 2: The submucosal tumour shows peritumoral lymphoid cuff (arrow). (H&E, 4x).

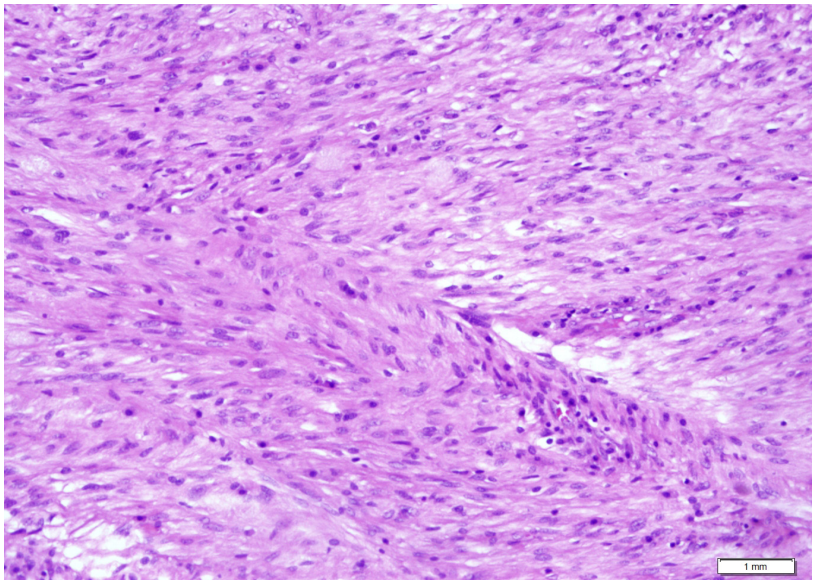

FIG. 3: The tumour is composed of bland spindle cells with oval to elongated slender nuclei arranged in fascicles with scattered lymphocyte infiltrates. (H&E, 20x).

although is a common soft tissue and central nervous system tumour, are rarely found within the gastrointestinal tract (GIT). It is important to differentiate schwannoma from GIST as the latter may have malignant potential. GIT schwannoma has generally benign behavior.[5,6] Other differential diagnosis includes leiomyoma and leiomyosarcoma which are also uncommon and occur in older patients as well as inflammatory myofibroblastic tumour.[1]

Schwannoma is a mesenchymal tumour that arises from Schwann cells. Patients with stomach schwannoma may be asymptomatic but symptoms such as abdominal discomfort, gastrointestinal bleeding, early satiety or even chest pain can be experienced.[6] A palpable mass may also be present when the tumour is large and exophytic.

The most common site for gastrointestinal schwannoma is the stomach with the occurrence within the lower GIT is relatively uncommon.[16] A study that included 33 cases of GIT schwannomas

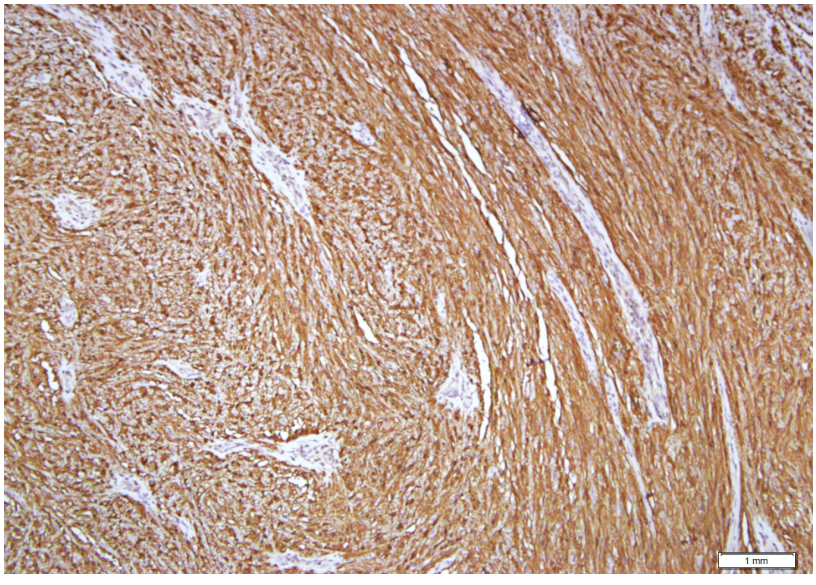

FIG. 4: Schwannoma showing diffuse S100 positivity (10x).

showed that the most common site for this tumour was stomach, with fewer cases reported in the oesophagus, colon and rectum.[5] Another study of 24 cases also revealed that 23 cases of schwannomas arise from the stomach with only one case from the ascending colon.[18] According to a literature review published by Voltaggio *et al*., there is an estimated 45 gastric GISTs reported for every 1 gastric schwannoma.[15]

Endoscopic examination may be normal or appear as non-specific secondary findings such as flattening of gastric folds or ulceration because of pressure necrosis from the tumour.[17] In this case, the scope findings only showed an extrinsic mass effect with no areas of ulceration.

Computed tomographic (CT) scan may show schwannomas as a homogenous, well defined, intramural mass without necrosis, hemorrhage or cystic degeneration, in contrast to GIST.[6] Magnetic resonance imaging (MRI) may also provide additional information regarding the location as well as its relation to adjacent structures. They appear as discrete, strongly enhancing tumours with low to medium signal intensity on T1 weighted images and high signal intensity on T2 weighted images.[7]

On macroscopic examination, schwannomas are well-circumscribed, yellowish-white lesions with cystic pattern. However, based on just gross appearance, it is difficult to differentiate schwannoma from other mesenchymal tumours such as GIST.[14]

Histologically, a typical schwannoma of the soft tissue and central nervous system are encapsulated and comprises of spindle cells arranged in alternating hypercellular (Antoni A) -hypocellular (Antoni B) pattern with presence of palisading nuclear area called Verocay bodies. Whereby, gastric schwannomas are usually encased by intact mucosa and principally involve the submucosa and muscularis propria, without invading the surrounding structures.[15] A prominent lymphoid aggregates, sometimes forming germinal centers surrounding the tumour with absence of Verocay bodies, Antoni A or Antoni B areas are typically seen in schwannoma of the GIT.[8] Peritumoral lymphoid cuff is not present in soft tissue and central nervous system schwannomas.[13]

A leiomyoma is typically a well circumscribed lesion arising in the muscularis layer and are composed of interlacing fascicles of benign looking smooth muscle bundles with low mitotic activity.[20,21] Leiomyosarcoma, the malignant counterpart, display infiltrative pattern, cellular atypia, increased mitotic activity and may be associated with necrosis. Histologically, inflammatory myofibroblastic tumour shows proliferation of cells with spindle/ epithelioid morphology in a variable background of myxoid and hyalinized stroma. Lymphoplasmacytic infiltrates are also evident within the tumour.[26] Immunohistochemical examination of the tumour cells is considered the optimal diagnostic tool for this type of tumour. It plays an important role for the differential diagnosis between the various types of intramural tumours such as GIST, leiomyoma, leiomyosarcoma and inflammatory myofibroblastic tumour.

Gastric schwannomas are typically positive for S100 protein, SOX10 and GFAP but they can be distinguished from GISTs since they are negative for CD117 and DOG-1, and also to CD34.[9,10,19] GIT schwannomas are more often GFAP positive compared to peripheral soft tissue schwannomas. GISTs frequently expressed CD117 and DOG-1. DOG-1 has been reported to be more sensitive and is positive in 90% of CD117 negative GISTs.[25] Schwannomas are also negative for smooth muscle markers SMA, in contrast to leiomyomas. Leoimyosarcoma share similar antigenic properties as leiomyomas. These smooth muscle neoplasms are positive for SMA, desmin and h-caldesmon.[22] On the other hand, inflammatory myofibroblastic tumours (IMT) are found more commonly in young adults. The tumour cells in IMT are often positive for ALK with variable positivity for SMA but are negative for CD117, DOG-1.[11,12]

Mutational analysis is another helpful investigation that can be used to differentiate schwannomas from GIST. GIST is known to have specific mutations in the c-KIT and PDGFRA genes while schwannomas do not.[23] Gastric schwannomas rarely exhibit somatic NF2 mutations. They are more commonly encountered in soft tissue schwannomas.[24] The aetiology of gastric leiomyomas and leiomyosarcomas have not be ascertained.

In conclusion, gastric schwannoma is a rare gastric neoplasm with good prognosis. Histopathological examination of the tumour with immunohistochemical studies is paramount for accurate diagnosis. Complete resection of the tumour is considered the best treatment of choice as recurrence and metastasis are considered rare events given the benign nature of the tumour.

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revision of manuscript critically for important intellectual content (RJ), revision of manuscript critically for important intellectual content & final approval of manuscript (SHMP).

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REFERENCES

1. Nishida T, Hirota S. Biological and clinical review of stromal tumors in the gastrointestinal tract. *Histol Histopathol*. 2000; 15(4): 1293–301.
2. Miettinen M, Sobin LH, Lasota J. Gastrointestinal stromal tumors of the stomach: a clinicopathologic, immunohistochemical, and molecular genetic study of 1765 cases with long-term follow-up. Am J Surg Pathol. 2005; 29: 52-68.
3. Melvin WS, Wilkinson MG. Gastric schwannoma: clinical and pathologic considerations. Am Surg. 1993; 59: 293-6.
4. Miettinen M, Sarlomo-Rikala M, Lasota J. Gastrointestinal stromal tumours. Ann Chir Gynaecol. 1998; 87: 278–81.
5. Hou YY, Tan YS, Xu JF, *et al*. Schwannoma of the gastrointestinal tract: a clinicopathological, immunohistochemical and ultrastructural study of 33 cases. Histopathology. 2006; 48:536–45.
6. Levy AD, Quiles AM, Miettinen M, *et al*. Gastrointestinal schwannomas: CT features with clinicopathologic correlation. Am. J. Roentgenol. 2005; 184: 797–802.
7. Karabulut N, Martin DR, Yang M. Gastric schwannoma: MRI findings. Br J Radiol. 2002; 75: 624–6.
8. Prévot S, Bienvenu L, Vaillant JC, *et al*. 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.
9. Miettinen M, Lasota J. Gastrointestinal stromal tumors--definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. Virchows Arch. 2001; 438: 1–2.
10. Sarlomo-Rikala M, Kovatich AJ, Barusevicius A, *et al*. CD117: a sensitive marker for gastrointestinal stromal tumors that is more specific than CD34. Mod. Pathol. 1998; 11: 728– 34.
11. Lorenzi L, Cigognetti M, Medicina D, *et al*. ALK-positive inflammatory myofibroblastic tumor of the abdomen with widespread microscopic multifocality. Int J Surg Pathol. 2014; 22(7): 640-4.
12. Kataoka TR, Yamashita N, Furuhata A, *et al*. An inflammatory myofibroblastic tumor exhibiting immunoreactivity to KIT: a case report focusing on a diagnostic pitfall. World J Surg Oncol. 2014; 12: 186.
13. Bae H, Van Vrancken M, Kang TW, *et al*. Peritumoral lymphoid cuff correlates well with lymph node enlargement in gastrointestinal schwannomas. Oncotarget. 2018; 9(16):12591-8.
14. Shah AS, Rathi PM, Somani VS, *et al*. Gastric Schwannoma: A Benign Tumor Often Misdiagnosed as Gastrointestinal Stromal Tumor. Clin Pract. 2015; 5(3): 775.
15. Voltaggio L, Murra R, Lasota J, *et al*. Gastric schwannoma: a clinicopathological study of 51 cases and critical review of the literature. Hum Pathol. 2012; 43: 650–9.
16. Stift A, Friedl J, Gnant M, *et al*. Gastrointestinal autonomic nerve tumors: a surgical point of view. World J Gastroenterol. 2004; 10: 2447–51.
17. Hong HS, Ha HK, Wona HJ, *et al*. Gastric schwannomas: radiological features with endoscopic and pathological correlation. Clin Radiol. 2008; 63: 536-42.
18. Daimaru Y, Kido H, Hashimoto H, *et al*. Benign schwannoma of the gastrointestinal tract: a clinicopathologic and immunohistochemical study. Hum Pathol. 1988; 19: 257–64.
19. Agaimy A, Markl B, Kitz J, *et al*. Peripheral nerve sheath tumors of the gastrointestinal tract: a multicenter study of 58 patients including NF1-associated gastric schwannoma and unusual morphological variants. Virchows Arch. 2010; 456: 411-22.
20. Agaimy A, Wünsch PH. True smooth muscle neoplasms of the gastrointestinal tract: Morphological spectrum and classification in a series of 85 cases from a single institute. Langenbecks Arch Surg. 2007; 392: 75–81.
21. Miettinen M, Sobin LH, Sarlomo-Rikala M. Immunohistochemical spectrum of GISTs at different sites and their differential diagnosis with a reference to CD117 (KIT). Mod Pathol. 2000; 13:1134–42.
22. Fletcher CDM, Bridge JA, Hogendoom PCW. WHO classification of tumours of soft tissue and bone. Lyon: IARC Press. 2013; 111-3.
23. Lasota J, Miettinen M. Clinical significance of oncogenic KIT and PDGFRA mutations in gastrointestinal stromal tumours. Histopathology. 2008; 53: 245–66.
24. Lasota J, Wasag B, Dansonka-Mieszkowska A, *et al*. Evaluation of NF2 and NF1 tumor suppressor genes in distinctive gastrointestinal nerve sheath tumors traditionally diagnosed as benign schwannomas: a study of 20 cases. Lab Invest. 2003; 83: 1361-71.
25. Kang GH, Srivastava A, Kim YE, *et al*. DOG1 and PKC-θ are useful in the diagnosis of KIT negative gastrointestinal stromal tumors. Mod Pathol. 2011; 24: 866–75.
26. Coffin CM, Hornick JL, Fletcher CD. Inflammatory myofibroblastic tumor: comparison of clinicopathologic, histologic, and immunohistochemical features including ALK expression in atypical and aggressive cases. Am J Surg Pathol. 2007; 31(4): 509–20.