



Case Report

Vagal Nerve Schwannoma Clinically Mimicking Gastrointestinal Stromal Tumor: Report of a Case

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Abstract

Introduction: Mesenchymal tumors of the gastrointestinal (GI) tract are mainly comprised of a spectrum of spindle cell tumors which include gastrointestinal stromal tumors (GISTs), leiomyomas or leiomyosarcomas, and schwannomas. As all of these tumors of the GI are located in the submucosal layer of the bowel wall, differential diagnosis is very difficult. Histopathological evaluation using immunohistochemical staining is required for the definitive diagnosis.

Presentation of case: A 20-year-old female patient was presented with an upper abdominal pain and dyspepsia since 2 months duration. This case is initially thought as GIST but finally diagnosed as vagal nerve schwannoma by histopathological evaluation after resection of the mass.

Conclusion: Schwannomas should be considered in the differential diagnosis of other mesenchymal tumors, such as gastrointestinal stromal tumors and leiomyomas or leiomyosarcomas, indeed it can grow in any part of the peripheral nerves along the gastrointestinal tract. Histopathological evaluation including immunohistochemical staining is required for the definitive diagnosis.

Keywords: Schwannoma; Neurilemoma; Mesenchymal tumor; Gastrointestinal stromal tumor

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Consent: We confirm that the patient has given their informed consent for the case report to be published.

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Introduction

Schwannomas (also known as neurilemmomas or neuromas) originate from schwann cells, which normally wrap around the axons of the peripheral nerves. Theoretically, schwannomas can develop anywhere along the peripheral course of the nerves. Although schwannomas are usually found in the head and neck region, they also have rarely been reported in the gastrointestinal tract in which stomach and small intestine are the most involved sites [1]. We report a case clinically and radiologically diagnosed as gastrointestinal stromal tumor due to submucosal gastric localization but finally diagnosed as vagal nerve schwannoma by histopathological evaluation after the resection of the mass. Vagal nerve schwannomas generally originate from the thoracic or cervical compartment of the vagal nerve whereas it originated from the abdominal compartment in the present case.

Case Presentation

A 20-year-old female patient was presented with an upper abdominal pain and dyspepsia for 2 months duration. There was not any notable finding on her medical history and physical examination. Laboratory results revealed a normal complete blood count and biochemical analysis. The serum carbohydrate antigen (CA 19-9) level was < 2.5 U/ml, serum carcino-embryonic antigen (CEA) level was 0.39 ng/ml and serum alpha-fetoprotein (AFP) level was 1.83 IU/ml. A retroperitoneal mass with a 9 cm of diameter reaching to the left medial lobe of the liver and posterior to the pancreas was detected on abdominal ultrasonography. Magnetic resonance imaging (MRI) demonstrated a well-defined mass which was located closely to the left diaphragm and stomach (Figure 1). The patient underwent an esophagogastroduodenoscopy (EGD) and no intraluminal lesion was detected in the stomach.

Ultrasonography-guided fine needle aspiration cytology was performed and the cytopathological examination was reported as spindle cell neoplasm. After presenting the case at the Multidisciplinary Oncology Board Conference, we decided to perform laparotomy for resection of the mass. A mercedes-typed incision was performed. The stomach was mobilized by opening the gastrocolic ligament. We observed the mass which was originated from the left diaphragmatic crus and reaching to the spleen laterally and to the left medial lobe of the liver medially (Figure 2). The mass was en-bloc resected followed by bilateral truncal vagotomy and the diaphragmatic cruses were repaired with primary sutures. A V-shaped polypropylene mesh was placed over the cruses. Mean operative time was 150 minutes and mean blood loss was 200 ml. The patient had an uneventful recovery. Macroscopic examination of the encapsulated mass measuring 9.5x6x4.5 cm diameters, showed a tan-white colored solid lobulated cut surface with degenerative myxoid areas. Histopathologic examination revealed that the neoplastic mass was comprised of spindle cells (Figure 3). The neoplastic spindle cells showed diffuse immuno-reactivity with S-100 protein but lacked immunoreactivity with CD 117, CD 34, smooth-muscle actin (SMA), HMB45 and desmin (Figure 4). There were no necrosis and the mitotic activity was 1/50 high-power fields (HPF). Additionally, the ki67 proliferative index was 3-5%. The histopathologic and immunohistochemical features were consistent with schwannoma. To rule out vestibular schwannoma or metastatic primary origin, cranial MRI and temporal bone MRI were performed which did not reveal any focus. According to our oncology tumor board decision, the patient is followed up without adjuvant chemotherapy. At the end of 3 months follow-up an abdominal CT is performed, which showed no loco-regional recurrence.

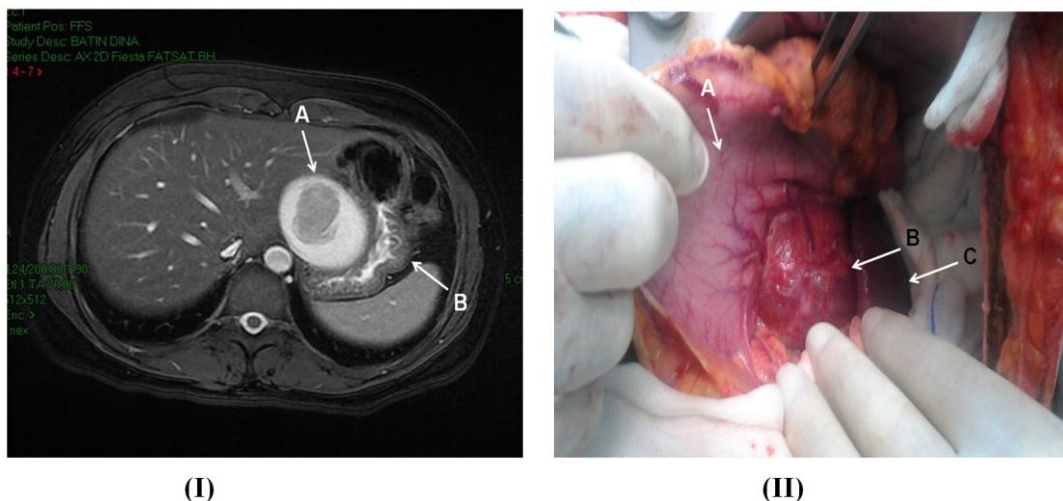


Figure 1 I. Magnetic Resonance Image (A- The vagal schwannoma, B- The stomach is pushed laterally by the tumoral mass) and **II.** Intraoperative image of the schwannoma (A- The stomach, B- The vagal schwannoma, C- The spleen).

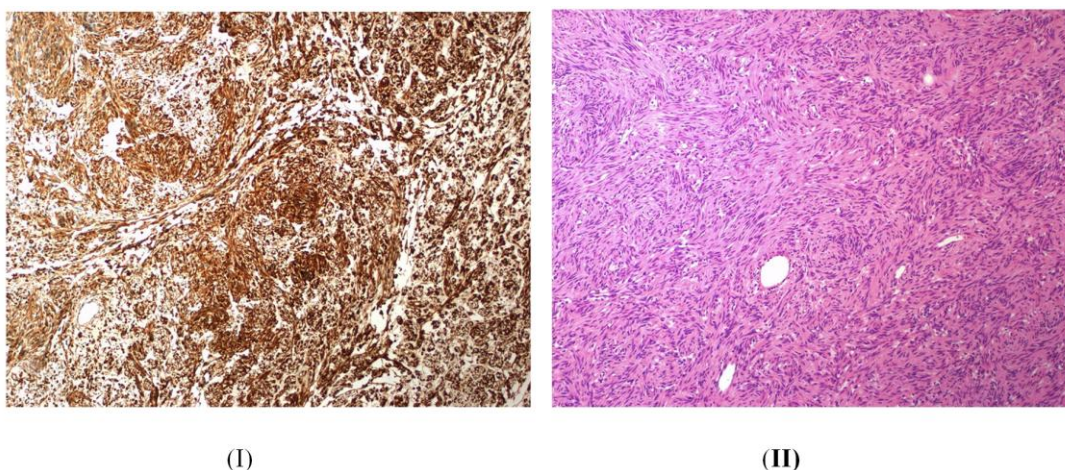


Figure 2 I. Cellular tumor area composed of spindle cells and thin-walled vascular channels (HE, x10) and **II.** Diffuse S-100 positivity in tumor cells (DAB, x10)

Discussion

Schwannomas are commonly benign, encapsulated, solitary and well circumscribed tumors. Schwannomas can develop anywhere along the peripheral nerve. They usually grow slowly and do not invade adjacent organs. Owing to subclinical tumor growth, the diagnosis is usually delayed. Schwannomas can occur at any age but are generally diagnosed between the second and fifth decades of life. There is no gender predilection. Patients are usually asymptomatic and laboratory tests are non-specific. Preoperative differentiation of

schwannoma from other submucosal tumors is very difficult and investigations such as fine needle aspiration cytology (FNAC) have low specificity [2, 3]. On contrast enhanced computed tomography (CT), necrotic areas and cystic degeneration are the most characteristic findings. Magnetic Resonance Imaging (MRI) evaluation typically shows the mass of intermediate signal on T1-weighted images and increased signal intensity on T2-weighted images with smooth, well-delineated margins and a homogeneous overall appearance. The 2-[18F]-fluoro-2-deoxy-D-glucose positron emission tomography with CT (FDG PET/CT) has

a low specificity because schwannomas have a high affinity to FDG, and it is unreliable in differentiating schwannomas from malignant peripheral nerve sheath tumors [4, 5].

Histologic examination mostly show a hypercellular portion of spindle cells with nuclear palisading (Antoni A) and a hypocellular portion of cells with cystic degeneration, focal calcification, and hemangiomatic vascular changes (Antoni B). Presence of S-100 protein and absence of smooth muscle antigens sustain a nerve sheath origin of the tumor. A malignant transformation may rarely occur. The definitive diagnosis of the malignant schwannoma can be a challenge and it needs an absolute histological examination. Malignancy depends on the presence of frequent or atypical mitotic figures, hypercellularity, tumor necrosis and nuclear atypia. The most reliable factor that correlates with malignancy is mitotic rate of 5 or more in 50 HPF [1, 6].

Mesenchymal tumors of the gastrointestinal (GI) tract are mainly comprised of a spectrum of spindle cell tumors which include gastrointestinal stromal tumors (GISTs), leiomyomas or leiomyosarcomas, and schwannomas. GISTs are the most common mesenchymal tumors of the gastrointestinal tract, and 60–70% of them occur in the stomach. GISTs and schwannomas seem grossly similar. However, the prognosis for schwannomas and GISTs is very different. Although schwannomas are usually in benign character, GISTs have malignant behavior. Schwannomas can vary from leiomyomas by SMA and desmin negativity, from GIST by CD34 and CD117 negativity and perivascular epithelioid cell tumor (PEComa) by HMB45 negativity [7-9].

The main treatment is complete surgical excision with clear margins. Prognosis is generally excellent and they have a low recurrence rate. Recurrent disease has been only observed after incomplete resection [10].

Because of the possibility of development anywhere along the peripheral course of nerves, schwannomas have been reported in various localizations of the body. Although, head and neck regions are the most involved ones, there are some case reports of schwannomas that were found in

the gastrointestinal tract, retroperitoneum, and diaphragm, respectively [11-13]. To our knowledge -after research of the English written literature- this is the first case who presented with vagal nerve schwannoma involving the diaphragmatic crus in the abdominal compartment.

Conclusion

Schwannomas should be considered in the differential diagnosis of other mesenchymal tumors, such as gastrointestinal stromal tumors and leiomyomas or leiomyosarcomas, indeed it can grow in any part of the peripheral nerves along the gastrointestinal tract. Histopathological evaluation including immunohistochemical staining is required for the definitive diagnosis.

References

1. Kurtkaya-Yapici O, Scheithauer B, Woodruff JM. The pathobiologic spectrum of Schwannomas. *Histol Histopathol.* 2003, 18: 925-934
2. Prévot S, Bienvenu L, Vaillant JC, de 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: 431-436
3. M. R. Sreevatsa, Ranganath Vitlapur Srinivasarao. Three Cases of Vagal Nerve Schwannoma and Review of Literature. *Indian J Otolaryngol Head Neck Surg.* 2011, 63: 310-312
4. M. H. Raber, C. M. P. Z. des Plantes, R. Vink, R. Vink, J. M. Klaase. Gastric schwannoma presenting as an incidentaloma on CT-scan and MRI. *Gastroenterology Research.* 2010, 3: 276-280
5. Benz MR, Czernin J, Dry SM, Tap WD, Allen-Auerbach MS, Elashoff D, et al. Quantitative F18-fluorodeoxyglucose positron emission tomography accurately characterizes peripheral nerve sheath tumors as malignant or benign. *Cancer.* 2010, 116: 451-458
6. 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: 650-659

7. Fülöp E, Marcu S, Borda A, Moldovan C, Fülöp EF, Loghin A, et al. Histopathological and immunohistochemical features of gastrointestinal stromal tumors. *Rom J Morphol Embryol*. 2011, 52:555-562
8. Boşoteanu M, Boşoteanu C, Deacu M, Aşchie M. Differential diagnosis of a gastric stromal tumor: case report and literature review. *Rom J Morphol Embryol*. 2011, 52: 1361-1368
9. William Yoon, Kari Paulson, Paul Mazzara, Sweetie Nagori, Mohammed Barawi, Richard Berri. Gastric Schwannoma: A Rare but Important Differential Diagnosis of a Gastric Submucosal Mass. *Case Rep Surg*. 2012, 2012:280982
10. Theodosopoulos T, Stafyla VK, Tsiantoula P, Yiallourou A, Marinis A, Kondi-Pafitis A, et al. Special problems encountering surgical management of large retroperitoneal schwannomas. *World J Surg Oncol*, 2008, 6:107
11. Langner E, Del Negro A, Akashi HK, Araújo PP, Tincani AJ, Martins AS. Schwannomas in the head and neck: retrospective analysis of 21 patients and review of the literature. *Sao Paulo Med J*. 2007, 125:220-222
12. Koichi Oishi, Toshiyuki Itamoto, Sho Okimoto, Takashi Nishisaka. Multiple Retroperitoneal Schwannomas Mimicking Adrenal Tumors. *Case Rep Oncol*. 2012. 5: 222-228
13. David J. Hobbs, Jennifer McLellan, Marc G. Schlatter. Schwannoma of the diaphragm- a pediatric case report and review of the literature. *J Ped Surg*. 2012, 47: E5-E7