Gastric Schwannoma mimicking malignant gastrointestinal stromal tumor and misdiagnosed by $^{18}$F-FDG PET/CT

Abstract

Gastric Schwannoma is a rare benign mesenchymal tumor that accounts for only 0.2% of all gastric tumors. The current study presents a case of gastric Schwannoma misdiagnosed as malignant gastrointestinal stromal tumor (GIST) by esophagogastroduodenoscopy, endoscopic ultrasonography, and contrast-enhanced or not enhanced and $^{18}$F-FDG PET/CT imaging.

Introduction

Schwannomas, also known as neurilemomas, are tumors originating from nerve cells that have Schwann cell sheaths. Gastrointestinal Schwannomas are rare, approximately 50 times less common than the most common gastrointestinal stromal tumor (GIST). It often occurs in older adults and forms intramural masses of 2-10cm that can ulcerate and present in a manner similar to GIST, gross features included [1, 2].

Case report

A 69 years old woman presented for evaluation of a 1 year history of nonspecific epigastric abdominal pain. Esophagogastroduodenoscopy (Figure 1A) and endoscopic ultrasonography (Figure 1B) revealed smooth normal gastric mucosa overlying a 4.0×3.2cm subepithelial (intramural) lesion located at the greater curvature of the antrum, suggestive of a GIST. Contrast-enhanced computed tomography (CT) scan of the abdomen revealed a solitary, exophytic soft tissue mass in the antrum (Figure 2A), and pathological lymph nodes measured 1.9×1.3cm adjacent to the lesion (Figure 2B). In addition, fluorine-18-fluorodeoxyglucose ($^{18}$F-FDG) positron emission tomography/CT (PET/CT) scan showed obvious $^{18}$F-FDG accumulation in the gastric tumor with maximum standardized uptake value (SUVmax)=4.5 (Figure 3A and B) and lymphadenopathy (SUVmax 1.3, Figure 3C). These findings were highly suspicious of malignant GIST with metastatic lymphadenopathy.

The patient underwent laparoscopic subtotal gastrectomy and regional lymph node dissection. Macroscopic examination of the stomach revealed a grayish-white, solid and well-circumscribed tumor 5×4cm with a rubbery and jelly cut surface located at the lesser curvature close to pylorus with exophytic character, beginning from the tunica muscularis without infiltrating the mucosa. Microscopically, the tumor tissue was spindle shaped, showing a fascicular or palisade arrangement (Figure 4A). Lymphocytic cuffing was identified at the periphery of the tumor (Figure 4B). Immunohistochemically, the tumor was positive for S-100 protein (Figure 4C), neuronal specific enolase (NSE) (Figure 4D) and glial fibrillary acidic protein (GFAP) (Figure 4E), but negative for CD 117, CD 34, smooth muscle actin (SMA) and desmin. There was no mitosis, and the Ki 67 index was only 1% (Figure 4F). The histopathologic features and immunohistochemical staining pattern were consistent with a benign gastric Schwannoma. Additionally, lymphadenopathy adjacent to the gastric tumor was confirmed to be chronic lymphadenitis (Figure 4G).
Figure 1. Esophagogastroduodenoscopy (A) and endoscopic ultrasound (B) revealed a large submucosal tumor measuring 4.0×3.2cm in the greater curvature of the gastric antrum.

Figure 2. Contrast-enhanced CT showed a well-defined, inhomogeneous, enhanced gastric mass (A) and pathological lymph nodes (B, arrow) adjacent to the lesion.

Figure 3. Maximum intensity projection images showed an intense hypermetabolic focus in the stomach (A, arrow). Transaxial views of the antral mass were observed with an overlay of CT, PET, and PET/CT images with a SUVmax of 4.5 (B). An 18F-FDG PET/CT scan also showed 18F-FDG accumulation in the lymphadenopathy with a SUVmax of 1.3 (C).

Discussion

The differentiation of Schwannoma from GIST by medical imaging procedures is difficult [3-5]. Fluorine-18-FDG PET/CT is accepted as a powerful and noninvasive metabolic/anatomic imaging modality for identifying and evaluating various tumors for distinguishing their malignant potential preoperatively, for staging and monitoring the therapeutic response [6, 7]. The 18F-FDG uptake has been demonstrated to have a significant correlation with the malignant potential of gastric GIST [8, 9]. Although gastric Schwannomas are normally benign, and their malignant transformation is extremely rare [10, 11], this tumor is considered as with high uptake of 18F-FDG [12, 13]. It remains unclear why high 18F-FDG uptake is found in benign tumors such as neural Schwannoma. This could possibly be due to over-expression of the glucose transporter by the tumor cells. In particular, glucose transporter type 3 is found in all human tissues and is the major glucose transporter on the neuronal surface [5, 12, 14].

Figure 4. (A-H) Histopathological findings. (A): The resected neoplastic mass comprised of spindle cells showing a fascicular or palisade arrangement (H&E, ×100). (B): Cuff-like lymphoid aggregate was recognized around the tumor (arrow, H&E, ×50). (C): S100 protein-positive stain (×50). (D): Neuron specific enolase (NSE)-positive stain (×50). (E): Gliarial fibrillary acid protein (GFAP)-positive stain (×50). (F): No mitosis was observed and the Ki67 index was only 1% (×200). (G): The lymphadenopathy adjacent to the gastric tumor was confirmed to be chronic lymphadenitis (H&E, ×25).

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Diagnosis is usually made only after excision and histologic examination owing to nonspecific radiologic and endoscopic appearances. The cellular structures of Schwannomas and GIST are spindle-shaped and appear similar under light microscopic examinations. However, there are differences in the immunohistochemical staining between these spindle cell tumors. Positive CD34 and CD117 indicate GIST, and positive S-100 indicates Schwannomas [1, 16]. The tumors in the present case, revealed spindle cells, strongly positive for S-100 staining, which thus indicated the diagnosis of Schwannomas.

Although benign, definitive treatment of gastric Schwannomas in fit patients entails surgical excision with clear margins, as it is frequently impossible to distinguish these tumors from other mesenchymal tumors, which are malignant or have malignant potential [17].

The authors declare that they have no conflicts of interest.

Bibliography

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