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Case Report

Gastric schwannoma: Two case reports and review of the literature ☆

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ABSTRACT

Gastric schwannomas are rare, slow-growing tumors whose clinical presentation is nonspecific. These are mostly benign, with a low probability of malignant transformation and an excellent prognosis. We present 2 cases of gastric schwannomas with distinct clinical features and imaging patterns, whose therapeutic approach differed. Case 1 is a 73-year-old woman with a voluminous subepithelial lesion in the greater gastric curvature, with predominantly endoluminal growth. Clinically the patient presented with nonspecific abdominal complaints and underwent complete surgical excision. Case 2 is a 69-year-old woman with an exophytic lesion adjacent to the gastric antrum, diagnosed incidentally and managed conservatively, with imaging follow-up, for the last 5 years and stable ever since. This article aims to focus on this rare disease, illustrating its main imaging findings, particularly in magnetic resonance imaging, along with pathological correlation, as well as reviewing the literature, discussing the differential diagnosis, and exploring clinical management and prognosis.

Introduction

Gastrointestinal schwannomas are uncommon tumors that account for 2%-6% of all gastrointestinal mesenchymal tumors [1,2]. They were first described in 1988 by Daimaru et al. [3] and are most commonly found in the stomach (60%-70% of cases), followed by the colon and rectum.

Gastric schwannomas (GS) are even rarer, accounting for only 0.2% of all gastric tumors. They consist of excessive pro-

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liferation of Schwann cells from the nerve bundles of Auerbach's plexus or, less commonly, Meissner's plexus [4,5].

Given their nonspecific clinical presentation, GS are usually identified incidentally. However, the absence of specific features on imaging makes their diagnosis difficult before histological confirmation. Pathological and immunohistochemical evaluation is fundamental in establishing the final diagnosis, differentiating gastric schwannomas from other stromal tumors [6].

We present 2 patients with histological evidence of GS and emphasize the magnetic resonance imaging (MRI) findings of these tumors. In addition, we review the topic, including its clinical presentation, imaging and endoscopic features, clinical management, and prognosis.

Case report

Case 1

A 73-year-old woman presented to our institution for abdominal discomfort, nausea, and early satiety. She had a past medical history of multinodular goiter, hyperparathyroidism, dyslipidemia, and allergy to iodinated contrast. There was also a family history of gastric adenocarcinoma (father). Physical examination and blood tests were unremarkable.

Upper endoscopy revealed a large bilobed subepithelial lesion on the posterior wall of the gastric body/antrum (Fig. 1A). Complementary endoscopic ultrasound (EUS) showed a hypoechoic, heterogeneous, well-defined lesion dependent on the muscularis propria, with 56 mm in greatest diameter (Fig. 1B). An endoscopic ultrasound-guided fine-needle biopsy (EUS-FNB) was performed (Fig. 1C).

As the patient was allergic to iodinated contrast, she underwent an abdominal MRI, which showed a lesion with 51 mm x 29 mm in the greater gastric curvature hypointense on T2-weighted images (WI), although with some areas of intermediate signal intensity, low ADC on diffusion-weighted imaging (DWI), and progressive enhancement after intravenous gadolinium-based contrast agent administration. It showed predominantly endoluminal growth, but there was a slight external protrusion of the gastric wall (Fig. 2). No signs of disease spreading or large size lymph nodes were detected.

The histology of the specimens provided by EUS-FNB revealed a mesenchymal lesion with morphology and immunohistochemical profile compatible with schwannoma: diffuse and intense nuclear and cytoplasmic expression for S-100 and no expression for CD34, CD117 (c-kit), DOG1, or smooth muscle actin (SMA) with a Ki67 index of 1% (Fig. 3).

The patient underwent atypical gastric resection, and the pathological study of the surgical specimen confirmed the diagnosis of schwannoma. The tumor was centered in the wall, well delimited, with a yellowish aspect, fasciculated and elastic, without necrosis (Figs. 4 and 5). The serosa was intact, and the surgical margins were negative (R0 resection). The patient has been on follow-up and disease-free for one year.

Case 2

In a 69-year-old female who underwent an abdominal MRI to characterize complex hepatic cysts, an exophytic nodule with rounded morphology adjacent to the gastric antrum with 24 × 22 mm was incidentally identified. The lesion had intermediate signal intensity in T2-WI, showing a well-defined hypointense wall, hypointense on fat saturation T1-WI, hyperintense on DWI, but no restriction on the ADC map, and progressively enhanced, more pronounced in the delayed phase (Fig. 6).

Physical examination and blood tests, including tumor markers, were unremarkable.

^{18}F-fluoro-deoxy-glucose positron emission tomography-computed tomography (^{18}F-FDG PET/CT) revealed the lesion was metabolically active with avid FDG uptake, reaching a maximum standardized uptake value (SUVmax) of 7.7 (Fig. 7).

The EUS showed a well-defined homogeneous hypoechogenic lesion originating from the muscularis propria (Fig. 8), which underwent EUS-FNB. The histology study showed a mesenchymal tumor whose immunohistochemical profile was compatible with schwannoma (Fig. 9). After explaining the diagnosis to the patient, she chose not to excise the lesion and has been under imaging surveillance for 5 years, with dimensional stability of the lesion so far.

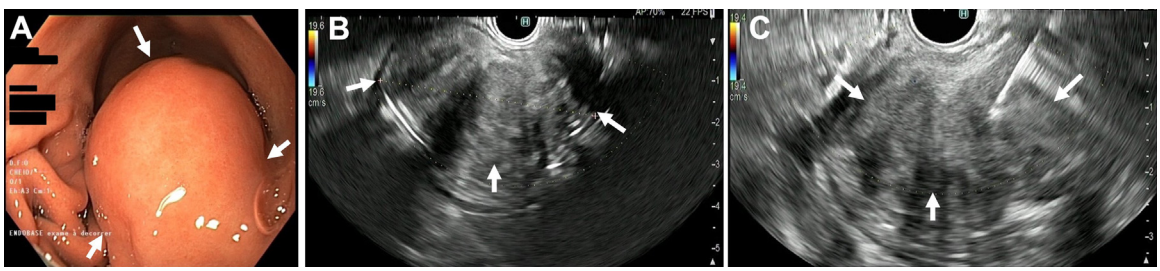

Fig. 1 – Endoscopic ultrasound case 1. Upper digestive endoscopy (A) showing a large bilobed subepithelial lesion on the posterior wall of the body/gastric antrum (arrows). Endoscopic ultrasound shows the lesion is hypoechoic, heterogeneous, well-defined, and dependent on the muscularis propria, measuring 56 mm in diameter (B). The lesion underwent an EUS-FNB (C).

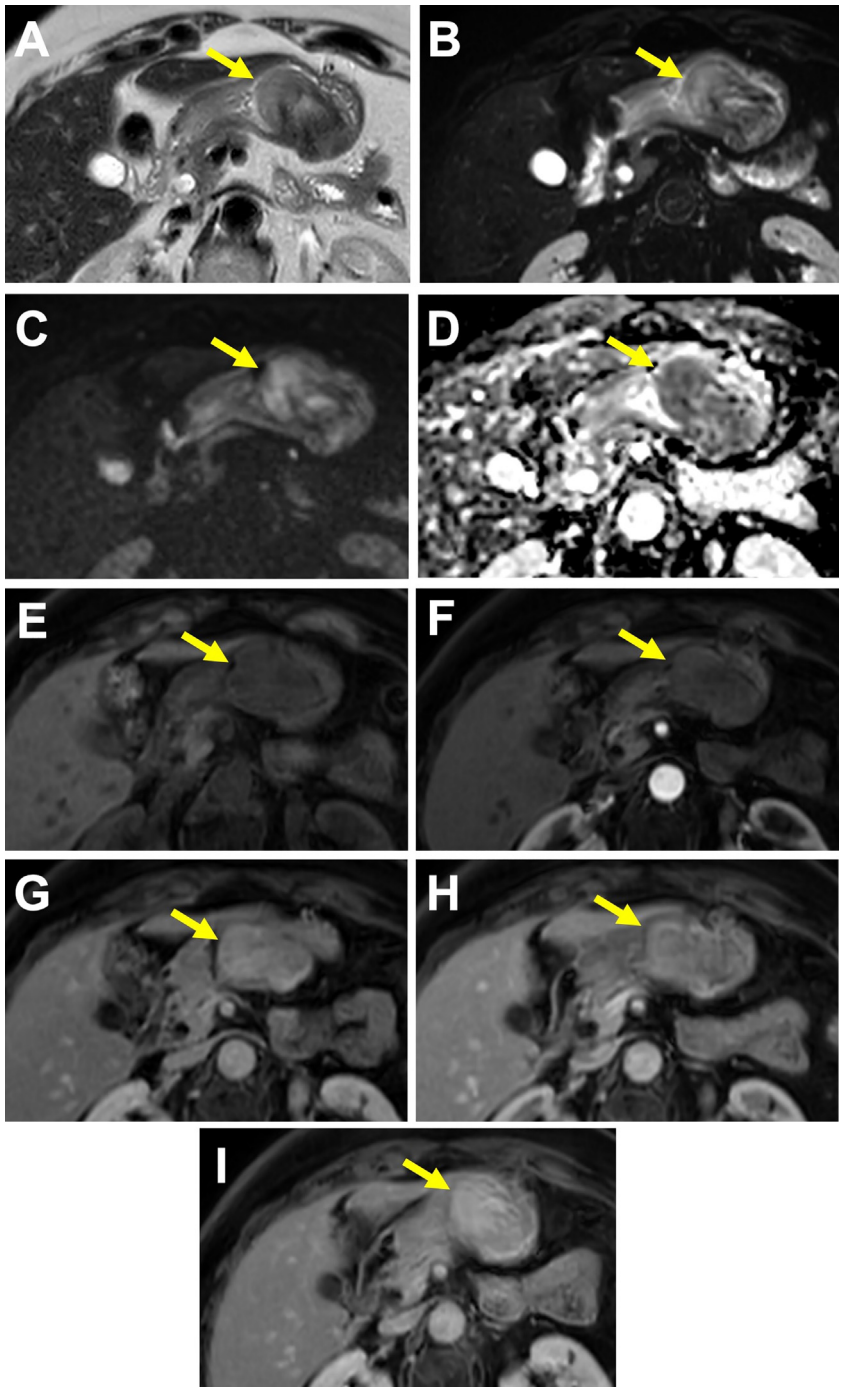

Fig. 2 – Abdominal MRI case 1. MRI shows the lesion in the large gastric curvature, predominantly endoluminal growth but with slight external protrusion of the gastric wall (arrows). It is hypointense on T2-WI (A) and DWI b10 (B), although with some areas of intermediate signal intensity. Show restricted diffusion with high signal intensity in DWI b900 (C) and low values on ADC map (D). It is homogeneously hypointense on FS T1-WI (E) with progressive enhancement after intravenous gadolinium-based contrast agent administration. The enhancement in the arterial phase is minimal (F), with progressive enhancement in the portal venous (G), interstitial (H), and delayed (I) phases.

Discussion

GS usually arise as solitary lesions in the submucosa or muscularis propria of the gastric wall and are most frequent in the stomach body (50%), followed by the antrum (32%) and fundus (18%) [5]. They may have an endophytic or exophytic growth pattern or a combination of both [6,7]. They most commonly appear in women between the fifth and sixth decades of life. Their slow growth means that they are usually asymptomatic

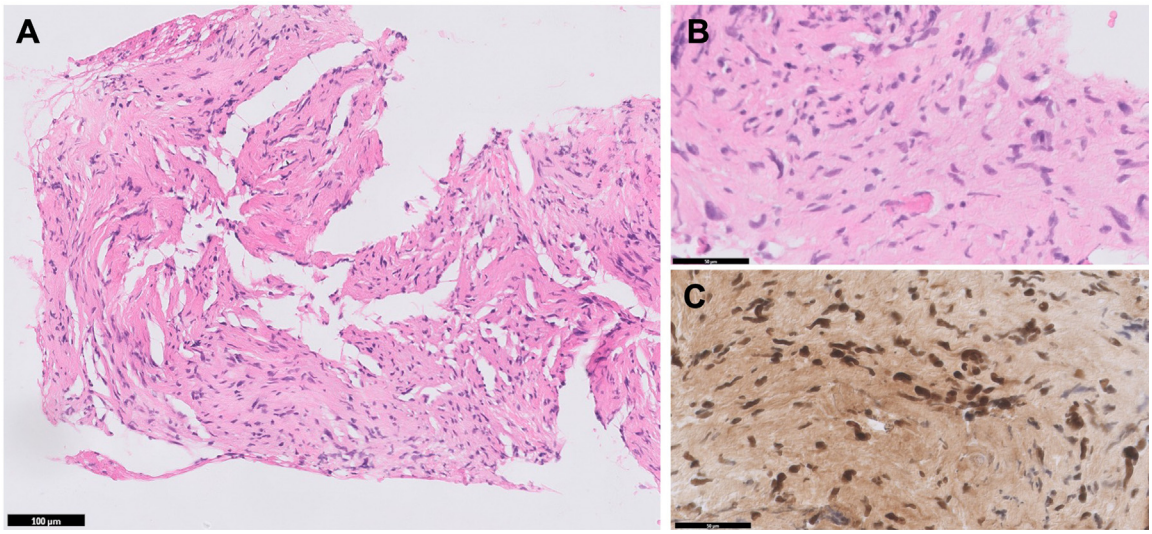

Fig. 3 – Biopsy histology case 1. (A) Histologically, the biopsy was composed of a proliferation of fusiform cells, with mild nuclear atypia, quite cellular and focally forming bundles (in B. at higher magnification); (C) the neoplastic cells showed intense and diffuse nuclear and cytoplasmic expression of S-100.

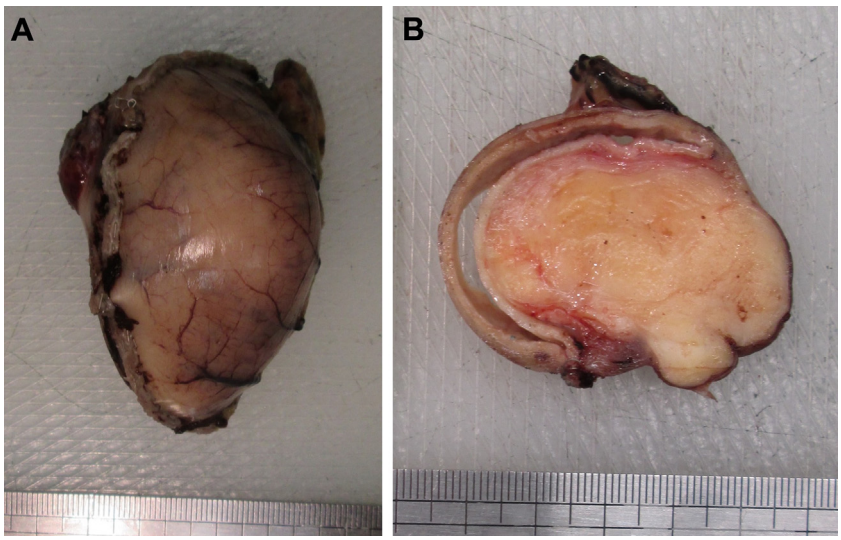

Fig. 4 – Surgical specimen. (A) Atypical gastric resection surgical specimen. (B) Cross-section reveals a 55 mm submucosal and well circumscribed lesion with white fasciculated and shiny surface.

and identified incidentally [4,6,8]. When symptomatic, most patients present with abdominal pain, gastrointestinal bleeding, palpable masses, and, less often, gastrointestinal obstruction [6].

On computed tomography (CT), they are well-defined lesions, rounded or lobulated, with low attenuation on unenhanced images due to their dense spindle cell composition [2]. Multiphase contrast-enhanced CT study shows mild enhancement in the arterial and venous phases with moderate enhancement in the late phase. They rarely have cystic degeneration or calcifications. The main differential diagnosis of GS is gastrointestinal stromal tumors (GIST), the most common mesenchymal neoplasm of the stomach [2,7,9]. It is estimated that there are approximately 45 gastric GISTs for each GS [6,10,11]. The most notable difference between the 2 entities is the greater heterogeneity of GIST due to their usually present hemorrhage, necrosis, and cavitation [2,9]. Moreover, according to Ji et al. [9], GS's progressively homogeneous enhancement pattern may help differentiate them from GISTs, which often have a more heterogeneous enhancement in the arterial phase and decreases in subsequent phases. Although the 2 cases presented the patients had not undergone CT, the enhancement pattern assessed on MRI agrees with that described in the literature.

There are few published data on the characteristics of GS on MRI, with some cases described in which the tumors

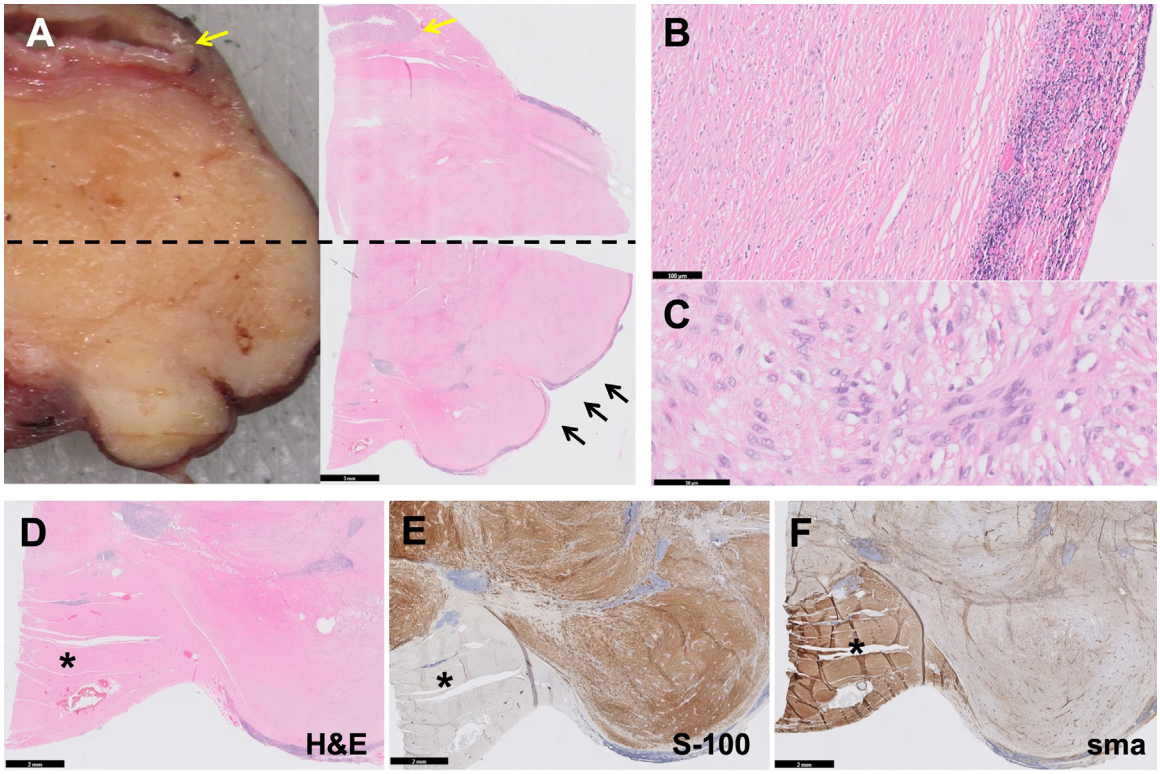

Fig. 5 – Histology case 1. (A) Histologically, an homogeneous proliferation was observed under the gastric mucosa (yellow arrow), buldging under the muscular layer (asterisk); in the subserosa at the periphery of the tumor, a lymphocytic infiltrate was identified (black arrows, in (B) at higher magnification); (C) the neoplasm is composed of fusiform cells without atypia; (D) schwannoma cells show diffuse expression of S-100 (E) and no expression of smooth muscle actin (SMA) (F), which is expressed by the gastric muscular layer cells (asterisks in D-F). Scale bars represent the different sizes described in them.

showed hypointensity on T1WI, hyperintensity on T2WI, and restricted water diffusion (consistent with their high cellularity) [7,12,13].

In case 1, the tumor presented a T2 predominant hypointense signal, an atypical finding not previously described in GS. In case 2, the tumor does not have a true diffusion restriction, a finding that might be explained by the lesion's low cellularity proven histologically.

The features of GS on ^{18}F-FDG PET/CT are also poorly reported, with data suggesting that gastrointestinal schwannomas are prone to higher SUVmax values than schwannomas in other locations [4,12,14]. On the other hand, ^{18}F-FDG PET/MR appears to have a limited role in differentiating between GISTs and other non-GIST subepithelial gastric lesions but may help distinguish between high- and low-risk GISTs [15].

On endoscopy, GS appear as protruding masses, which may be associated with a central ulcer in 25%-50% of cases [4]. EUS allows the location and boundaries of the tumor to be determined, as well as performing EUS-guided biopsy, which is currently considered the standard method for sampling submucosal lesions. In GS, a conventional endoscopic biopsy may produce false-negative results since the mucosa overlying the lesions may be normal or show only chronic inflammatory changes [1,4,13].

The lack of reliably specific findings in the techniques described above makes the definitive diagnosis dependent on pathological findings. On histology, GS show atypical spindle-shaped cells arranged in a trabecular or fascicular pattern, and there may be a prominent lymphoid cuff in the periphery of the tumor, a finding that is characteristic of gastrointestinal schwannomas and uncommon in schwannomas elsewhere [7,13]. On immunohistochemistry, they are positive for S100 protein, a specific marker for schwannomas in general. Negative expression of other markers, such as cKIT, desmin, and smooth muscle actin (SMA), helps differentiate them from other mesenchymal tumors [7,8,13,16].

The therapeutic approach depends on the location, layer of origin, size, and growth pattern of the tumors, as well as their relationship to surrounding structures. Available therapeutic modalities include endoscopic or surgical resection; preoperative imaging evaluation is essential in the decision [10,13,16]. Endoscopic or imaging surveillance has been proposed for small (<2 cm) schwannomas, although it is a poor consensus recommendation and rarely adopted in reported cases [1].

GS are mostly benign tumors whose malignant transformation is rare. Although there are no specific data for GS, in the universe of gastrointestinal schwannomas, malignancy occurs in only 2% of cases. With complete surgical excision, they have an excellent prognosis, and, as far as we know, there are no reported cases of recurrence of benign GS [5,6,8,10,13].

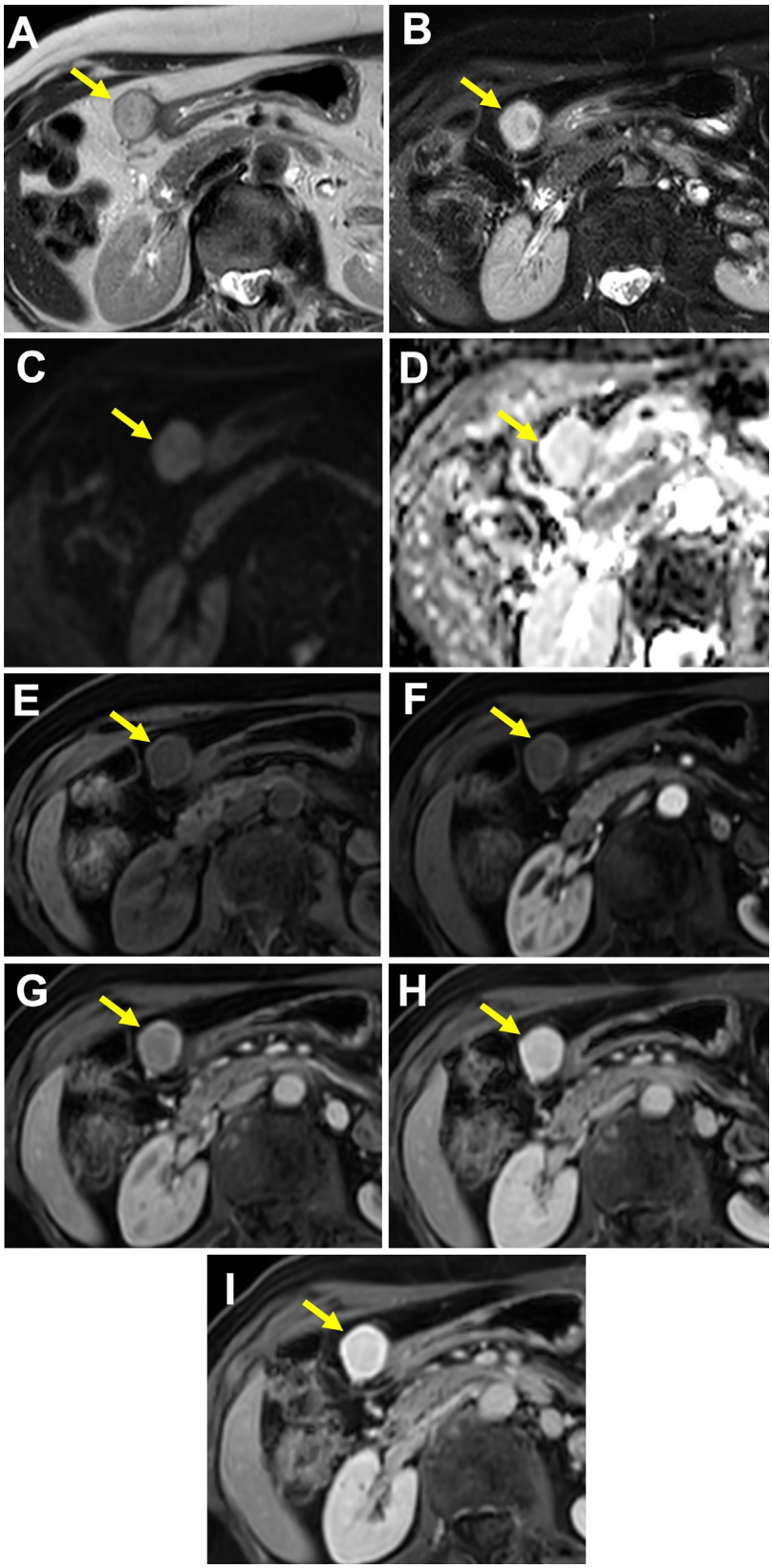

Fig. 6 – Abdominal MRI case 2. MRI shows an exophytic lesion with rounded morphology adjacent to the gastric antrum (arrows). The lesion had an intermediate signal intensity on T2-WI with a well-defined hypointense wall (A), hyperintense on DWI b10 (B), and DWI b900 (C), but no restriction on the ADC map (D). Was hypointense on fat saturation T1-WI (E) with progressively enhanced. The enhancement in the arterial phase is minimal (F), with progressive enhancement in the portal venous (G), interstitial (H), and delayed (I) phases.

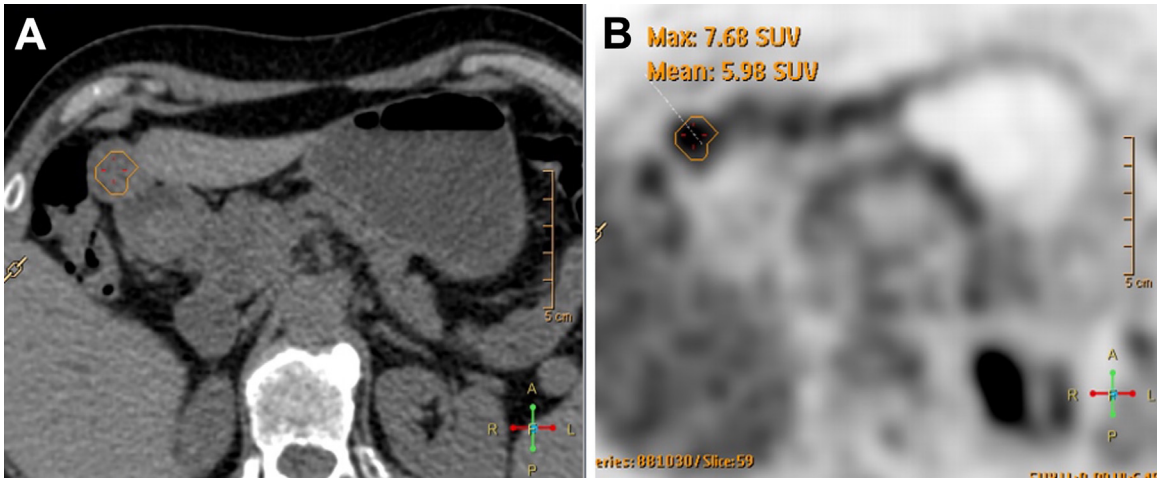

Fig. 7 – ^{18}F-FDG PET/CT case 2. The axial low-dose CT (A) and ^{18}F-FDG PET images (B) show the lesion adjacent to the gastric antrum has avid FDG uptake, reaching a SUVmax of 7.7.

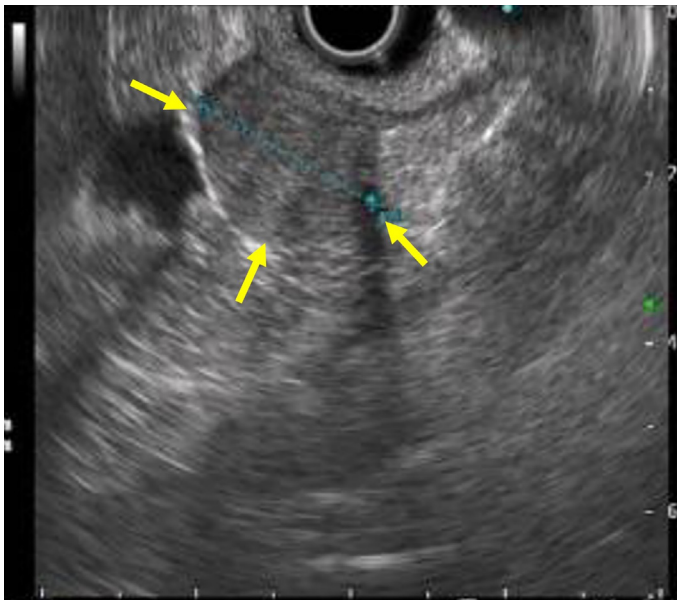

Fig. 8 – Endoscopic ultrasound case 2. Endoscopic ultrasound showed a well-defined homogeneous hypoechogenic lesion originating from the muscularis propria (arrows).

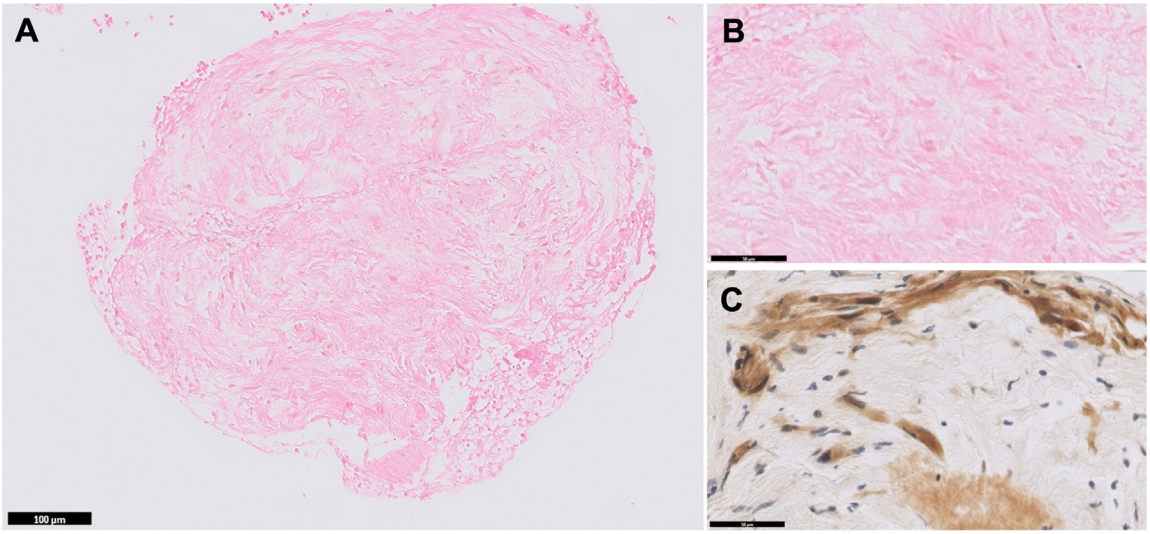

Fig. 9 – Biopsy histology case 2. (A) Histologically, the biopsy was poorly cellular, with fusiform cells with elongated nuclei embedded in a collagenized stroma (in B. at higher magnification); (C) the neoplastic cells showed intense and diffuse nuclear and cytoplasmic expression of S-100.

Conclusion

GS is a rare, slow-growing tumor whose clinical presentation is nonspecific. Imaging, namely CT, MRI, and EUS, helps early detection and defines the therapeutic approach. However, the lack of specific findings in conventional imaging studies makes the preoperative diagnosis of GS quite challenging. We reinforce this difficulty by presenting 2 cases in which the T2 and DWI signal patterns were atypical on MRI compared to the currently available data. The definitive diagnosis is made by pathological examination. The therapeutic approach usually includes endoscopic or surgical resection. These are mostly benign tumors with a low probability of malignant transformation and an excellent prognosis.

Patient consent

The authors obtained written informed consent from the patients to publish this case report.

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