Asploro Journal of Biomedical and Clinical Case Reports
(ISSN: 2582-0370)

Case Report

DOI: https://doi.org/10.36502/2020/ASJBCCR.6201

Early Diagnosis and Treatment of a Small Gastric Stromal Tumor – A Case Report and Literature Review

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Received date: 21 May 2020; **Accepted date**: 06 June 2020; **Published date**: 14 June 2020

Citation: Binh LT, Mao NV, Huy TV, Tri NH, Khoan LT. Early Diagnosis and Treatment of a Small Gastric Stromal Tumor – A Case Report and Literature Review. Asp Biomed Clin Case Rep. 2020 Jun 14;3(2):135-40.

Abstract

Gastrointestinal stromal tumor (GIST) is the most common mesenchymal tumor of the gastrointestinal tract and the stomach is the most affected site. Clinical manifestation is erratic depending on the tumor's location, size, histological type, and stage. Ultrasound is useful in detecting gastric intramural tumor whereas computed tomography is the imaging modality of choice in further evaluating tumor characteristics and expansion, treatment planning, and monitoring recurrence. Definite diagnosis of GISTs is based on the immunohistochemical study. Early diagnosis and complete surgical resection of the tumor yield good long-term outcomes. We present a case of a small gastric GIST which was early detected and treated and the patient has tumor-free during 8 years of follow-up.

Keywords

Gastrointestinal Stromal Tumor, Ultrasound, Computed Tomography, Immunohistochemical, Surgical Resection

Background

Gastrointestinal stromal tumors (GISTs) represent a minority of the gastrointestinal malignancies but are the most common (80%-90%) mesenchymal subepithelial tumors of the gastrointestinal tract [1-3]. These tumors occur most commonly in the stomach (60-70%), followed by the small intestine (20–30%), duodenum (4-5%), rectum (4-5%), colon (<2%), esophagus (<1%) and are rarely found in the peritoneum, mesentery and omentum [1,2]. The true incidence of GISTs is unknown and the tumors are probably much more common than current data suggest [2]. One study reported very small gastric stromal tumors in 22% of autopsies in adults greater than 50 years of age [2]. Clinical symptoms of GISTs are nonspecific and the tumors are usually found parenthetically by endoscopy or imaging, especially for those smaller than 2 cm [1]. Small gastric GISTs less than 2 cm have a 100% cure rate after complete surgical resection [4]. Therefore, early diagnosis and prompt treatment of GISTs is very crucial to improve the prognosis of the disease [1,4]. We report a case of a

small GIST which was early diagnosed and treated and the patient has tumor-free during 8 years of follow-up.

Case presentation

Based on our institutional policy, the Institutional Review Board is waived for a case report. A 57-year-old woman presented to our institution with a chief complaint of a long lasting epigastric discomfort and intermittent pain. The pain was irregular, vague without spreading or progression or correlation to alimentary habit. She had no nausea or vomiting, neither hematemesis nor hematochezia. She did not report any changes in her bowel habit and had not experienced any recent fevers. She denied any history of alcohol consumption, cigarette smoking, and non-steroid anti-inflammatory medications intake and weight loss. Her past medical history and previous routine abdominal ultrasound check were unremarkable. On physical examination, her heart rate, blood pressure, respiratory rate, and body temperature were within the normal range. Cardiovascular and respiratory findings were unremarkable. There was little to no tenderness to palpation in the upper abdomen. Neither bowel distension nor abnormal motility was noted. Auscultation of bowel sound revealed normal. General blood and urine tests were within normal range. Gastritis was the primary clinical diagnosis. An esophagogastroduodenoscopy was indicated for further assessment and an abdominal ultrasound was also performed to rule out other etiologies.

Abdominal ultrasound (US) showed a homogenous hypoechogenic mass located within the posterior gastric wall, protruding into the inner lumen. The mass was oval in shape, had a regular and well-defined margin without infiltration. There was no gastric wall thickening and the layers remained intact. No evidence of hypervascularity on Color Doppler ultrasound (CDUS) of the mass (**Fig-1A**, **Fig-1B**). No other abnormalities were seen on US. On endoscopy, an oval submucosal lesion was found in the antrum bulging into the stomach cavity. The mass was relatively firm on compression and the mucosa was nearly intact without evidence of ulceration or linitis plastica (**Fig-1C**). An abdominal computed tomography (CT) was then indicated. Axial contrast-enhanced computed tomography (CECT) showed a well-defined, ovoid, homogeneous, intramural mass arising from the gastric antrum (arrow). The mass was measured 2cm in diameter. After contrast administration, a slight and homogenous enhancement was noted. There was no evidence of adjacent tissue infiltration or lymphadenopathy or ascites. CT scan findings were suggestive of an intramural stromal tumor (**Fig-2**).

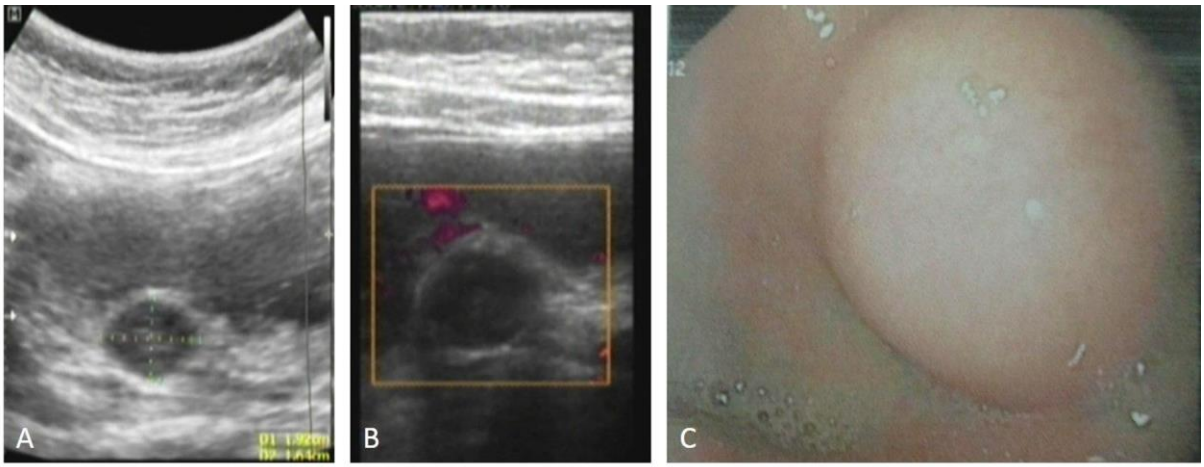

Fig-1(A,B,C)

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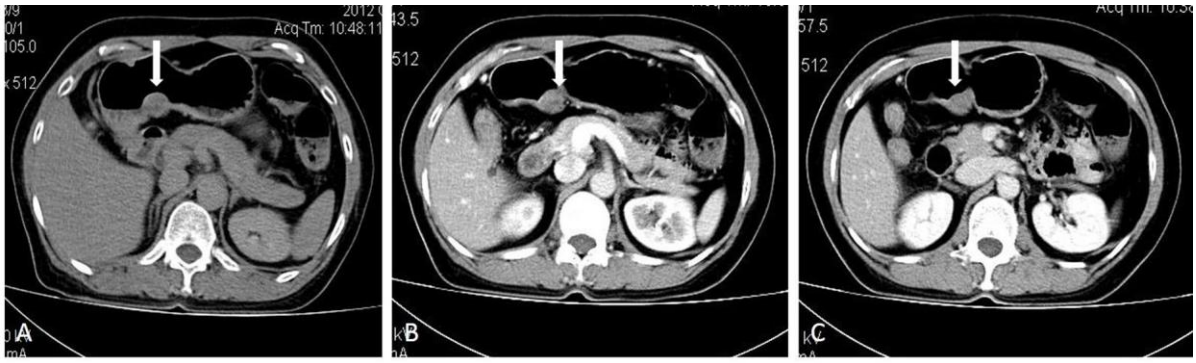

Fig-2

A subtotal gastrectomy was performed after a multidisciplinary discussion. Gross section showed a submucosal tumor located in the pyloric antrum which is firm and fleshy in nature (**Fig-3**). The mucosal surface remained normal. The diagnosis of low-grade GIST was confirmed histopathologically with CD117 (+), CD34 (+), and Desmin (-) (**Fig-4**). No further treatment was necessary. The patient has been followed up regularly during 8 years at the outpatient clinic without evidence of recurrence.

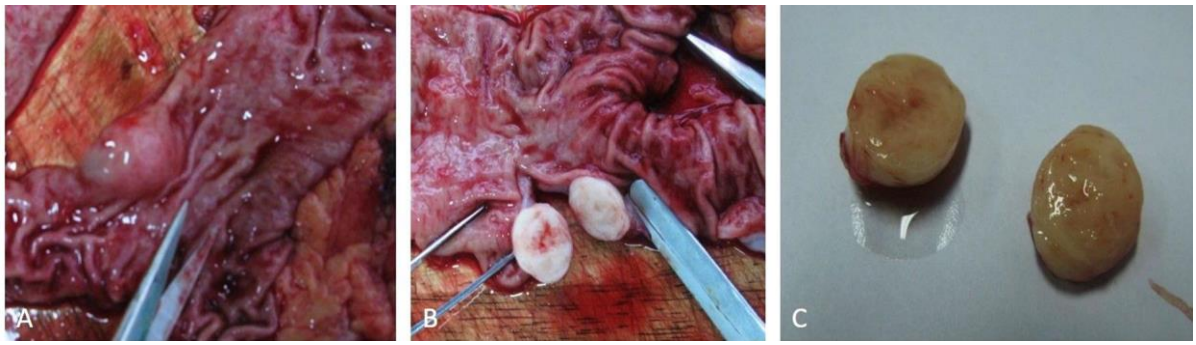

Fig-3

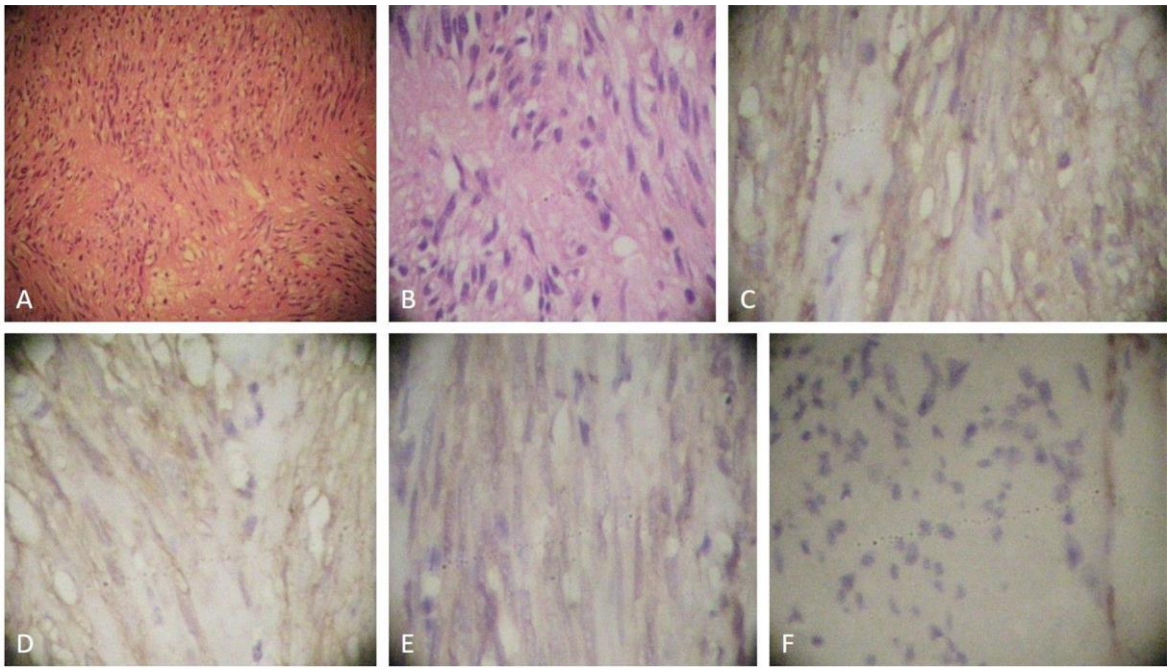

Fig-4

Discussion

Intramural gastric tumors are typically mesenchymal in origin and include GISTs, non-GIST sarcomas, lipomas, leiomyomas, schwannomas, glomus tumors, hemangiomas, inflammatory fibroid polyps (IFPs), inflammatory myofibroblastic tumors (IMFTs), and plexiform fibromyxoma [3]. The term of the stromal tumor was first described as a separate entity by Mazur and Clark in 1983 and Schandelbrand and Appleman in 1984 [5]. GISTs derive from a precursor of the interstitial cells of Cajal (ICC), normally present in the myenteric plexus in the muscular layer of the GI tract, and are clearly distinct from other mesenchymal tumors, such as leiomyoblastoma, leiomyomas or leiomyosarcomas [2,3,5]. Over 90% of GISTs occur in adults over 40 years old though the tumors have been reported in all ages without sex predilection [6].

Clinical manifestation of GIST is erratic. Common presentations include vague abdominal pain, palpable mass, gastrointestinal bleeding, fever, anorexia, weight loss, and anemia. Clinical symptoms mostly depend on the size and site of the lesion, with the most common symptom being bleeding into the bowel or abdominal cavity secondary to ulceration of the tumor mucosa [5,7]. Our patient also presented with non-specific symptoms. Sonographic findings were suggestive of an intramural gastric tumor. In our institution, US is usually indicated as a first-line imaging modality. Even though US is not sensitive in examining the gastric surface, it can initially detect intramural mass with a fluid filled stomach which is necessary to provide a good sonographic window.

To date, CECT is the imaging modality of choice for the localization, characterization, and staging of GISTs [5,6]. Tumor size, shape, margins, growth pattern, enhancement pattern, and enlarged vessels were significantly different between the low-grade and high-grade malignant potential groups [7]. Small GISTs (<5 cm), which are usually benign, appear on CT as sharply margined, smooth-walled, homogeneous, and intraluminal soft-tissue masses involving the wall of the GI tract [8]. Larger lesions often have well- or ill-defined margins; inhomogeneous density both on unenhanced and CECT due to hemorrhage, necrosis, or cystic degeneration; combined intraluminal/extraluminal growth and a tendency to spread to surrounding structures. Mural calcification is rarely seen [7-9].

The clinical diagnosis is mainly based on imaging, as biopsy runs the risk of tumor rupture or seeding of the biopsy tract, in otherwise resectable disease [8,9]. Besides, endoscopic ultrasound (EUS) allows assessment of the tumor size, border, internal echoes, layer of the GI wall from which the lesion arises, and other tumor morphological characteristics [7]. Differential diagnosis of GISTs includes schwannomas, leiomyomas, leiomyosarcomas, congenital cyst, carcinoma and neuroendocrine tumor [2,3,5]. Occasionally, gastric adenocarcinoma or lymphoma may demonstrate intramural growth and mimic a GIST [3]. Therefore, incorporating other imaging modalities is beneficial in making the diagnosis and evaluating the tumor components and origination [10].

Histologic characteristics of GISTs usually consist of interlacing whorls of spindle-shaped cells with eosinophilic cytoplasm and elongated nuclei, although tumors completely composed of ovoid cells are not uncommon. ICC are KIT, or CD117 positive, therefore, immunoreactivity to c-KIT or DOG1 confirms the diagnosis of GISTs in 80% of patients whereas immunoreactivity to desmin helps differentiate a leiomyoma from GISTs [2,3,9,10].

GISTs are assessed for risk of progressive disease on the basis of their mitotic rate, size, and location. Origin in the stomach is a favorable prognostic factor, and gastric GISTs less than 2 cm in size may have no or extremely low malignant potential [3]. Danti et al summarized the prognostic value of CECT in GIST from various investigations (**Table-1**). Other findings which imply unfavorable prognosis are intratumoral hemorrhage, necrosis, cavitation and cystic degeneration while calcification does not affect prognosis [6].

Previous studies recommend active surveillance of a GIST < 2 cm without signs of malignancy, though a small GIST does not exclude the malignant potential [1]. Conversely, some investigators advocate that every

GIST should be considered potentially malignant and all GISTs without metastasis need resecting [4]. Our patient was highly suspected a stromal tumor on imaging, thus a complete surgical resection was attempted after a multidisciplinary discussion and obtaining patient consensus. The pathological result confirmed a low-grade GIST. The prognosis of a low-risk GIST after complete resection is promising, with a 5-year survival rate up to 30% [10] and nearly 60% of all patients with GISTs are cured by surgery [5]. Recently, the introduction of neoadjuvant imatinib improves overall survival compared to resection alone [5,10]. In addition, interventional procedures such as radio-frequency ablation, transarterial yttrium-90 radioembolization, and transarterial chemoembolization have all been attempted in patients with GIST metastases [5].

Table-1: Relationship between different CECT features and prognostic outcomes in GISTs [6]

CT characteristics	Favorable prognosis	Intermediate prognosis	Unfavorable prognosis
Site	Stomach	Duodenum or rectum	Jejunum or Ileum
Size	< 5cm	5 – 10 cm	> 10 cm
Single/Multiple	Not related	Not related	Not related
Margin	Regular	-	Irregular
Enhancement	Homogenous	-	Heterogenous

Conclusion

The present case shows that ultrasound can play an important role as a useful imaging modality to detect an intramural gastric mass and assist in proper diagnosis. Prompt detection and diagnosis together with a comprehensive multidisciplinary approach are critical to provide the patient with proper treatment and a better prognosis.

Conflict of Interest

All authors have read and approved the final version of the manuscript. The authors have no conflicts of interest to declare.

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