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Department of Surgery, Mubarak Al-Kabeer Hospital, Jabriya, Kuwait

Corresponding author:
Sarah Qassim
Department of Surgery,
Mubarak Al-Kabeer Hospital,
Jabriya,
KUWAIT
Tel: +96566447299
E-mail: sarahqassim89@gmail.com

ABSTRACT

Gastrointestinal stromal tumors (GIST) are the 3rd common mesenchymal tumors. This case report is one of the rare presentations of a cystic GIST. It starts when a 57-year-old male complained of abdominal pain and mass. Several investigations were performed including an abdominal ultrasound. It showed an abdominal cystic lesion 5cm in size. A contrast-enhanced computed tomography scan of the abdomen was done and revealed same finding with uncertain origin. Due to that Magnetic resonance cholangiopancreatography and endoscopic ultrasound were performed and showed a cystic mass originating from the pancreas. Fine needle aspiration showed gastrointestinal stromal tumor. The patient underwent an 8cm mass resection of the gastric body via laparotomy. Immuno-histology study showed GIST. After that, the patient recovered well with no recurrence in follow-up. Gastrointestinal stromal tumors are commonly presented as a solid lesion and cystic appearance is rare and can be misdiagnosed as pancreatic pseudocyst.

KEY WORDS: gastrointestinal tract, mesenchymal tumors, oncology

INTRODUCTION

Gastrointestinal stromal tumors (GIST) are the most common subtype of mesenchymal neoplasms of the gastrointestinal tract with a prevalence of less than 1%.[1] GISTs typically occur in older adults, with a male predominance above 50 years of age, median age of 60-65 patient age and rarely under the age of 40.[2] They are most commonly located in the stomach followed by the small intestine, colon and rectum, and esophagus; with incidence of 60%, 20%, 5% and less than 5% respectively.[3] Initial patient evaluation should include a computed tomography (CT) scan and endoscopic ultrasound (EUS) might be of additional value to rule out neoplastic cells.[3] Cystic based GIST are uncommon as the majority are submucosal in origin; this case study represent a GIST with a cystic changes.

CASE REPORT

A 57-year-old gentleman presented with epigastric and right upper quadrant pain of two months duration. The pain was intermitted, radiating to the left side with no aggravating or relieving factors. The patient also gave a history of chronic diarrhea of 6 months duration with no blood or mucus discharge. There was no nausea, vomiting, loss of appetite or weight. His abdomen was soft with no tenderness and epigastric mass was felt. The laboratory investigations were within accepted range. The patient had an abdominal ultrasound showed irregular thickened wall with rounded cystic structure 5x7 cm. Computerized tomography (CT) scan of the abdomen showed left hypochondriac central mass. Tumor Markers CEA, Ca 125, Ca 19.9, Ca 15.3, AFP were all within normal range.

Magnetic Resonance Cholangiopancreatography (MRCP) showed a cystic mass with irregular thickened wall seen in the left Para-pancreatic area, around its tail, splenic hilum and retro-stomach. 7x8x9 cm. The mass has almost rugae like anterior wall and it is inseparable from the greater curve of stomach. The impression was a left upper quadrant abdominal mass, more in favor of exophyatic gastric tumor (GIST) (Figure 1).

EUS was performed and an external compression was noticed at the greater curvet at 40 cm from the

incisors. Difficult to assess its origin, but most likely arising from the tail of pancreas. Largest diameter 8X6.5 cm lesion was cystic in nature with nodular components and thickened walls. The largest nodule 2.4x0.7cm. The thickened wall diameter measured 3.6cm. single septum identified. Lesion was Doppler negative. It was hypo/isoechoic in nature and anechoic of the cystic component. Three passes were performed and samples were taken, 65cc of serosanguinous material was drained.

The impression was that the lesion is cystic in nature with nodular component and thickened wall suggestive of possible malignant transformation. (Figure 2) FNAC showed a positive CD117, DOG1 and negative S-100, which conclude a GIST. At first, a diagnostic laparoscopy was performed and a mass was noted behind the stomach; however, mass origin couldn't be identified via laparoscopy and conversion to laparotomy was done after failure of progression. So in laparotomy, a pedunculated gastric mass from posterior wall of the stomach was identified and wedge resection with 2cm margin was performed. Histopathology of the gastric mass showed a GITS, grade 1, (T3 Nx Mx). Tumor size was 8.5 greatest diameter, unifocal, spindle cell subtype, 1/50 HPF mitotic rate, low risk with negative margins. Immuno-histopathology showed positive CD117, DOG1 and negative CD34+, SMA (focal+), S100 and Desmin.

The operation was uneventful with high recovery rate. Case was discussed with a multidisplinary team of radiologist, histopathologist, surgeon and oncologist; in which, only follow up was advised with no need of adjuvant therapy. On follow up, patient was symptom free with no recurrence of tumor on further investigation.

DISCUSSION

GISTs are the most common sarcomas of the gastrointestinal tract. They can occur from esophagus to rectum, but most commonly the have been found in the stomach.[4] Gastrointestinal stromal tumors originated from intestinal cell of Cajal (ICCs). GISTs are the only tumors that express both c-Kit and CD34 as the development of ICCs and other stem cells are dependent on SCF-KIT oncogene. Also, ICCs are located in and near the circular muscle layers of the gastrointestinal tract, for that GIST usually originate from the submucosa and muscularis propria and present as solid tumors.[5] Mostly patients experience melena due to central necrosis in the lesion and others are usually symptomatic if the lesion is 5cm or larger.[6] In this case the patient experienced only diffuse abdominal pain and mass. CT scan is the gold standard method and with the MRCP and EUS-guided fine needle aspiration revealed a cystic pancreatic mass with a malignant transformation. However, EUS-guided fine-needle aspiration did not reflect the tumor subtype and mitotic ratio and thus proper management and prognosis cannot be achieved via FNAC result.[7] Exophytic stromal tumors with cystic changes have been previously reported; however, they are rarely observed and during preoperative diagnosis, these masses may be misdiagnosed as duodenal, pancreatic or liver cyst, duplication cyst, diverticulum or even a metastatic lesion from a primary liver or pancreatic tumor.[8-9] The patient underwent a diagnostic laparoscopy that revealed a gastric mass from the posterior wall of the stomach. The surgery was converted to upper midline laparotomy in which wedge resection of the pedunculated mass was done. Histopathology of the resected lesion showed a grade 1 GIST, spindle subtype with mitotic rate of 1/50 HPF positive CD117. So, an accurate diagnosis and prognosis of the lesion is only done after surgical resection. GISTs are relatively rare tumors that require a multimodal approach to management. Surgery remains the definitive treatment and is recommended for

primary disease (tumors ≥2 cm). This is followed by tumor classification in to low or high risk of malignancy according to tumor size, mitotic count, and location. And the introduction of imatinib will improved the clinical outcome of metastatic GIST.[10] For that, a multidisciplinary team was conducted in which the patient will be followed by the surgical and ontological team for the progression of the disease.

CONCLUSION

Most of GISTs are solid tumors, and cystic change is uncommon. Preoperatively, an intra-abdominal cyst may be considered pancreatic in origin. Resection of the mass is the standard treatment and with malignant lesion imatinab is prescribed.

Take home message:

1- GISTs are solid tumors and they can present as a cystic lesion.
2- Considering GIST as a differential diagnosis of a cystic lesion.
3- Appropriate diagnosis and management of GIST is resection of lesion. So, Endoscopic ultrasound FNAC is not the gold standard for diagnosis.

ACKNOWLEDGMENT

Author's Contribution

Sarah Qassim, the corresponding author has written the case report and the co-authors have collected the recourses and patient consent. The authors read and approved the final manuscript.

No conflict of interest.

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FIGURES

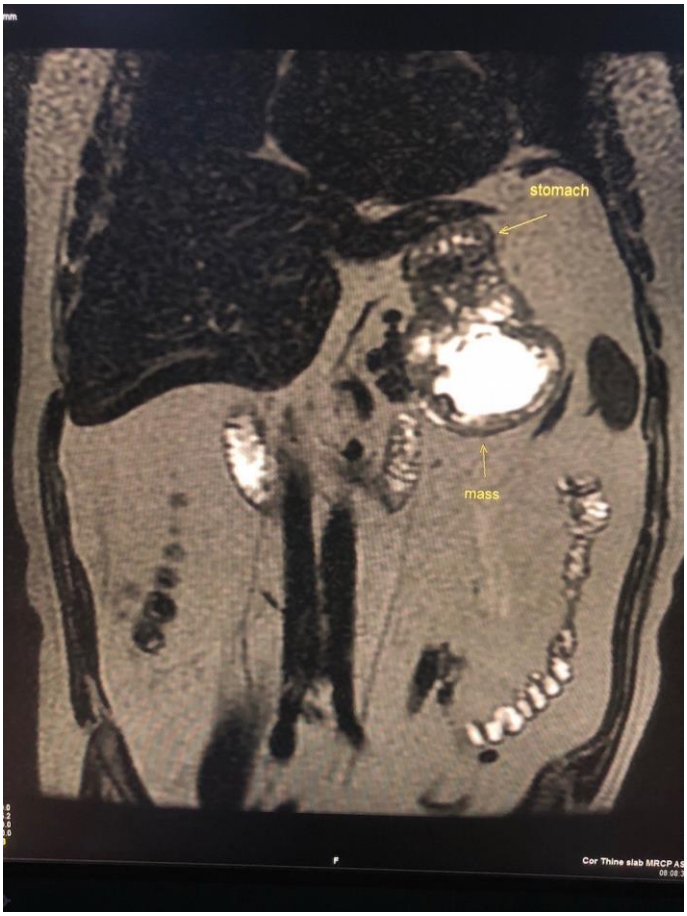

Figure 1: Endoscopic Ultrasound. Compression noted at the greater curvature.

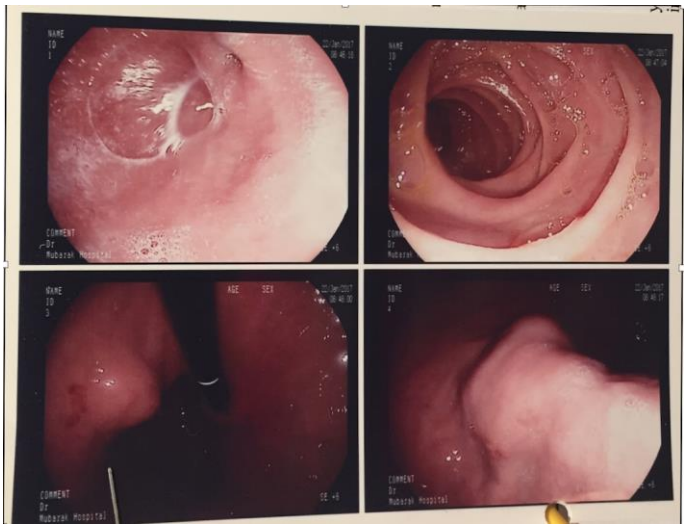

Figure 2: Magnetic Resonance Cholangiopancreatography (MRCP). A cystic mass noted, retro gastric 7x8x9cm.