—Case Report—

A duodenal gastrointestinal stromal tumor with a large central area of fluid and gas due to fistulization into the duodenal lumen, mimicking a large duodenal diverticulum

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) can occur anywhere along the gastrointestinal tract especially the stomach and upper small bowel. They are usually solid, but cystic degeneration, necrosis, and focal hemorrhage have been described in larger tumors leading to central necrotic cavitation. The most sensitive marker of GIST is CD117 (c-kit). In computed tomography (CT) scan, it is often difficult to decide the origin of the primary tumor, especially in large GISTs. We report an incidental case of a large duodenal GIST fistulizing into the second part of the duodenum with a large amount of fluid and gas inside, mistaken for a cystic pancreatic neoplasm by CT and mistaken for a duodenal diverticulum by endoscopic ultrasound.

Key words: Duodenal diverticulum, endoscopic ultrasound, gastrointestinal stromal tumor, pancreatic cyst

INTRODUCTION

Cystic lesions in the upper abdomen usually originate from the pancreas being mostly nonneoplastic as pseudocysts, however, cystic neoplasms as mucinous and serous cysts also represent a good percentage of such lesions. Gastrointestinal stromal tumors (GISTs) can occur anywhere along the gastrointestinal tract especially the stomach and upper small bowel. A few cases of pancreatic GISTs have been described.[1] They usually present as solid tumors, however large areas of internal hemorrhage and cystic degenerations may occur.[2] We report a case of a large duodenal GIST fistulizing into the second part of the duodenum with a large amount of fluid and gas inside, mistaken for a cystic pancreatic neoplasm by computed tomography (CT) and mistaken for a duodenal diverticulum by endoscopic ultrasound (EUS).

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Website: www.eusjournal.com

DOI: 10.4103/2303-9027.163018

CASE REPORT

A 42-year-old female was presented by vague central abdominal discomfort not responding to symptomatic treatment for months. She was admitted to our hospital with an attack of melena not associated with hematemesis. Upper gastrointestinal (GI) endoscopy showed a smooth bulge in the second part of the duodenum, measuring 30 mm × 30 mm, just above the papilla [Figure 1] with a small depression at its summit, too small to allow the passage of the upper GI endoscope [Figure 2]. Abdominal CT revealed a large epigastric complex cystic lesion, most likely a cystic pancreatic neoplasm [Figure 3] with fluid and gas inside but no ascites, lymphadenopathy or evidence of metastasis. EUS showed a large complex

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Received: 2014-06-27; **Accepted:** 2014-07-26

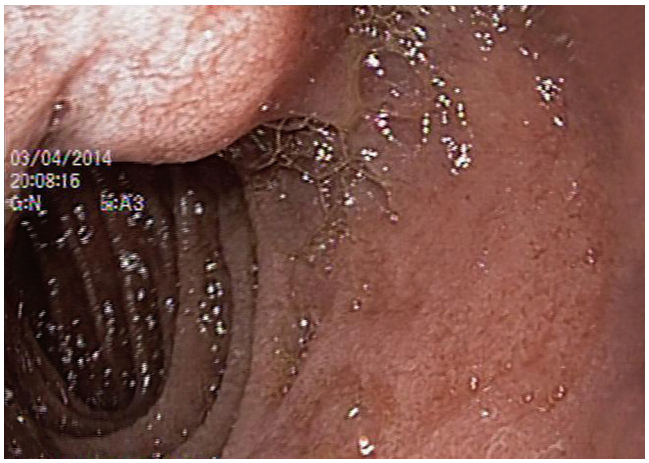

Figure 1. Gastrointestinal stromal tumor seen as a bulge in the mid-second part of the duodenum

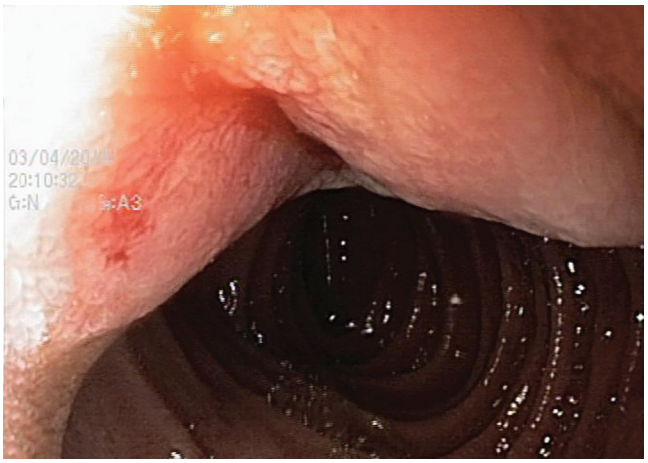

Figure 2. Gastrointestinal stromal tumor with a small depression at its summit

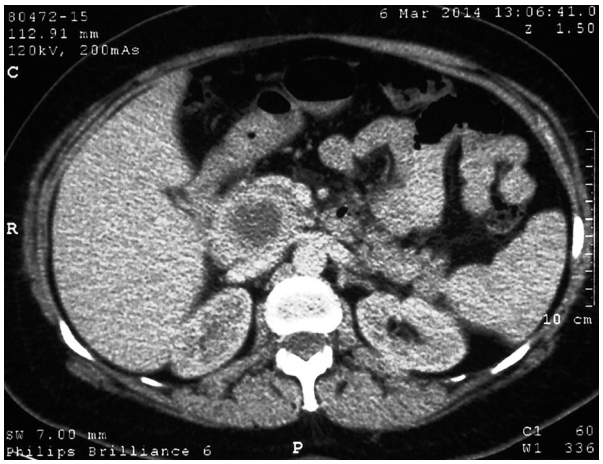

Figure 3. Computed tomography appearance of the duodenal gastrointestinal stromal tumor mistaken for cystic pancreatic neoplasm

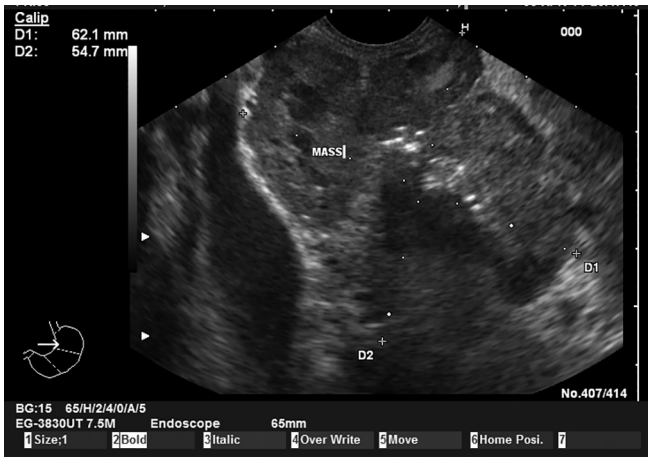

Figure 4. Endoscopic ultrasound appearance of the duodenal gastrointestinal stromal tumor with a large area of fluid and gas inside mistaken for a duodenal diverticulum

cyst, measuring 55 mm × 62 mm, with thick irregular wall and a large central area of fluid and gas shadow inside [Figures 4 and 5]. The lesion appeared separable from the pancreatic head, but it was inseparable from the duodenal wall. Due to the presence of fluid and gas inside the duodenal related cystic lesion, the diagnosis of a large duodenal diverticulum with a narrow mouth was suggested. As the wall of the cystic lesion was thick and irregular, EUS-fine needle aspiration (EUS-FNA) was done to verify the possibility of a mass developing inside a duodenal diverticulum [Figure 6]. However, EUS-FNA revealed nonspecific inflammatory process. After consulting the hepatobiliary surgical team, surgical exploration was decided. Laparotomy revealed a large mass originating from the duodenal wall and encroaching upon the pancreatic head [Figure 7]. Intraoperative biopsy and frozen section revealed a muscular tumor. Pancreaticoduodenectomy was done. Postoperative incision in the lateral wall of the resected second part of the duodenum showed a pin hole opening of the mass inside the medial wall of the duodenum [Figure 8]. This explains the presence of a large amount of fluid and gas inside the mass simulating a duodenal diverticulum. Histopathological examination and immunohistochemical staining revealed a spindle cell neoplasm, positive for c-kit 34 and 117 and the final diagnosis of a duodenal malignant GIST was made.

DISCUSSION

Gastrointestinal stromal tumors are rare neoplasms, representing 1% of all benign gastrointestinal tumors with an annual incidence of approximately 4/million.[3] They were initially classified as leiomyomas and leiomyosarcomas due to the similar morphological appearance, but now with the discovery of mutational activation of c-kit proto-oncogene, GIST became a separate entity.[4]

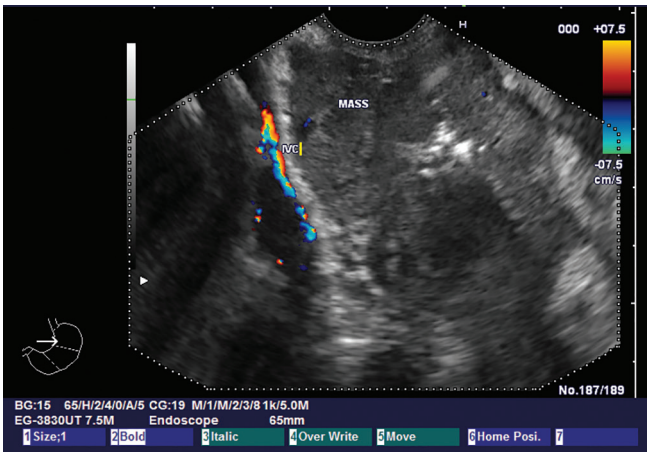

Figure 5. Inferior vena cava adjacent to the large duodenal gastrointestinal stromal tumor

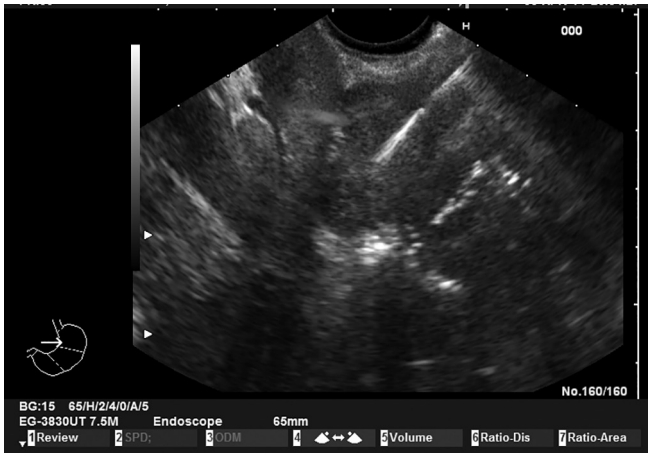

Figure 6. Endoscopic ultrasound-fine needle aspiration from the solid part of the large cystic lesion

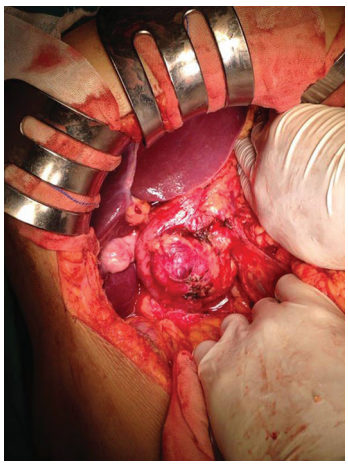

Figure 7. Intraoperative findings of the duodenal gastrointestinal stromal tumor

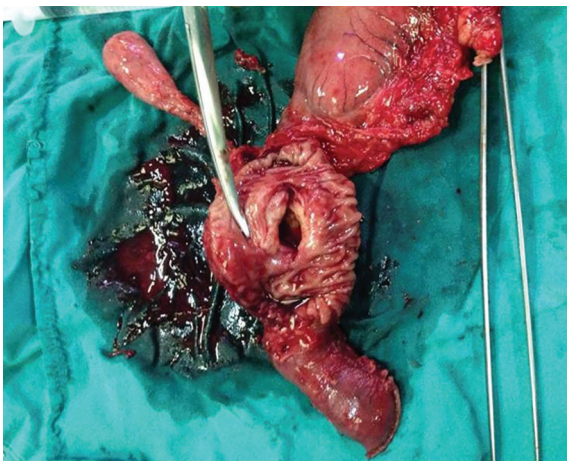

Figure 8. Postoperative finding of the duodenal gastrointestinal stromal tumor with a hole connecting it to the duodenal lumen

Gastrointestinal stromal tumors with cystic changes may be observed in the following situations:

1. Primary cystic GIST, in which the main structure comprises cystic tissue with a pseudocapsule, rarely invading the surrounding organs;
2. Malignant GIST with cystic degeneration, caused by rapid growth of the tumor, which due to insufficiency of the internal blood supply results in necrosis and liquefaction;
3. When the tumor metastasizes to the liver and pancreas, the metastatic lesion is usually cystic in nature, often confused with liver cysts and pancreatic cysts; and
4. On treatment with imatinib, malignant GISTs show cystic degeneration.[5-7]

In our case, the prominent cystic changes were produced by two factors; cystic degeneration due to its large size and invasion of the duodenal wall by the tumor causing fistula formation between the two structures. The rarity of our case was the presence of a large amount of gas [Figures 4 and 5] transmitted from the duodenal lumen to the tumor and hence that it was mistaken for a duodenal diverticulum during EUS examination.

CONCLUSION

Gastrointestinal stromal tumors may present with a large area of cystic breakdown mimicking complex cystic lesions. There may also be gas shadow inside if it is communicating with the gastrointestinal lumen mimicking a gastrointestinal diverticulum.

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How to cite this article: Okasha HH, Amin HM, Al-Shazli M, Nabil A, Hussein H, Ezzat R. A duodenal gastrointestinal stromal tumor with a large central area of fluid and gas due to fistulization into the duodenal lumen, mimicking a large duodenal diverticulum. Endosc Ultrasound 2015;4:253-6.

Source of Support: Nil. **Conflicts of Interest:** None declared.