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Case Report

Uncommon presentation of duodenal gastrointestinal stromal tumor as large paraduodenal cyst: case report

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) are rare solid mesenchymal neoplasms. Small bowel GISTs have predominantly an outward growth and multifocal when associated with neurofibromatosis-1 (NF-1). When the duodenum is involved, lesions may be more challenging if growing towards the pancreas. Their prevalence is unknown, and diagnosis is usually incidental. Owing to the complex anatomy of the duodenum and pancreatic head, GISTs can be misdiagnosed as pancreatic head tumors, however they presenting as cyst is very rare. Literature shows very few reported cases of cystic GISTS, all arising from the stomach or pancreas where they have been mistaken for pseudocysts or even a mucinous cystadenocarcinoma of the pancreas and only one case of duodenal GIST presenting as cyst has been reported so far. Here, we report a case where investigations including radiology and upper gastrointestinal endoscopy suggested it to be a paraduodenal cyst, but histopathological examination of the specimen resulted in an unexpected diagnosis of GIST. We concluded that non-invasive imaging like CT, MRI, EUS and whenever possible FNA are important for the diagnosis and treatment of GISTs. Surgical resection with clear margin is the desired treatment for a localised duodenal GIST. Also, the feasibility of limited resections such as duodenectomy without pancreatic/ampullary involvement as in our case report where we preserved the pancreas reduces the morbidity and complications associated with extensive surgeries like pancreaticoduodenectomy. Also, Imatinib is an effective therapeutic agent with a remarkable response and prolonged survival.

Keywords: Paraduodenal cyst, Masquerading GIST, Duodenectomy

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) although rare tumors, usually present as solid lesions anywhere in the gastrointestinal tract but mostly in stomach and small intestine. But their atypical presentation as cystic lesions pose a diagnostic dilemma. Also, duodenal cysts themselves are uncommon lesions and are mostly benign.[1] Moreover, duodenal GIST presenting as paraduodenal cyst is extremely rare and to the best of our knowledge only one case has been reported so far.[2] Our case is being reported because although investigations including radiology and upper gastrointestinal endoscopy suggested it to be a para-duodenal cyst, histopathological examination of the specimen resulted in an unexpected diagnosis of GIST.

CASE REPORT

A 57-year gentleman with no comorbidities had an incidentally detected paraduodenal cyst. He had a history of significant weight loss (over 10 kg in 3-5 months) with loss of appetite. Well-defined, mobile, right hypochondrial firm to hard lump (about 6×8 cm) was palpable.

CT scan showed 7.1×6.9×6.4 cm well defined thick walled peripherally enhancing lesion arising from

paraduodenal "C- loop" region abutting D2 segment, head and uncinate process of pancreas (Figure 1). Endoscopy suggested extrinsic compression of D2 segment with fistulous tract and fluid coming out through it in D2 distal to the ampulla. However, these findings were not diagnostic for malignancy. The rest of the laboratory reports were normal.

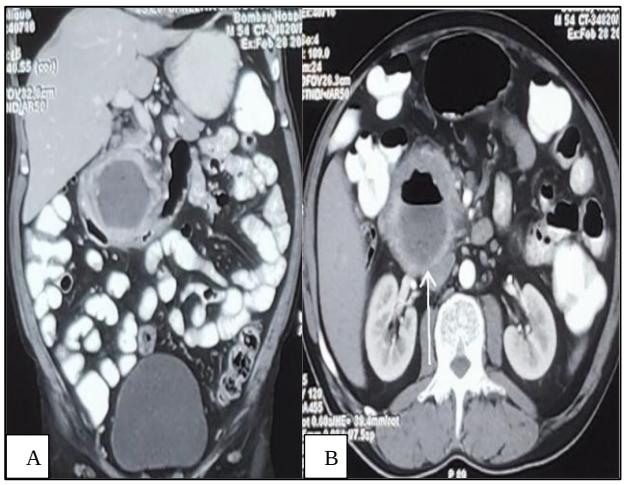

Figure 1 (A and B): Coronal and axial views of CT abdomen and pelvis showing para-duodenal cyst with air-fluid levels.

Patient underwent pancreas preserving duodenectomy surgery (Figure 2). Intraoperatively the cyst was freely mobile within the lesser sac with no attachment to the liver but abutting head and uncinate process of pancreas although fat planes were preserved and part of its wall inseparable form D2 segment. He underwent pancreas preserving duodenectomy of D2 and D3 segment followed by gastrojejunostomy. Postoperatively patient had delayed gastric emptying which resolved gradually with conservative management.

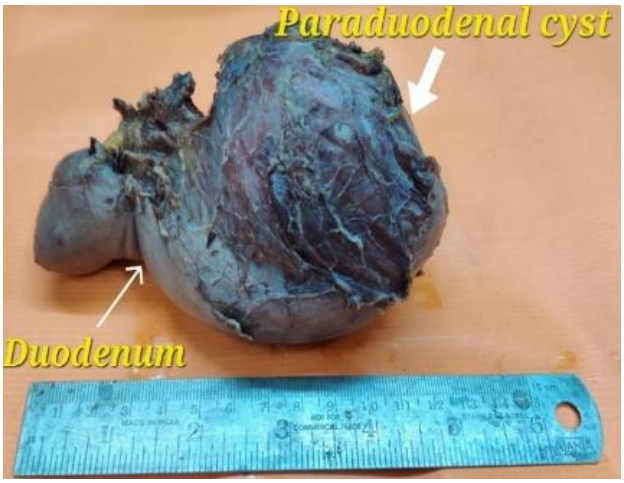

Figure 2: Gross specimen of paraduodenal cyst.

Histopathological examination revealed spindle cell tumor with central necrosis, hemorrhagic areas and extensive cystic degeneration with rare mitoses indicating the unexpected diagnosis of duodenal GIST, which was confirmed on immunohistochemistry (CD117, DOG-1, Vimentin, SMA, Calponin as positive and Desmin, CD-34, S-100 as negative; Ki-67 very low).

DISCUSSION

GISTs are mesenchymal neoplasms of the gastrointestinal (GI) tract featuring interstitial cell of Cajal differentiation. Interstitial cells of Cajal serve as pacemakers of GI motility, providing an interface between autonomic nerve stimulation and the muscle layer of the GI wall.[3]

More than half of GIST cases arise from the stomach, 25% from the small bowel, 5% from the rectum, and a small minority from the esophagus.[4]

Classic gastric GISTs can either have an outward growth or grow within the gastric wall. They are typically multifocal, instead, when associated with SDH deficiency. Small bowel GISTs mostly have an outward growth. When the duodenum is involved, lesions may be more challenging if growing toward the pancreas.

GISTs can grow outward from the gastrointestinal (GI) wall, which can make them difficult to detect early. As they grow, they can become large enough to be felt as abdominal masses. Additionally, they can cause symptoms such as GI bleeding, hemoperitoneum (blood in the abdominal cavity), or perforations, which often lead to their discovery. This late detection can complicate treatment and prognosis, highlighting the importance of early detection methods.[5]

GISTs are typically identified as solid masses, varying in size and location within or outside the gastrointestinal tract. They are well defined but lack a true capsule, and they can exhibit either an endophytic (growing into the GI wall) or exophytic (growing outward from the GI wall) growth pattern. While larger GISTs may show small cystic areas, they are not primarily cystic tumors.

Such uncommon presentations of GIST are very few as reported in literature.[6-12] In a retrospective study by Xue et al twenty cystic GISTs were analyzed from a single institution.[13] The study found that the majority of cystic GISTs were in the stomach (50%) or small intestine (45%), with one case in the omentum (5%). In fifteen cases, the diagnosis was either indistinct or misdiagnosed based solely on preoperative radiology.

Gross examination of the cystic GISTs in the study showed that the cystic component made up most of the masses and was filled with dark bloody fluid and necrotic debris in 18 cases. The study concluded that cystic GISTs represent an indolent subset of GISTs with favorable prognoses. They suggested that considering adjuvant imatinib therapy could be a prudent approach in

managing these cases. This recommendation aligns with the growing understanding of imatinib's effectiveness in managing GISTs, especially in cases where the tumors exhibit certain characteristics, such as being cystic.

Due to the complex anatomy of the duodenum and pancreatic head, GISTs can be misdiagnosed as pancreatic head tumors. However, it is exceedingly rare for GISTs to present as cysts in this region. This highlights the importance of thorough evaluation and consideration of various differential diagnoses when encountering cystic lesions in the duodenum and pancreatic head.

Surgical resection is typically the primary treatment for localized duodenal GISTs. Noninvasive imaging plays a crucial role in the diagnosis and treatment planning for GISTs. Imaging modalities such as computed tomography (CT), magnetic resonance imaging (MRI), and endoscopic ultrasonography (EUS) can provide valuable information for diagnosing duodenal GISTs. These imaging techniques can also help differentiate GISTs from other adjacent tumors, aiding in the development of an appropriate treatment strategy.[14-16]

Obtaining a preoperative diagnosis for duodenal GISTs can be challenging. In some cases, using endoscopic ultrasound (EUS) combined with fine needle aspiration cytology (FNAC) may help clarify the diagnosis. For instance, if imaging shows a cystic lesion with no solid component, this might lower the suspicion for malignancy. However, it is important to remember that GISTs can occasionally appear as predominantly cystic lesions, underscoring the need for thorough evaluation and consideration of all diagnostic possibilities.

Surgical R0 resection, which aims to remove the tumor with clear margins, remains the only curative approach for duodenal GISTs. There are several operative methods available for managing these tumors. Segmental resection options include duodenal transection with Roux-Y or Billroth II gastrojejunostomy (G-J), as well as end-to-end duodeno-duodenostomy (D-D). More extensive procedures, such as pancreaticoduodenectomy (PD), pancreas-preserving duodenectomy, and the Whipple procedure with pancreatojejunostomy, may be necessary in certain cases depending on the location and extent of the tumor. The choice of surgical approach depends on various factors, including the size and location of the tumor, as well as the surgeon's expertise and the patient's overall health.

In this case, a pancreas-preserving duodenectomy with gastrojejunostomy was performed because the fat planes were preserved, even though the tumor was abutting the pancreas. This approach aims to remove the tumor while preserving the pancreas. After surgery, careful follow-up is essential. If the resection margin is positive (indicating that some tumor cells may remain), targeted therapy with imatinib mesylate (Gleevec) may be necessary. Imatinib is a tyrosine kinase inhibitor that can be effective in treating GISTs, especially when used in a targeted manner based on the specific genetic characteristics of the tumor.

CONCLUSION

The cystic presentation of duodenal GISTs requires careful evaluation preoperatively due to the duodenum's complex anatomy and the need for a high index of suspicion for malignancy. Surgical resection with clear margins is the desired treatment approach, aiming to remove the tumor completely. Whenever feasible, limited resections should be considered to preserve organ function and minimize surgical morbidity.

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