CASE REPORT

A Gastrointestinal Stromal Tumor of the Stomach Mimicking as a Cystic Neoplasm of Pancreas: A Case Report and Review

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) are usually solid but cystic degeneration, necrosis, and focal hemorrhage have been described in larger tumors leading to central necrotic cavitation. Gastric GISTs can be divided into histologic subgroups including spindle cell and epithelioid variants. The most sensitive marker of GISTs from all sites is CD117(c-kit). A weak or absent expression of CD117 is characteristic for gastric GISTs with epithelioid differentiation. In CT scan, it is often difficult to decide the origin of the primary tumor especially in large GISTs. We presented a rare case of gastric cystic lesion mimicking the cystic neoplasm of the pancreas and have reviewed the literature. The problems of identifying tumor origin are highlighted.

Key words: Gastrointestinal stromal tumor - Pancreatic pseudocyst - Cystic neoplasm of pancreas - Gastric cystic lesion

Introduction

Cystic lesions of the pancreas are usually pseudocysts. The differential diagnosis of pancreatic cyst includes inflammatory pseudotumors with regressive cystic change, solid-pseudopapillary tumors and serous or mucinous cystic pancreatic neoplasms. A few cases of pancreatic gastrointestinal stromal tumor (GIST) have been described[1]. Occasionally cystic malignancy of structures surrounding the pancreas may secondarily involve it. Here, we present a rare case of gastric cystic lesion mimicking the cystic neoplasm of the pancreas. The problems of identifying tumor origin are described.

Case Report

A 48 year old male presented with upper abdominal discomfort since three months. Since 2 weeks he also noted an abdominal lump. There was no history of trauma. On examination, pallor was present and an ill defined, smooth, nontender, firm mass of approximately 10 cm x 8 cm was palpable in the epigastrium and left hypochondrium. It was relatively fixed and to get above the swelling was not possible. Routine laboratory investigations were normal except for anemia (hemoglobin: 9.5 g/dL). Abdominal computed tomography (CT) scan showed a 12.4 cm X 10.1 cm complex cystic mass (with solid component) with an eccentric enhancing nodule closely related to pancreatic tail (Figure 1). The mass was found to be splaying and indenting the stomach. The mass did not communicate with the pancreatic duct. There were no ascites, lymphadenopathy and evidence of metastasis. CT-guided cystic fluid aspiration showed few atypical cells. However, fluid amylase, CEA and CA 19-9 were normal. Serum CA 19-9 and CEA were also normal. Esophagogastroduodenoscopy revealed a gastric antral bulge, possibility of submucosal lesion and extrinsic compression was kept a possibility (Figure 2). The differential diagnoses of cystic tumor in this location were considered with possible...
origins from pancreas or stomach. Cystic neoplasm arising from the pancreas was the first possibility in view of CT scans appearances and it being relatively common.

Laparotomy revealed a unifocal cystic mass of dimension 15 cm x 12 cm x 8 cm occupying the lesser sac and involving stomach, the transverse colon and mesocolon. The en-block excision of mass with R0 margins with partial gastrectomy was done. Grossly the tumor was attached with the stomach wall. On cut section a large blood filled cavity with necrotic material was seen. A thin solid part was seen at one pole. Microscopy demonstrated a mesenchymal tumor arising from gastric submucosa comprising of epithelioid cells (Figures 3 and 4). Tumor cells were patchy positive for CD117 (Figure 5), and focally positive for CD34, and cytokeratin. It was also patchy positive for SMA and diffusely positive for vimentin, but negative for desmin, S-100 and...
chromogranin. Mitotic rate of 40 / 50 HPF and necrosis of about 90% extent was noted. There was no regional lymph node metastasis (0/3). A diagnosis of gastric GIST with epithelioid cell differentiation with high risk behavior was made. Postoperative recovery was good. He is doing well and under our follow up for last 4 months.

Discussion

GIST represents the most common type of mesenchymal tumor that arises from the alimentary tract. GIST is currently defined as a gastrointestinal tract mesenchymal tumor showing CD117 (c-kit protein) positivity at immunohistochemistry. GIST most commonly occur in the stomach (60%) followed by jejunum and ileum (30%). Only rarely they present as apparently primary extraintestinal tumours in the vicinity of the gastrointestinal tract[2].

The cut surface of GIST is typically granular, but as these tumors assume great dimensions over time, they may outgrow the blood supply of their tissue of origin, resulting in necrosis and cavitation[3, 4].

Gastric GISTs can be divided into spindle cell (70%) and epithelioid variants (30%)[5].Benign gastric GISTs often have an epithelioid morphology. This subtype often has a low mitotic rate and favorable prognosis[6-8]. However, our case had high risk behavior. About 10-30% of GISTs are malignant[5]. GISTs may invades adjacent structures or evoke local inflammatory reaction. Therefore, origin of large GISTs in the upper abdomen may be difficult to determine[6]. Gastric and duodenal GISTs may mimic as pancreatic lesions conversely, pancreatic cystic neoplasm may mimic gastric GIST[6,9-11]. Published reports of gastric GIST mimicking as pancreatic cystic lesions are summarized in Table. The epithelioid cell differentiation noted as common feature in reported cases of gastric GISTs mimicking as pancreatic cyst. Our case also had epithelioid cell differentiation as shown in Table.

On the basis of stratification, GISTs are classified into very low, low-, intermediate-, and high-risk categories based

<table>
<thead>
<tr>
<th>Ref</th>
<th>Age, sex</th>
<th>Clinical features</th>
<th>Initial investigation</th>
<th>Initial diagnosis</th>
<th>Gastric GIST suspected</th>
<th>HPE</th>
<th>Immunohistochemistry, Genetic study</th>
<th>Treatment, Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>6</td>
<td>51, F</td>
<td>Abdominal mass</td>
<td>CT - cystic tumor present between the stomach and The pancreas. EUS-Changes of chronic pancreatitis</td>
<td>Pancreatic pseudocyst</td>
<td>During laparotomy</td>
<td>Gastric GIST (epithelioid subtype) - low mitotic rate (1/50 HPF), Vimentin and CD34 (strong +), KIT (weak +), Cytokeratin (-), Synaptophysin (-), S100 (-), SMA (-)</td>
<td>PDGF Ralpha mutation not detected.</td>
<td>Resected. Relapse free follow-up 8 years.</td>
</tr>
<tr>
<td>8</td>
<td>65, F</td>
<td>Abdominal Discomfort</td>
<td>USG-pancreatic cystic tumor</td>
<td>Pancreatic pseudocyst</td>
<td>During laparotomy</td>
<td>Gastric GIST (epithelioid subtype) - low mitotic rate (1/50 HPF), KIT (weak +), Vimentin (+), CD34 (+), BCL-2(+), Cytokeratin (-), Synaptophysin (-), S100 (-), SMA (-)</td>
<td>PDGF Ralpha mutation detected.</td>
<td>Resected. Relapse free follow-up 3 years.</td>
</tr>
<tr>
<td>11</td>
<td>NA</td>
<td>Diarrhea, Pain, Abdominal mass</td>
<td>CT - pancreatic cystic lesion</td>
<td>Pancreatic cyst</td>
<td>During laparotomy</td>
<td>Epithelioid and spindle cells embedded in myxoid stroma, Vimentin (+) and CD34 (+), Genetic study-NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>Present case</td>
<td>48, M</td>
<td>Abdominal discomfort, Abdominal lump</td>
<td>CT-pancreatic cystic neoplasm. Cystic fluid- atypical cells</td>
<td>Pancreatic cystic neoplasm</td>
<td>During laparotomy</td>
<td>Gastric GIST (epithelioid subtype)- high mitotic rate (40/50 HPF), KIT (Patchy +), CD34 (focally +), Cytokeratin (focally +), SMA (patchy +), Vimentin (+), Chromogranin (-), Desmin (-), S-100 (-)</td>
<td>Genetic study not done.</td>
<td>Resected Relapse free followup 4 months</td>
</tr>
</tbody>
</table>

HPE, histopathology examination; CT, computerized tomography; USG, ultrasonography; EUS, endosonography; NA, information not available; +, strong/diffuse positive; -, negative
on size and on the number of mitoses[4]. The most sensitive markers of GISTs from all sites are CD117 followed by CD34. A weak or absent expression of CD117 is characteristic for gastric GISTs with epithelioid differentiation. Approximately 30% of GISTs express muscle markers, including SMA, calponin, and caldesmon. The rare expressed markers are BCl-2, S100, cytokeratin, nestin, desmin and vimentin [4]. In our case tumor cells were patchy positive for CD117 and focally positive for CD34 (Table).

CT scan is the single most important diagnostic test for demonstrating the size and anatomic location of the mass and treatment planning. Large GISTs (>10 cm) often have irregular border and inhomogeneous density. The disadvantage of CT is its inability to differentiate inflammatory adhesions from actual involvement of contiguous organs. In large GISTs, it is often difficult to decide if the primary tumor arises from the stomach, liver, pancreas, spleen, or colon[4,12].

The endoscopic appearance of GISTs is that of a submucosal lesion or of an extrinsic mass with or without mucosal lesion. Endoscopic ultrasound [EUS] usually coupled with fine-needle aspiration is the principal modality for diagnosis of gastric GISTs[13]; however, it is operator dependant. EUS done in one of the previously discussed case failed to identify origin of cyst[6]. Moreover, EUS and its expertise are not widely available in world. Therefore, treatment decision is often based on the CT-scan. Unfortunately, we also did not perform EUS.

Optimal surgical treatment of GISTs entails complete removal of the localized tumor with clear surgical margins including adjacent involved organs. Surgical resection has very good outcome as shown in our case.

**Conclusion**

In conclusion, the cases of gastric GISTs mimicking as pancreatic cyst mostly have epithelioid cell differentiation. A weak or absent expression of CD117 is characteristic for gastric GISTs (epithelioid subtype). In CT scan, it may be difficult to decide the origin of the large GISTs. Therefore, large gastric GIST (epithelioid subtype) with cystic changes may masquerade as pancreatic cyst and should be considered in differential diagnosis.

**References**


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