**CASE REPORT**

**SOLITARY FIBROUS TUMOR OF THE PANCREAS: A CASE REPORT AND REVIEW OF THE LITERATURE**

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**Abstract**

Pancreas is an extremely rare abdominal localization of the solitary fibrous tumor (SFT). It usually grows asymptptomatically for a long time before a diagnosis can be made on the basis of symptoms and/or mechanical complications. Due to the rarity and nonspecific clinical presentation, this entity is diagnostically challenging.

We present a 47-year-old man with a history of progressive epigastric pain for the last two weeks, and jaundice, who was admitted to hospital for further investigation. Cystadenocarcinoma was suspected based on the radiologic findings, and a pancreatoduodenectomy was performed. The removed portion of the pancreas contained a 3.5 × 2 × 1.8 cm well-circumscribed, but not encapsulated white tumor mass with smooth cut surface, cystic component and duct dilatation within the tumor and within the adjacent pancreatic tissue. Based on the histology and immunostaining profile, a diagnosis of the solitary fibrous tumor was made. One week post-operatively, the patient died due to surgical complications. Microscopic and immunohistochemical examinations are necessary for accurate diagnosis of cystic SFT of the pancreas. Because there is limited data regarding the biological behavior of SFT with extra-pleural localization the authors recommend clinical follow-up for SFT treatment if the criteria of malignancy are not met.

**Key words:** pancreas, solitary fibrous tumors

**Introduction**

Pancreas is an extremely rare abdominal localization of the solitary fibrous tumor (SFT) with only 16 cases reported to date [1–16]. It usually grows asymptptomatically for a long time before a diagnosis can be made on the basis of symptoms and/or mechanical complications. Due to the rarity and nonspecific clinical presentation, this entity is diagnostically challenging.

We describe a 47-year-old man with a solitary fibrous tumor arising in the head of the pancreas and review of the diagnostic literature.

**Case report**

A 47-year-old man with a history of progressive epigastric pain for the last two weeks, and jaundice, was admitted to hospital for further investigation. His past medical history revealed a conservative cholecystectomy performed 5 years ago. The family history including malignancy or inherited disease was unremarkable. The physical examination revealed
icteric sclera and yellowish skin. Laboratory investigations showed a normal hemogram: hemoglobin concentration 140 g/L (reference range: 140–180 g/L), hematocrit 40% (reference range: 37–54%), platelet 290 (reference range: 140–340), white blood cell 8000 (reference range: 4000–8000), glucose (ser) 6.1 mmol/L (reference range: 3.5–6.1 mmol/L) alpha amylase (ser) 119 U/L (reference range: 30–110 U/L). Abnormal laboratory findings included elevated LDH 605 U/L (reference range: 248 U/L), total bilirubin 133 umol/L (reference range: 6.8–20.5 umol/L), indir.bilirubin 14 umol/L (reference range: 5.1–13.6 uml/L), direct bilirubin 119 umol/L (reference range: 1.5–6.8 umol/L), and alkaline phosphatase 608 U/L (reference range: 38–126 U/L). The serum tumor markers were within the normal limits for carcinoembryonic antigen 2. 4 ng/mL (reference range: 1–3.4 ng/mL) and increased for carbohydrate antigen 19–9, 198.0 U/ml (reference range: 1–37 U/ml). The abdominal ultrasonography showed a hypoechoic mass, 3.5 cm in cross diameter, located in the pancreatic head, without stones. The computed tomography (CT) imaging of the abdomen confirmed a 3.5 cm mass with enhanced contrast uptake in both arterial and venous phases on the pancreatic head. The tumor mass was well-delimited but not encapsulated, mainly solid but with a cystic component. The surrounding bile duct of the pancreaticoduodenal arcade was dilated. There was no sign of loco-regional invasion or metastasis. The diagnosis of cystadenocarcinoma was suspected and a pancreateoduodenectomy was performed.

The removed portion of the pancreas measured 10 × 4.5 × 3.5 cm and contained a 3.5 × 2 × 1.8 cm well-circumscribed, but not encapsulated white tumor mass with smooth cut surface, cystic component and duct dilatation within the tumor and within adjacent pancreatic tissue [Fig. 1]. The transection margin was tumor-free. The histological analysis showed that tumor had infiltrated the surrounding pancreatic parenchyma and consisted of spindle cells with eosinophilic cytoplasm and hyperchromatic nucleus with minimal cytological atypia, arranged in a fascicular pattern and with branched hemangiopericytoma-like vessels [Fig. 2, 3]. No necrosis was found and mitotic figures were very rare, 1–2 mitosis per 10 high-powered fields. Hyalinization and myxoid degeneration areas were seen in parts, which were hypocellular. The cystic component was related to retention cysts and duct dilatation. The tumor invaded the muscularis propria of the duodenum. The complete pathological examination revealed no vascular or nervous invasion. All seventeen lymph nodes were tumor-free.

**Figure 1** – Solitary, circumscribed, neoplasm with white cut surfaces, in the head of the pancreas

**Figure 2** – Spindle cell with minimal nuclear pleomorphism (HE × 200)

**Figure 3** – Mixoid background and haemangiopericytoma-like vascular pattern (HE × 100)
Immunohistochemical analysis on the resected tumor revealed that the tumor cells express diffusely positive for CD34, vimentin and CD99, focally positive for bcl-2, nuclear beta-catenin and actin and were negative for CD117, EMA, Caldesmon, Desmin, S100 and Cytokeratins [Fig. 4–6]. Ki-67 proliferation index was observed below 1%.

Based on the histology and immunostaining profile, a diagnosis of solitary fibrous tumor was made. One week post-operatively, the patient died due to surgical complications.

Discussion

The 2010 World Health Organization classification defines SFT as a ubiquitous mesenchymal tumor of probable fibroblastic type with a prominent haemangiopericytoma-like branching vascular pattern [15]. SFT was first described as a distinct entity among primary neoplasms in the pleura by Klemperer and Rabin in 1931 [14]. SFT is quite a rare tumor; it accounts for 0.03% of all neoplasms and 3% of soft tissue tumors, and its incidence has been estimated to be 2–4 cases per million per year in the general population. SFT is typically found in the pleura (65%), fascial and musculo-aponeurotic tissues, but can occur in intra-abdominal localization. Intra-abdominal SFTs are most often associated with familial adenomatous polyposis or Gardner syndrome (familial adenomatous polyposis with multiple osteomas and mesenchymal tumors of the skin and soft tissues) in up to 70% of the cases while sporadic cases are uncommon. In contrast to the intra-abdominal forms, sporadic pancreatic fibrous tumors are more frequent than those associated with familial adenomatous polyposis. SFTs in extra-pleural localizations generally exhibit benign behavior [14]. However, it has been reported that several clinical and pathological features can predict more aggressive behavior and metastasis can be seen approximately in 10–15% of tumors [14].

In 65% of SFT cases, the tumor arises from the pleura, but they can also be found in other sites such as lung parenchyma, thyroid gland, liver, kidney, adrenal gland, salivary gland, soft tissue, head and neck. Pancreas is an extremely rare extrapleural location. Including our patient, only 16 cases of pancreatic SFTs have been reported with the clinical findings summarized in Table 1. Other mesenchymal tumors located in the pancreas include GIST, leiomyosarcoma, schwannoma, fibromyxoid sarcoma, perivascular epithelioid cell tumor (PECOoma), and vascular tumors. Pancreatic SFT is often asymptomatic because it is generally a benign and slow-growing tumor. Symptoms differ according to the location and size of the mass and include abdominal pain, constipation, jaundice, and weight loss. In our case, the patient presented with abdominal pain...
and jaundice. The patients’ age ranges from 24–78 years at diagnosis [1–15]. Although extrapleural SFT has no gender bias reported, the current pancreatic cases have female to male ratio of 12 : 3. Our patient was a 47–year-old man. In 10 cases, the radiologic impression favored an endocrine tumor, as these tumors can appear similarly well-circumscribed and hypervascular. In our case, the initial diagnosis was a cystadenocarcinoma. This was suggested because of the cystic component and female gender. In fact, the cystic component corresponded to retention cysts above the tumor. Only 4 published pancreatic SFT presented as cystic tumors. These cases confirm the difficulties of the radiologic diagnosis of the cystic pancreatic tumors. The size of the tumors ranged from 1.5 to 18.5 cm in diameter. SFTs were mostly localized in the head of pancreas (9/13) as in our case. Pancreatic SFT may show a wide range of histological patterns including palisading, diffuse sclerosing areas and stori-form or hemangiopericytic patterns and can thus mimic other mesenchymal neoplastic and non-neoplastic proliferations [1, 4, 7, 8]. Mixed fibrotic, hyalinized and myxoid changes might be found in stroma. The diagnosis of SFT has been refined by the availability and the immunohistochemical markers such as CD34, vimentin, bcl-2, and CD99. Nuclear beta-catenin may occur in approximately one third of SFT.

The differential diagnosis of pancreatic SFTs includes several spindle cell neoplasms such as GIST, leiomyosarcoma, schwannoma, and fibromyxoid sarcoma [10, 11, 14]. Immunohistochemically SFT expresses CD34 and vimentin in 80–90% of cases and CD99 and bcl-2 in 70% of cases. They are usually negative for c-kit (CD117), smooth muscle actin, desmin, S-100 protein and cytokeratins which are markers of GIST, leiomyosarcoma, schwannoma, and fibromyxoid sarcoma, respectively [4, 9]. Diffusely positive staining for CD34, vimentin and CD99, focally positive for bcl-2, nuclear beta-catenin and actin and negative for CD117, EMA, Caldesmon, Desmin, S100 and Cytokeratins has been obtained in our case.

There is limited data regarding the biological behavior of SFTs with extra-pleural localization, because they are rare tumors. Approximately 10–15% of the extra-pleural SFTs are malignant. A study in literature showed local recurrence after 168 months; however, most of the metastasis or a local recurrence was seen within 2 years after treatment. Lung, liver, bone, mesentery, omentum, mediastinum and retroperitoneum were distant metastasis areas in this study [14]. The criteria for malignancy include large tumor size (> 50mm), disseminated disease at presentation, infiltrative margins, and histologic features consistent with high cellularity, nuclear pleomorphism, areas of tumor necrosis, and an increased mitotic index (> 4 mitoses/10 high powered fields) [15]. Malignant SFTs have reduced CD34 immunoreactivity. Relapse was seen in 80% of these cases. However, in our case no pleomorphism and necrosis was found and mitotic figures were very rare, less than 1–2 mitosis per 10 per high-powered fields. Ki-67 proliferation index has been observed below 1% and tumor was diagnosed as a benign.

In the cases of intra-abdominal SFTs, complete tumor resection is the treatment of choice [1, 3, 4, 6, and 7].

In conclusion, we report a rare case of pancreatic SFT. Cystic SFT of the pancreas is difficult to radiologically distinguish from other cystic pancreatic tumors. Microscopic and immunohistochemical studies are necessary for accurate diagnosis. Because there is limited data regarding biological behavior of SFT with extra-pleural localization the authors recommend clinical follow-up for SFT treatment if the criteria of malignancy are not met.
Table 1

Comparative data of patients with solitary fibrous tumors of the pancreas

<table>
<thead>
<tr>
<th>Case</th>
<th>Author</th>
<th>Age</th>
<th>Gender</th>
<th>Clinical presentation</th>
<th>Tumor size (cm)</th>
<th>Location in the pancreas</th>
<th>Immunostaining (+)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Lütgtes 1999</td>
<td>50</td>
<td>Female</td>
<td>Absent/incidental finding</td>
<td>5.5</td>
<td>Body</td>
<td>CD34, CD99, bcl-2, vimentin</td>
</tr>
<tr>
<td>2</td>
<td>Chatti 2006</td>
<td>41</td>
<td>Male</td>
<td>Abdominal pain</td>
<td>13</td>
<td>Body</td>
<td>CD34, CD99, bcl-2, vim</td>
</tr>
<tr>
<td>3</td>
<td>Gardini 2007</td>
<td>62</td>
<td>Female</td>
<td>Abdominal pain</td>
<td>3</td>
<td>Head</td>
<td>CD34, CD99, bcl-2, vim</td>
</tr>
<tr>
<td>4</td>
<td>Miyamoto 2007</td>
<td>41</td>
<td>Female</td>
<td>Right upper quadrant</td>
<td>2</td>
<td>Head/body junction</td>
<td>CD34, bcl-2</td>
</tr>
<tr>
<td>5</td>
<td>Kwon 2008</td>
<td>54</td>
<td>Male</td>
<td>Absent/incidental finding</td>
<td>4.56</td>
<td>Body</td>
<td>CD34, CD99, vim</td>
</tr>
<tr>
<td>6</td>
<td>Srinivasan 2008</td>
<td>78</td>
<td>Female</td>
<td>Back pain, weight loss</td>
<td>5</td>
<td>Body</td>
<td>CD34, CD99, bcl-2, vim</td>
</tr>
<tr>
<td>7</td>
<td>Amiot 2008</td>
<td>51</td>
<td>Female</td>
<td>Epigastric pain</td>
<td>6</td>
<td>Tail</td>
<td>Anti-beta-catenin</td>
</tr>
<tr>
<td>8</td>
<td>Chetty 2009</td>
<td>67</td>
<td>Female</td>
<td>Absent/incidental finding</td>
<td>2.6</td>
<td>Head</td>
<td>CD34, CD99, bcl-2</td>
</tr>
<tr>
<td>9</td>
<td>Ishiwatarii 2009</td>
<td>58</td>
<td>Female</td>
<td>Absent/incidental finding</td>
<td>3</td>
<td>Head</td>
<td>CD34, bcl-2</td>
</tr>
<tr>
<td>10</td>
<td>Sugawara 2010</td>
<td>55</td>
<td>Female</td>
<td>Absent/incidental finding</td>
<td>7</td>
<td>Body</td>
<td>CD34</td>
</tr>
<tr>
<td>11</td>
<td>Santos 2012</td>
<td>40</td>
<td>Female</td>
<td>Absent/incidental finding</td>
<td>3</td>
<td>Body</td>
<td>CD34, β-catenin</td>
</tr>
<tr>
<td>12</td>
<td>Tasdemir 2012</td>
<td>24</td>
<td>Female</td>
<td>Abdominal pain</td>
<td>18.5</td>
<td>Body</td>
<td>CD34, vim</td>
</tr>
<tr>
<td>13</td>
<td>Chen 2013</td>
<td>49</td>
<td>Female</td>
<td>Mild pain in the upper abdomen</td>
<td>13</td>
<td>Head</td>
<td>CD34, bcl-2, vim, muscle-specific actin (MSA), CD68, Ki67</td>
</tr>
<tr>
<td>14</td>
<td>Hwang 2014</td>
<td>53</td>
<td>Female</td>
<td>Absent/incidental finding</td>
<td>5.2 and 1.8</td>
<td>Head</td>
<td>CD34, bcl-2, muscle-specific actin (MSA), CD10, ER, PR</td>
</tr>
<tr>
<td>15</td>
<td>Hee Han 2015</td>
<td>77</td>
<td>Female</td>
<td>Jaundice without other symptoms</td>
<td>1.5</td>
<td>Head</td>
<td>CD34, CD99</td>
</tr>
<tr>
<td>16</td>
<td>Baxter 2015</td>
<td>58</td>
<td>Female</td>
<td>Left lower quadrant</td>
<td>3.5</td>
<td>Head</td>
<td>CD34, bcl-2,</td>
</tr>
<tr>
<td>17</td>
<td>Current case</td>
<td>47</td>
<td>Male</td>
<td>Jaundice, abdominal pain</td>
<td>3.5</td>
<td>Head</td>
<td>CD34, CD99, bcl-2,</td>
</tr>
</tbody>
</table>

REFERENCES

Солитарните фиброзни тумори (СФТ) исключително ретко се јавуваат во панкреасот. Тие обично се развиваат асимптоматски и тоа по-долго време поради долгот период пред да може да се постави дијагноза врз основа на симптомите и/или механички предизвиканите компликации. Поради реткоста и неспецифичната клиничка слика, овој ендитет претставува вистински дијагностички предизвик.

Прикажуваме случај на 47-годишен маж со историја на прогресивна епигастрична болка во тек на две недели и појава на жолтица, кој беше хоспитализиран за понатамошно испитување. Врз основа на радиолошките наоди, поставена е дијагноза на цистаденокарцином, по што е направена панкреатодуоденектомија. Отстапениот дел од панкреасот содржи добро одграничен, неинкапсулиран, бела туморска маса со димензии 3.5 x 2 x 1.8 см. На пресек творбата беше мазна. Видливи беа цистични компоненти и дилатација на каналите во туморското ткиво и во ткиво на панкреасот во неопосредна близина.

За поставување точна дијагноза на цистичен СФТ на панкреас, неопходни се микроскопски и имунохистохемиски испитувања. Податоците за биолошкото однесување на СФТ со екстраплеврална локализација се ограничени, па затоа, и кога не се исполнети критериумите за малигнитет, авторите препорачуваат клиничко следење при третманот на СФТ.