CASE REPORT

Solitary fibrous tumor of the pancreas

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Abstract

Solitary fibrous tumors (SFTs) are rare mesenchymal neoplasms of fibroblastic origin. Most commonly they affect the pleura but they have been described in other viscera. SFT of the pancreas is extremely rare, and only eight cases have been reported to date. We perform a literature review and report a ninth case. The patient is a 54-year-old African-American female who presented with several months of abdominal pain. Abdominal radiography demonstrated a lesion in the head of the pancreas, and she underwent a Whipple operation. Pathology demonstrated SFT of the pancreas. She is alive and well 1 year post-operatively. SFT of the pancreas predominately affects middle-aged women. These tumors are difficult to distinguish radiologically from neuroendocrine tumors. While SFT of the pancreas tend to have an indolent course, there is the potential for malignancy. We recommend complete surgical excision.

INTRODUCTION

Solitary fibrous tumors (SFTs) constitute a group of non-epithelial tumors with spindle cell features. SFTs of the pancreas are exceptionally rare, and only eight cases have been reported to date [1–8]. These tumors present with vague abdominal symptoms or are discovered incidentally on abdominal radiography. Radiologically, they are difficult to distinguish from neuroendocrine tumors. They occur predominantly in middle-aged women and have an indolent course. Herein, we report an additional case of SFT of the pancreas and provide a review of the literature with outcomes and follow-up of cases reported so far.

CASE REPORT

The patient is a 58-year-old African-American female who presented with several months of left lower quadrant pain. A computed tomography (CT) and magnetic resource imaging (MRI) of abdomen/pelvis demonstrated a 3.8-cm hyper vascular mass in the head of the pancreas without any evidence of distant metastasis or spread to the surrounding vasculature (Figs. 1 and 2). The differential diagnosis includes endocrine tumor, gastrointestinal stromal tumor, sarcoma, solid pseudo-papillary tumor, inflammatory pseudotumour/tumor forming autoimmune pancreatitis and SFT. An endoscopic ultrasound with fine-needle aspiration was nondiagnostic. The carcinoembryonic antigen (CEA) and cancer antigen 19-9 (CA 19-9) were both mildly elevated at 6.5 (nl range 0–5.0) and 39 (nl range 0–37), respectively. Functional endocrine hormone studies were significant only for a mildly elevated gastrin level of 214, but the insulin, glucagon and vasoactive intestinal peptide (VIP) levels were all within normal limits. Her review of symptoms was negative for jaundice or weight loss. A Whipple procedure was performed. The patient had an uncomplicated hospital course and was discharged Post-op Day 7. She is alive and well 2 years post-operatively.

The specimen was a 3.5 x 3 x 3-cm white, firm, well-circumscribed mass located in the head of the pancreas (Fig. 3). The mass was found to be compressing the distal aspect of the main pancreatic duct. All 15 lymph nodes received in the specimen were negative. Histologically, the tumor is well circumscribed from normal pancreatic tissue and composed of spindle cells that in some areas form short ill-defined fascicles and in
other areas are randomly arranged in a dense fibrohyaline stroma. A well-developed vascular network was visualized throughout the tumor with vessels of a stag horn appearance with thin
Table 1: Synopsis of the nine cases of SFT of the pancreas reported in the literature, including our own case with the reference, age, sex, type of surgery and follow-up.

<table>
<thead>
<tr>
<th>Sex</th>
<th>Age</th>
<th>Tumor location in the pancreas</th>
<th>Tumor size</th>
<th>Type of Surgery performed</th>
<th>Follow-up</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female</td>
<td>50</td>
<td>Body</td>
<td>5.5 cm</td>
<td>Distal pancreatectomy</td>
<td>Disease free 20 months post-op</td>
<td>Luttges et al.</td>
</tr>
<tr>
<td>Male</td>
<td>41</td>
<td>Body</td>
<td>13 cm</td>
<td>Enucleation</td>
<td>Died Post-op Day 3 from surgical complications</td>
<td>Chatti et al.</td>
</tr>
<tr>
<td>Female</td>
<td>41</td>
<td>Neck</td>
<td>2 cm</td>
<td>Laparoscopic enucleation</td>
<td>Disease free 7 months post-op</td>
<td>Miyamoto et al.</td>
</tr>
<tr>
<td>Female</td>
<td>62</td>
<td>Head</td>
<td>3 cm</td>
<td>Pylorus sparing pancreatectomy</td>
<td>Disease free 16 months post-op</td>
<td>Gardini et al.</td>
</tr>
<tr>
<td>Male</td>
<td>54</td>
<td>Body</td>
<td>7.6 cm</td>
<td>Central Pancreatectomy</td>
<td>Disease free 70 months post-op</td>
<td>Kwon et al.</td>
</tr>
<tr>
<td>Female</td>
<td>78</td>
<td>Body</td>
<td>5 cm</td>
<td>Distal pancreatectomy</td>
<td>Disease free 70 months post-op</td>
<td>Srinivasan et al.</td>
</tr>
<tr>
<td>Female</td>
<td>67</td>
<td>Uncinate process</td>
<td>2.6 cm</td>
<td>Whipple</td>
<td>Alive and well 6 months post-op</td>
<td>Chetty et al.</td>
</tr>
<tr>
<td>Female</td>
<td>55</td>
<td>Head</td>
<td>6 cm</td>
<td>Whipple</td>
<td>Alive and well 3 years post-op</td>
<td>Sugawara</td>
</tr>
<tr>
<td>Female</td>
<td>58</td>
<td>Head</td>
<td>3.3 cm</td>
<td>Whipple</td>
<td>Alive and well 1 year post-op</td>
<td>Current case</td>
</tr>
</tbody>
</table>

These tumors are difficult to distinguish radiologically from endocrine tumors as they appear hypervascular and well circumscribed on CT scan. The mainstay of diagnosis is histological. Particularly, the growth pattern and immunohistochemistry are helpful in differentiating SFTs from other mesenchymal tumors. These tumors tend to demonstrate spindle cells that grow in patternless arrangements with varying amounts of collagen and that stain positive for the cell markers CD34, CD99 and BCL-2.

Surgical excision is the recommended treatment for this rare disease entity, and although there is limited experience with SFTs of the pancreas, it appears that the prognosis is good especially if they have a favorable histological profile on generous tissue sampling and thorough microscopic evaluation. In terms of the extent of surgical resection, complete surgical resection is acceptable. Enucleation has been attempted in only two patients, and one died Post-op Day 3 from surgical complication [1, 10]. No instances of recurrences have been reported in the pancreas thus far; however, the experience is limited, and longer term follow-up is necessary.

CONFLICT OF INTEREST STATEMENT

None declared.

REFERENCES


