CASE REPORT

Malignant Solitary Fibrous Tumor of the Pancreas

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ABSTRACT

Context Solitary fibrous tumor is an uncommon malignancy of spindle cell origin rarely found in the pancreas. A single description of a patient with malignant pancreatic solitary fibrous tumor has previously been published. **Case report** We report an incidentally found malignant solitary fibrous tumor in a 39-year-old woman who presented with low rectal cancer. The tumor was discovered on her rectal cancer staging workup, and imaging studies were suggestive of solid pseudopapillary neoplasm or pancreatic neuroendocrine tumor. Two attempts at endoscopic ultrasound with fine needle aspiration were not diagnostic. The patient underwent successful pancreaticoduodenectomy with resection of the mass and a total of 19 lymph nodes and adequate margins. Final pathology revealed malignant solitary fibrous tumor by histologic examination and immunohistochemistry, with up to six mitoses per 10 high-power fields. **Conclusion** Pancreatic solitary fibrous tumors are themselves extremely rare, with only 15 cases reported in the literature. The majority of tumors are discovered incidentally and are considered benign, but a subset of solitary fibrous tumors carry a malignant phenotype. Data on the appropriate management and treatment for malignant solitary fibrous tumor is scarce. However, given their potential for recurrence, lifelong surveillance is recommended.

INTRODUCTION

Solitary fibrous tumors, though rare, have been reported in a variety of tissues throughout the body. These tumors are predominantly benign lesions of spindle cell origin that typically arise in the chest wall. Extrathoracic solitary fibrous tumors have been found in the sigmoid mesentery [1], the orbit, the salivary glands [2], the kidney [3], and the pancreas. Pancreatic solitary fibrous tumor is an exceedingly rare occurrence, with a recent literature review tallying only 15 cases reported [4].

Malignancy in these tumors is uncommon [5, 6]. The majority of the reported cases are intrathoracic, though malignant solitary fibrous tumor has also been found in the retroperitoneum, extremities, kidney, and, once, in the pancreas [7]. Given the rarity of these tumors, treatment and surveillance recommendations are not clear, especially in young patients. We present a patient with a malignant solitary fibrous tumor arising from the mesenchyme of the pancreatic head, as well as a review of the literature regarding diagnosis, management, and follow-up.

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Abbreviations EUS Endoscopic ultrasound
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CASE REPORT

A thirty-nine-year-old woman with no significant past medical history presented to her primary care physician with persistent rectal bleeding. The patient reported three to four years of small amounts of rectal bleeding with a recent marked increase, but denied any nausea, vomiting, dysphagia, new reflux disease, abdominal pain, jaundice, or new-onset diabetes or exocrine dysfunction. Her laboratory results were within the range of normal. Endoscopic investigation revealed a 25-30% circumferential low rectal moderately differentiated adenocarcinoma on biopsy, and staging workup with CT and MRI was undertaken. Her rectal cancer was found to be T2N0M0, stage I. Her staging CT abdomen and pelvis, performed with IV contrast, revealed an incidental 1.8 cm × 1.2 cm well-circumscribed mass in the pancreatic head **(Figure 1a)**. The mass showed central hypodensity with contrast uptake in the surrounding rim, suspicious for a solid pseudopapillary neoplasm or a pancreatic neuroendocrine tumor, as suggested on MRI **(Figure 1b)**. Endoscopic ultrasound was performed with fine needle aspiration (FNA); histologic findings from this specimen were limited by hypocellularity of the sample but showed rare scattered oval to spindle cells with no sign of malignancy. A repeat endoscopic ultrasound with FNA showed only scattered normal pancreatic neuroendocrine cells and fragments of duodenal mucosa.

Due to the patient's concomitant rectal cancer, the decision was made to postpone pancreatic resection until after the necessary abdominoperineal resection (APR)

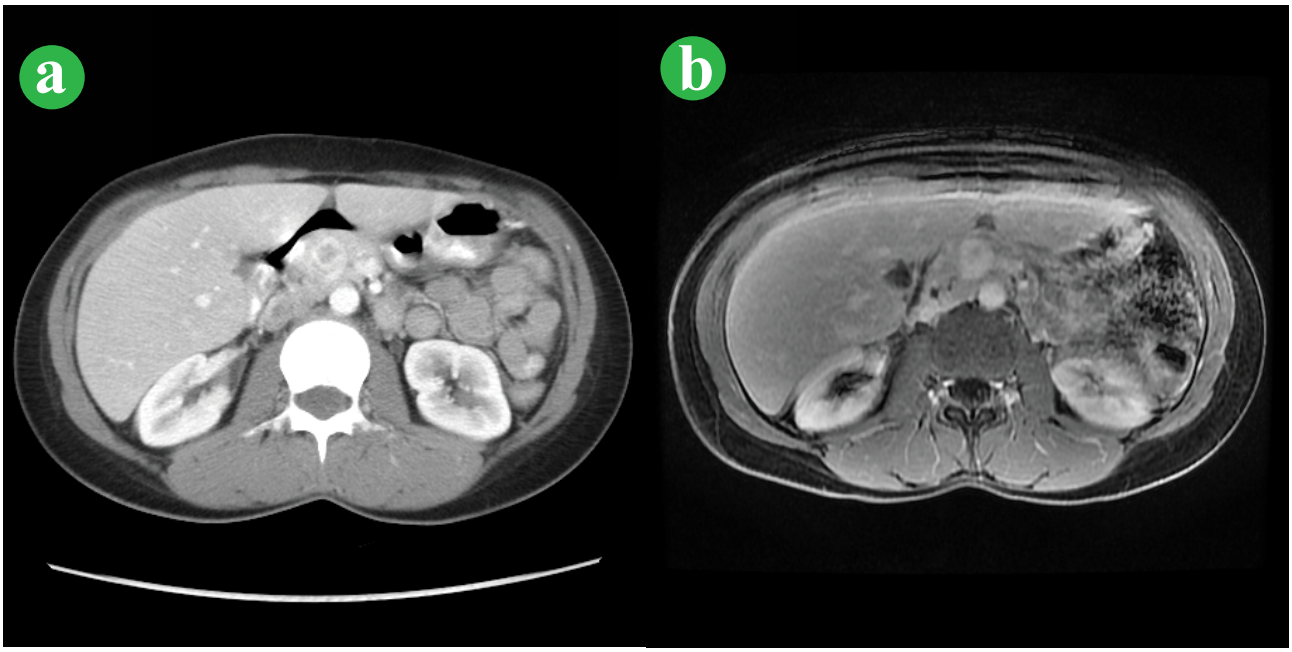

Figure 1. (a). Computed tomography of the abdomen with intravenous contrast showing 1.8 cm × 1.2 cm well-circumscribed mass in the pancreatic head. **(b).** Magnetic resonance imaging of the abdomen showing pancreatic head mass at 2 cm × 1.7 cm with central hypoenhancement.

and convalescence. The patient underwent APR six weeks after her initial diagnosis, and returned for consideration of resection of her pancreatic tumor six months later. The final stage of her rectal cancer was T2N0M0, and she was not recommended for adjuvant therapy. She had undergone extensive genetic testing for possible mutations, including Lynch syndrome, that might be the source of her multiple lesions, but this was negative. The patient was counseled on the risks of continued surveillance versus resection of the pancreas lesion given the lack of a clear histologic diagnosis, she opted for surgical management.

The patient was taken to surgery and a pylorus-preserving pancreaticoduodenectomy was performed. The mass was well-circumscribed and was contained entirely within the pancreas on intraoperative examination. On gross examination, the specimen measured 6.0 cm × 5.8 cm × 3.8 cm and contained a well-circumscribed, slightly bulging tan mass measuring 2.2 cm × 2.2 cm × 1.8 cm, abutting the portal vein groove and 1.0 cm from the pancreatic resection margin **(Figure 2a)**. A total of 19 lymph nodes were found within the specimen, all negative for metastatic disease. On histologic examination, the tumor was found to be a spindle cell neoplasm comprised of pattern-less infiltrate of round to spindle-shaped tumor cells in a fibrous background, without necrosis or vascular invasion **(Figure 2b)**. Mitoses measured up to six per 10 high-power fields, and immunohistochemical studies showed active expression of CD34, BCL2, CD56, nuclear STAT6, cytokeratin CAM5.2 and variable expression of AE1/AE3, and no expression of S-100, CD117, CDX2, desmin, SMA, ER, MNF116, CK7, CK20, synaptophysin, or chromogranin **(Figures 2c,2d)**. Based on histologic and immunochemical findings, a diagnosis of malignant solitary fibrous tumor was suggested. The specimen was sent for outside consultation given the rarity of this diagnosis, and a separate institutional review of the slides confirmed that the appearance was "undoubtedly (that) of a malignant solitary fibrous tumor".

The patient recovered well from the operation, and one month post-operatively is disease- and symptom-free. Her abdominal imaging surveillance for her malignant solitary fibrous tumor will be combined with her necessary pelvic imaging surveillance for her rectal cancer.

DISCUSSION

Asymptomatic pancreatic lesions, or pancreatic incidentalomas, are an increasingly common finding in the age of easily available cross-sectional imaging [8, 9]. Defined as asymptomatic pancreatic masses discovered on radiologic, endoscopic, or laboratory evaluation for unrelated conditions, these lesions are most commonly discovered during evaluation of the genitourinary/renal system, during evaluation for elevated LFTs, or during staging, screening, or surveillance for non-pancreatic neoplasm [10, 11, 12]. Of asymptomatic pancreatic lesions referred to tertiary care centers for management, 5-45% percent were discovered on cancer screening or surveillance [13]. Although the most likely diagnosis for symptomatic primary pancreatic tumor is adenocarcinoma, the differential diagnosis for asymptomatic pancreatic lesions is extremely broad, including papillary cystic and solid tumor, solid pseudopapillary tumor, serous cystadenoma, mucinous cystadenoma or cystadenocarcinoma, pancreatic neuroendocrine tumor, and intraductal papillary mucinous neoplasm [14, 15, 16, 17, 18, 19]. In the majority of studies, patients presenting with asymptomatic pancreatic lesions were relatively young without significant risk factors [20]. In one study of asymptomatic pancreatic lesions treated with resection, six percent of patients had more

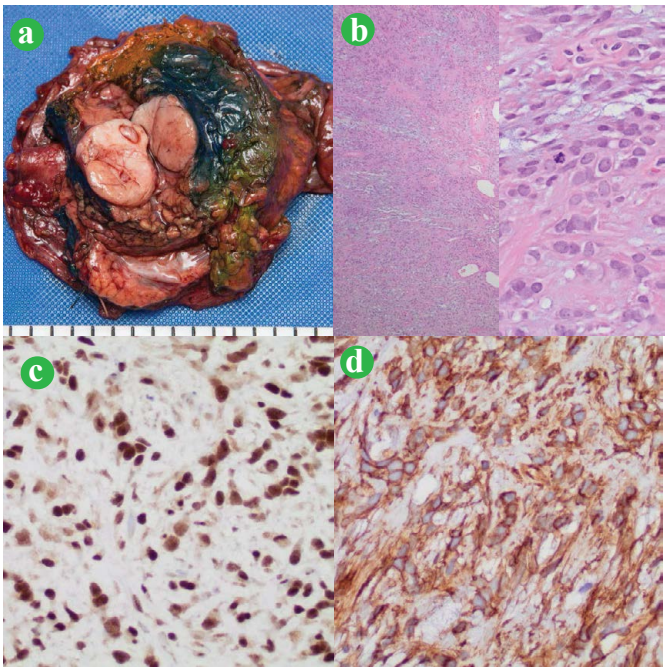

Figure 2. (a). Pancreatic resection specimen, bisected, containing a well-circumscribed, slightly bulging tan mass measuring 2.2 cm × 2.2 cm × 1.8 cm, abutting the portal vein groove and 1.0 cm from the pancreatic resection margin. **(b).** Hemotoxylin and eosin stain of the tumor specimen revealed spindle cell neoplasm comprised of patternless infiltrate of round to spindle-shaped tumor cells in a fibrous background, without necrosis or vascular invasion, but with up to six mitoses per 10 high-power fields (40x, 400x). **(c).** STAT6 immunohistochemical stain of the cell block specimen with tumor cells that have strong positivity in the nucleus (400x). **(d).** CD34 immunohistochemical stain of the cell block specimen with tumor cells that stain diffusely positive (400x).

than one distinct pathology on final histologic diagnosis, and 12% had no identified pancreatic lesion on pathology [11]. Endoscopic ultrasound and tissue biopsy is often sought preoperatively in order to better characterize asymptomatic pancreatic lesions, though this testing does not always prove diagnostic [8, 10, 20]. In our patient, the differential diagnosis was narrowed marginally by the distinctive imaging characteristics and FNA biopsy results, which indicated a possible solid pseudopapillary tumor versus a nonfunctional pancreatic neuroendocrine tumor. Both lesions carry a low but concerning malignant potential, and resection was indicated despite lack of a clear histologic diagnosis preoperatively [14, 17, 21].

Mesenchymal tumors of the pancreas, such as lymphangioma, liposarcoma, schwannoma, malignant fibrous histocytoma, and malignant and benign solitary fibrous tumors, are extremely rare [22, 23]. Accurate diagnosis of malignant solitary fibrous tumor on FNA is difficult, particularly considering that immunohistochemical markers such as CD34 expression, which help guide diagnosis of benign solitary fibrous tumor, are often lost in malignant degeneration. In a single case series of 11 malignant solitary fibrous tumor, none of the cases were correctly preoperatively diagnosed on FNA [6].

Only one other instance of a malignant solitary fibrous tumor of the pancreas has been reported; this presented symptomatically with biliary obstruction from a 12 cm tumor confirmed on imaging, and no preoperative tissue diagnosis was attempted [7]. Histology showed heterogenous fibrous tumor, in places typical of a benign solitary fibrous tumor, and in places appearing overtly sarcomatous with a moderate to marked degree of nuclear pleomorphism and 5-17 mitoses per 10 high-power fields. Twenty-four regional lymph nodes and all margins were free of tumor, and the patient was well with no evidence of disease at 40 months after resection.

Although malignant solitary fibrous tumor are characterized by high rates of mitosis per high-power fields and are considered at risk of recurrence and metastatic spread, there is evidence from intrathoracic case series that certain gross and histologic characteristics of the tumor predict a less-aggressive pattern of tumor behavior [5, 24]. In malignant solitary fibrous tumor of the pleura, cases of "grossly polypoid, well-circumscribed, and completely resectable" tumors with 10 or fewer mitoses per 10 high-power fields were less aggressive and had outcomes comparable to benign solitary fibrous tumors, with very low likelihood of recurrence and metastases [3]. Although there is sparse data on extrathoracic malignant and benign solitary fibrous tumor, they appear to behave similarly, apart from a slightly increased risk of local recurrence compared to intrathoracic solitary fibrous tumors [25]. However, case reports exist of malignant solitary fibrous tumor recurring years to decades from initial resection, and long-term surveillance imaging is recommended [7, 26]. There is no clear consensus on the optimal timing and modality for surveillance imaging, and some consideration must be given to minimizing risk of cumulative ionizing radiation from repeat CT scan. In our patient, her concurrent rectal cancer will require long-term surveillance, and therefore imaging will be combined as much as feasible.

In summary, we report the second patient with malignant solitary fibrous tumor of the pancreas in the English literature. Extrathoracic malignant solitary fibrous tumor is difficult to diagnose preoperatively despite imaging and advanced biopsy techniques. The risk of malignancy in incidentally found asymptomatic pancreatic lesions is low but cannot be dismissed, especially in young patients in whom long-term watchful waiting would put them at risk for malignant degeneration or spread. Providers must maintain a high index of suspicion for potential malignancy when confronted with incidentally found asymptomatic pancreatic lesions. Fine needle aspiration is an appropriate initial diagnostic test, but inability to make a preoperative histologic diagnosis of malignancy should not preclude operative intervention when imaging findings are concerning. Gross, histologic, and immunohistochemical examination are necessary to characterize extrathoracic malignant solitary fibrous tumor, and histologic findings may help predict the risk of recurrence or spread. All patients should be followed with long-term surveillance.

Conflict of Interest

The authors declare that they have no conflict of interest.

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