Solitary Fibrous Tumor of the Pancreas: A Case Report and Review Literature

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Abstract

Solitary fibrous tumor represents a single spectrum of mesenchymal tumors with fibroblastic differentiation. The pleura is the first organ that tumors originate and occur most commonly in the thoracic cavity. Solitary fibrous tumor of pancreas is extremely rare and slow growing with asymptomatic. We report the case of a patient with a primary solitary fibrous tumor of the pancreas with a review of the literature. This case is a 56-year-old woman, presented with right upper abdominal pain for 1 month. Whole abdominal CT scan revealed a well-defined hypervascular mass at right upper abdominal region, sized 6.3x5.9x5.7 cm, abutting duodenum and pancreas. The gross specimen displayed a well circumscribed solid mass at pancreatic head, measuring 7.5x6.5x6.5cm. On histomorphology, the mass comprises bland-looking spindle cell neoplasm. Immunohistochemical studies showed positive vimentin, Bcl-2, CD99, CD34 and STAT-6 compatible with solitary fibrous tumor. The patient had been still alive 1 year after surgery.

Keyword; Solitary fibrous tumor, pancreas, mesenchymal tumor

เนื้องอก Solitary fibrous tumor ของตับอ่อน: รายงานผู้ป่วยพร้อมทบทวนวรรณกรรม

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บทคัดย่อ

เนื้องอกโซลิทารี่ไฟบรัส (solitary fibrous tumor) เป็นเนื้องอกในกลุ่มเนื้อเยื่อเกี่ยวพัน ที่จัดอยู่ในกลุ่มเนื้องอกของไฟโบรบลาสต์ เยื่อหุ้มปอดเป็นอวัยวะแรกๆที่พบและพบเนื้องอกชนิดนี้ได้บ่อยที่สุดในช่องทรวงอก เนื้องอกในตับอ่อนนั้นถือว่าเกิดขึ้นได้ไม่บ่อย และส่วนใหญ่ผู้ป่วยมักไม่มีอาการ รายงานนี้จะรายงานผู้ป่วยที่พบเนื้องอกโซลิทารี่ไฟบรัสในตับอ่อนพร้อมทั้งทบทวนวรรณกรรม ผู้ป่วยเป็นหญิงอายุ 56 ปี มีประวัติปวดท้องด้านขวาบนมาเป็นเวลา 1 เดือน พบก้อนขอบเขตชัดเจนเมื่อตรวจด้วยเอกซเรย์คอมพิวเตอร์ มีหลอดเลือดจำนวนมากในก้อน ที่บริเวณด้านขวาบนของช่องท้อง ขนาด 6.3x5.9x5.7เซนติเมตร บริเวณติดกับลำไส้เล็กส่วนต้น (duodenum) และตับอ่อน ชิ้นเนื้อพบเป็นก้อนเนื้องอกขอบเขตชัดเจน ขนาด 7.5x6.5x6.5 เซนติเมตรบริเวณส่วนหัวของตับอ่อน ทางกล้องจุลทรรศน์พบเนื้องอกเป็นเซลล์รูปกระสวย การตรวจทาง immunohistochemistry พบเนื้องอกมีการแสดงออกของ vimentin, Bcl-2, CD99, CD34 และ STAT-6 จึงให้การวินิจฉัยเป็น solitary fibrous tumor ผู้ป่วยยังมีชีวิตอยู่หลังจากผ่าตัดได้ 1 ปี

Introduction

Extrapulmonary solitary fibrous tumor (SFT) is a mesenchymal tumor that exhibits fibroblastic-type differentiation, which shows a prominent hemangiopericytoma-like branching vascular pattern. In the past, most cases were termed hemangiopericytomas[1]. The tumor typically affects in middle-aged adult aged 20-70 years[1] and are found in any location; 28% found in abdomen/pelvis, 16% in extremity and 11% in head and neck [2]. In pancreas, tumor is extremely rare to be found and in past 18 years, there are only 19 cases that have been reported[3-21]. We report additional case of SFT in pancreas and review of the literature.

Case report

A 56-year old woman had right upper abdominal pain for 1 month. The pain was mild to moderate requiring no analgesia. It was persistent without alleviation or aggravation by any factors. No other associated symptoms were noted.

Physical examination revealed a moderate degree of tenderness over right upper quadrant of abdomen without any masses or other abnormal findings. Except for a marginal high fasting blood sugar of 120 mg/dl, other laboratory investigations of complete blood count, blood chemistry, and electrolytes were within normal limit. With a provisional diagnosis of chronic cholecystitis, whole abdominal CT scan was performed which revealed a well-defined hypervascular mass at right upper abdominal region, sized 6.3x5.9x5.7 cm, abutting duodenum and uncinate process of pancreas without a separate fat plane. No internal calcification or hemorrhage of the mass was seen. There was no regional lymph node enlargement. The radiologic diagnoses were neuroendocrine tumor, gastrointestinal stromal tumor, or metastatic cancer.

Whipple operation and cholecystectomy were successfully done. The specimens from Whipple procedure included whole duodenal C-loop, part of jejunum and pancreatic head with a well circumscribed solid mass at pancreatic head, measuring 7.5x6.5x6.5cm (Fig. 1A). Cut surface of the mass appeared fibrous solid-hard tumor at pancreatic head leaving only thin layer of normal pancreatic tissue. Hemorrhage and necrosis are not present (Fig. 1B). No involvement of bowel is identified. The gallbladder stone sized 4.5x2.5x2.5 cm was found occupying nearly the entire luminal cavity with thickening of gallbladder wall.

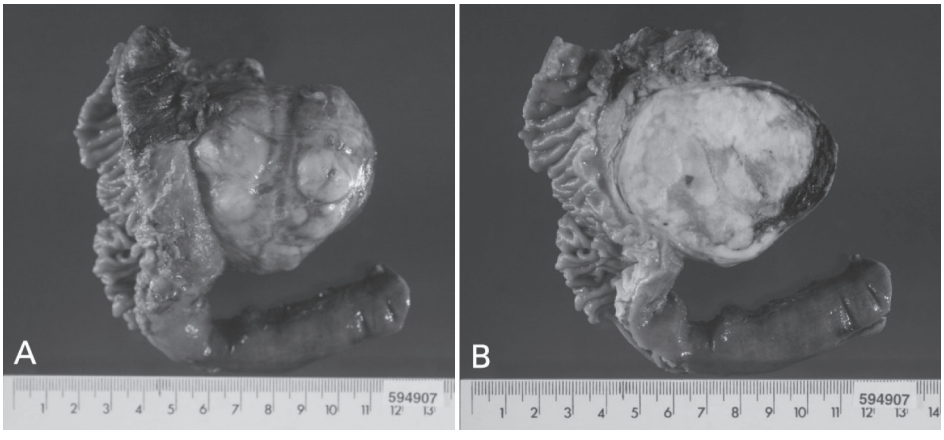

Figure 1: Gross appearance A, Solid grey-white and pale brown mass, in head of pancreas. B, Cut surface showed fibrous solid -hard and grey-white tumor at pancreatic head leaving only thin layer of normal pancreatic tissue. No necrosis is observed

Histologically, the pancreatic tumor was well circumscribed with focal entrapped pancreatic tissue (Fig. 2A). Proliferation of staghorn blood vessels were frequently seen (Fig. 2B) in collagenous stroma (Fig. 2C). The tumor was composed of spindle-shape cells arranging in interlacing bundles with minimally pleomorphic nuclei and fine chromatin (Fig. 2D). Mitosis was rarely observed, 1/10HPFs. Immunohistochemical studies were compatible with fibroblastic origin with positive vimentin, Bcl-2, CD99, CD34 and STAT-6 (Fig. 3A-D) and negative AE1/AE3, EMA, SMA, S-100, desmin and CD117.

She had postoperative febrile illness with intra-abdominal fluid collection from pancreaticoduodenal fluid leakage, lung atelectasis, and surgical site infection. The conditions were resolved with antibiotics and conservative treatment. Other incidental event was delirium which was determined to be irrelevant to her tumor. The patient had been alive 1 year after surgery.

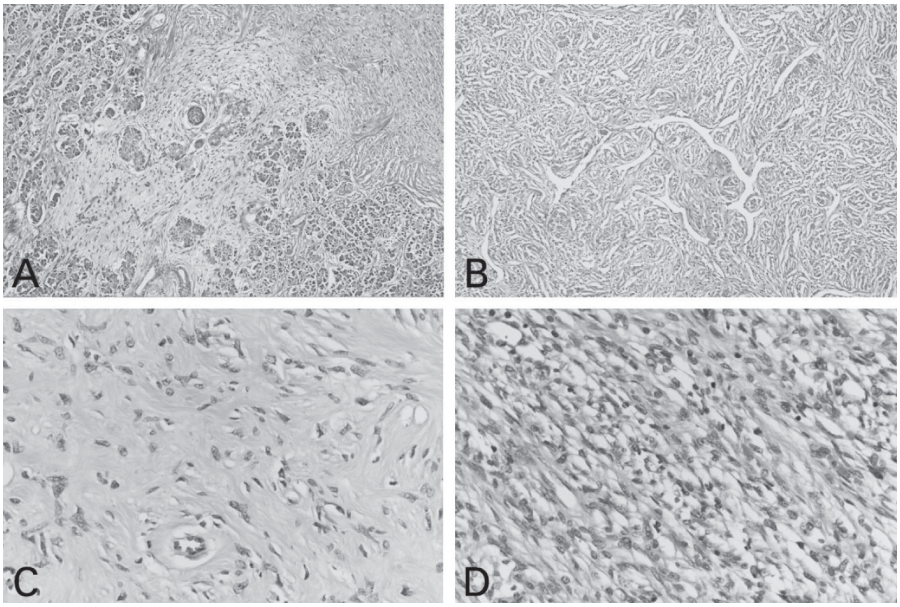

Figure 2: Histomorphology A, Rimming of the tumor and normal pancreatic tissue, the tumor entrapping normal pancreatic tissue (Hematoxylin-eosin, ob. x10). B, Increased vascularity with staghorn blood vessels (Hematoxylin-eosin, ob. x10). C, Abundant collagenous stroma (Hematoxylin-eosin, ob. X40). D, The tumor composed of spindle cells that are arranged in interlacing fascicular pattern. The tumor cells have minimal atypical nuclei, fine chromatin and eosinophilic cytoplasm (Hematoxylin-eosin, ob. X40).

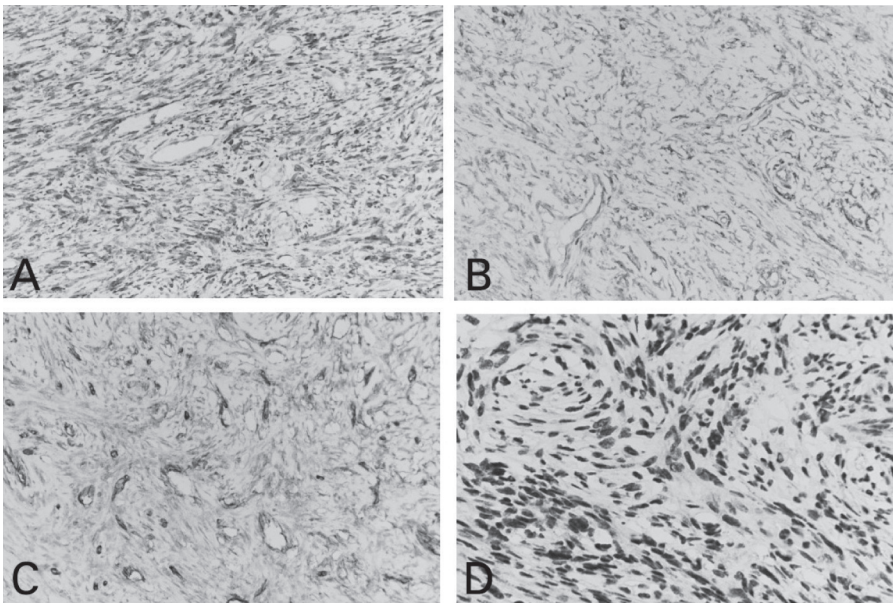

Figure 2: Immunohistochemical studies in solitary fibrous tumor. A, Nuclear and cytoplasmic expression of Bcl-2 (Immunohistochemistry, ob. X20). B, Cytoplasmic expression of CD99 (Immunohistochemistry, ob. X20). C, Cytoplasmic expression of CD34 (Immunohistochemistry, ob. X20). D, Nuclear expression of STAT-6 (Immunohistochemistry, ob. X40).

Discussion

Primary mesenchymal tumors of the pancreas are rare comprising only 0.3% of surgically resected or biopsy-examined pancreatic tumor[19]. Majority of the mesenchymal tumors were benign or borderline and only about one third were sarcomas. Histopathology of can mimic schwannomas, inflammatory myofibroblastic tumor, solid-cystic hamartoma, fibromatoses, cavernous hemangiomas, angiomyolipoma, solitary fibrous tumors, etc.[19].

SFT, a tumor of fibroblastic origin, is formerly known as hemangiopericytoma due to its prominent branching hemangiopericytoma-like vascular pattern[20]. It is more common at head and neck regions and is extremely rare at pancreas [21]. Although 70% of intra-abdominal SFTs are associated with familial adenomatous polyposis or Gardner syndrome (familial adenomatous polyposis with multiple osteomas and mesenchymal tumors of the skin and soft tissues), most pancreatic SFT are sporadic in nature[15]. Our patient had no history of underlying disease or family history of tumors. Hence, we assumed that our case was sporadic in nature. The World Health Organization reported extrathoracic SFT (without specific site of origin) were more common in male compared to female (ratio of 3:2), with have age incidence range from 20-70 years old [20]. From our literature review including our patient, focusing only to pancreatic SFT, we found age ranged from 1-82 years (median 54 years) with more numbers in female than male.

Most patients with extrathoracic SFTs presented with mass at the initial evaluation. Other symptoms were abdominal pain, change in bowel habit, and lower urinary tract symptoms from pressure effects. In one patient a pelvic mass is found incidentally during a routine pelvic examination[22]. In pancreas, no difference of symptoms among SFTs at different parts of pancreas (head, body, or tail) is observed (Table 1). Hypoglycemia has been reported in about 5% of SFTs, particularly in malignant SFTs, SFT associated with Doege-Potter syndrome, and in tumors located in the pelvis and retroperitoneum[23]. The secretion of non-suppressible insulin-like active substances and insulin-like growth factors from the tumor is assumed[24]. Our patient, on the contrary, showed an evidence of impaired glucose tolerance (120 mg/dl). This might be due to her pre-existing diabetes mellitus and a rapid diagnosis and treatment after one month of her symptom.

Pre-operative imaging study generally shows a circumscribed and hypervascularized mass leading to a radiologic impression of an endocrine tumor [22]. The whole abdominal CT scan performed in our patient had these features.

From our review, gross findings of the pancreatic SFT were well-circumscribed with or without capsule and may range from 2.0 to 18.5 cm (median 3.5cm; generally larger in body of pancreas) (Table 1). The mass is firm and elastic with variegated cut surfaces of greyish white, yellowish brown or yellowish- beige with occasional hemorrhage and cystic change in some cases. These features would help to differentiate SFT from malignant tumors which frequently have infiltrative margins and necrosis [25, 26, 27].

Histomorphology of SFT is variable, ranging from a paucicellular to a moderate to highly cellular tumor (table 2). Tumor cells are round to spindle with little cytoplasm, arranging in a haphazard pattern or short storiform or fascicular pattern along with thin-walled staghorn branching vessels or hemangiopericytoma-like pattern. The tumor matrix frequently showed collagenous, hyalinized, fibrous, and keloid-like of collagen bundles. Artifactual 'cracks' between the cells and collagen were observed [24]. Metaplastic ossification was rarely reported [24]. Atypical histological features, such as, increased cellularity, nuclear pleomorphism and atypia, necrosis and mitotic figure of 4 or more per 10 high-power fields, and stromal or vascular invasion were associated with clinically malignant behavior [10, 28,29,20,31].

Immunohistochemical study may help in any cases with suspicious diagnosis. From a literature review and as found in our case, 95-100% of SFTs were positive for CD34 [32] and 50-100% for Bcl-2 [23, 32]. In combination of positive CD34 and Bcl2, almost all cases of SFT can be diagnosed with certainty. The only exception to this is malignant and dedifferentiated cases of SFT in which the percentage of CD34 positivity may be lower [23, 32]. Other immunohistochemcal studies which had been reported are shown in (Table 3).

Table 1:

Clinical presentation of pancreatic solitary fibrous tumor (N=19)

Part of pancreas	N	Symptoms (n)	Size in cm, range (median)	Reference
Head	9	Yes (4), No (5)	2-7 (3.0)	4, 8, 7, 9, 12,13,15,18, our patient
Body	7	Yes (2), No (5)	2-13 (5)	1, 2, 5, 6, 11, 16, 17
Tail	1	Yes (0), No (1)	2.1	14
Head and body junction	1	Yes (1), No (0)	2.0	3
Unidentified location	1	Yes (1), No (0)	18.5	10

Table 2:

Histopathologic features of pancreatic solitary fibrous tumor (N=19)

Tumor morphology	N	Reference
Biphasic cellularity	6	1, 2, 7, 10, 14, 16
Patternless (haphazard)	6	1, 3, 5, 12, 14, 17
Storiform/fascicular pattern	7	1, 8, 9, 13, 15, 16, our patient
Stromal appearance		
Collageneous/ hyalinized/ fibrous/ keloid-like	14	1, 2, 3, 5, 6, 7, 8, 10, 12, 13, 14, 17, 18, our patient
Metaplastic ossification	1	12
Myxoid degeneration	3	10, 15, 16
Vascular pattern/change		
Hemangiopericytoma-like/ stag horn blood vessels	7	2, 5, 9, 12, 13, 15, 17, our patient
Hyalinized vascular wall	1	10
Mitosis		
No	6	2, 3, 7, 11, 13, 14
1-2/10HPF*	6	5, 10, 15, 16, 17, our patient
2-5/10HPF*	1	18
Not mentioned	4	1, 6, 9, 12, our patient
Necrosis		
Presence	0	
Absence	9	3, 5, 7, 11, 13, 14,15, 18, our patient
Not mentioned	8	1, 2, 6, 9, 10, 12,16, 17
Tumor involvement		
No	7	5, 6, 11, 12, 13, 14, 16
Pancreatic tissue	7	1, 3, 7, 9, 10, 15, our patient
Neighboring viscera (duodenum)	2	15
Note mentioned	2	2, 17

*HPF = High power flied

Table 3:

Immunohistochemical expression in pancreatic solitary fibrous tumor (N=19)

Immunohistochemical expression	N	Reference
Highly suggestive SFT		
CD34	18	1, 2, 3, 4, 5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16, 17, 18
STAT-6	2	14, 17
Suggestive		
Bcl-2	12	2, 3, 4, 5, 7, 8, 10, 12, 13, 14, 15, 16
CD99	10	2, 4, 5, 6, 7, 12, 13, 14, 15, 16
Occasional		
Beta-catenin	3	10, 11, 13
Smooth muscle actin	4	2 (focally), 4, 12, 18
CD117	1	2 (focally)
CD10	1	12
ER	1	12
PR	1	12

Surgical resection remains the gold standard of treatment [15, 18]. Pancreatic surgery could generate significant postoperative morbidity, especially when the location is at pancreatic head, therefore, a Whipple procedure to remove them in toto is required [17]. Few deaths were reported within a few days to a week after surgery [4, 15]. Others reported disease-free survivals ranging from 6 to 24 months (median 11-12 months) [1,3,4,5,7,13,14,16,17,18].

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