CASE REPORT

Pancreatic Metastasis from a Solitary Fibrous Tumor of the Central Nervous System

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ABSTRACT

Context Solitary fibrous tumor of the central nervous system is uncommon, with only around 200 reported cases. Further, extracranial metastasis is extremely rare, and only 5 cases of hematogenous metastases have been reported so far. To the best of our knowledge, there have been no reports of solitary fibrous tumor of the central nervous system metastasizing to the pancreas. **Case report** A 62-year-old woman was referred for evaluation of a pancreatic mass, which was strongly suspected to be a neuroendocrine tumor. However, the histological findings and immunohistochemical profile indicated the presence of a solitary fibrous tumor. Because the medical history revealed previous transcranial resection for intracranial meningioma 16 years ago, we conducted a pathological review of the brain specimen obtained by the first operation and found that it had the same histology and immunohistochemical profile as the current endoscopic ultrasound-guided fine-needle aspiration specimen. Consequently, the final diagnosis, on the basis of the brain specimen, was changed from meningioma to solitary fibrous tumor of the central nervous system, and the pancreatic mass was diagnosed as metastasis from solitary fibrous tumor of the central nervous system. The patient underwent middle pancreatectomy; the pancreatic specimen also had the same histology and immunohistochemical profile as the brain specimen. **Conclusion** Histological findings and immunohistochemical profile obtained by EUS-FNA are invaluable for the correct diagnosis to avoid excessive surgical procedures.

INTRODUCTION

Solitary fibrous tumor of the central nervous system was firstly reported by Carneiro *et al*. in 1996 [1] and subsequent reports are limited, with around 200 described cases [2, 3, 4], of which only five extracranial hematogenous metastases including lung, liver, bone, soft tissue, and a combination of two or more regions have been reported thus far [5, 6, 7, 8, 9] (Table 1). We here report a case of pancreatic metastasis from solitary fibrous tumor of the central nervous system diagnosed by endoscopic ultrasound-guided fine needle aspiration (EUS-FNA).

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Key words Endoscopic Ultrasound-Guided Fine Needle Aspiration; Immunochemistry; Neoplasm Metastasis; Pancreas; Solitary Fibrous Tumors

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CASE REPORT

A 62-year-old woman had chest discomfort and was examined by computed tomography (CT) with knowledge of her history of coronary heart disease. There were no findings of coronary arterial stenosis; however, a pancreatic mass with hypervascularity was incidentally detected, and she was subsequently referred to our hospital for evaluation of the mass. She had no clinical symptoms related to pancreas or hormonal hypersecretion. Her medical history included surgical resection for intracranial meningioma 16 years ago and γ-knife radiosurgery for intracranial recurrence 9 and 4 years ago. Blood examination upon admission showed normal serum levels of amylase (47 IU/L; reference range: 37-125 IU/L) and lipase (5 IU/L; reference range: 11-53 IU/L). Fasting blood sugar (206 mg/dL; reference range: 70-109 mg/dL) and glycated hemoglobin (6.6%; reference range: 4.6-6.2%) were increased, as she had previously been diagnosed with diabetes. Serum levels of tumor markers, including carcinoembryonic antigen and carbohydrate

Table 1. Previous reports of extracranial hematogenous metastases of solitary fibrous tumor of the central nervous system.

Case	Author	Year	Age (years)	Sex	Primary site	Diameter of primary site (cm)	Treatment for primary site	Resected margin	Metastatic site	Time to metastasis (years)
#1	Ng *et al*. [5]	2000	55	Female	Intracranial	4.5	Resection, radiation	Negative	Soft tissue of the neck, chest wall, and lung	9
#2	Kim *et al*. [6]	2004	55	Male	Intracranial	NS	Resection	NS	Liver, lung, and ribs	7
#3	Ogawa *et al*. [7]	2004	44	Female	Intracranial	3	Resection	NS	Lung	23
#4	Metellus *et al*. [8]	2007	34	Male	Intracranial	NS	Resection, radiation	Positive	Systemic (Otherwise not specified)	12
#5	Muñoz *et al*. [9]	2008	35	Male	Intraspinal	5.2	Resection	Negative	Lung and liver	4
#6	Present case	2013	46	Female	Intracranial	4	Resection, radiation	Positive	Pancreas	16

NS: not stated in the original articles by the reporting authors

antigenic 19-9 determinant were within normal ranges. Abdominal contrast enhanced CT revealed a mass, 3.3 cm in diameter, with main pancreatic duct stenosis at the junction of the head and body of the pancreas (Figure 1a). Magnetic resonance imaging and cholangiopancreatography clearly showed cystic degeneration in the central portion of the mass and pancreatic duct stenosis due to the mass (Figure 1bcd). At that time, the lesion was suspected as non-functioning neuroendocrine tumor (NET) of the pancreas with indication of possible malignancy; therefore, pancreaticoduodenectomy with lymph node dissection was scheduled. For definitive diagnosis, the patient underwent EUS-FNA using a 22-gauge Expect™ needle (Boston Scientific Japan, Tokyo, Japan) (Figure 1ef). However, pathological examination of the EUS-FNA specimen showed an unexpected finding: there were a large number of elongated cells with collagenous fibers that were positive for CD34,

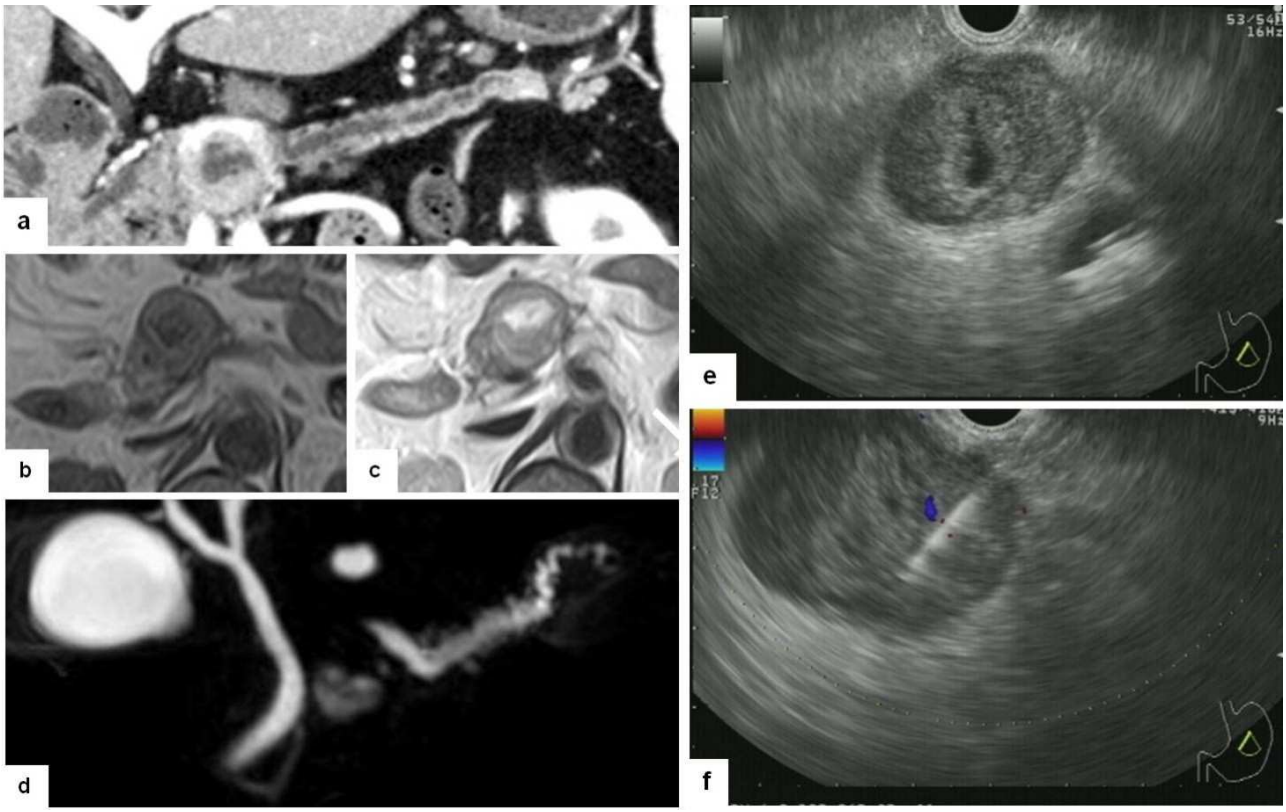

Figure 1. Findings of image examinations. Curved planar reformation of computed tomography made by tracing the main pancreatic duct showed a well-defined mass of the pancreas with strong enhancement, stenosis of the main pancreatic duct, and central cystic change (**a.**). Magnetic resonance imaging showed an iso-intensity mass with a central cystic portion and capsule like rim on (**b.**, **c.**). MR cholangiopancreatography clearly showed cystic degeneration in the central portion of the mass and pancreatic duct stenosis due to the mass (**d.**). Endoscopic ultrasound demonstrated a well-demarcated hypoechoic mass with a central hyperechoic area, which indicated intratumoral hemorrhage or degeneration (**e.**). Fine needle aspiration was performed using a 22-gauge needle (**f.**).

CD99, and bcl-2, but negative for chromogranin A, synaptophysin, and epithelial membrane antigen (EMA) by immunohistochemical staining (Figure 2a-g). On the basis of her medical history, we conducted a pathological review of the brain specimen obtained by the first operation and found that it showed the same histological findings and immunohistochemical profile as our current EUS-FNA specimen (Figure 2a'-d'). Consequently, the final diagnosis of the brain specimen was changed from meningioma to solitary fibrous tumor of the central nervous system, and the pancreatic mass was diagnosed as metastasis from solitary fibrous tumor of the central nervous system. A previous report suggested that gross resection is the most important key to a good prognosis [2]. There was no evidence of further metastasis to other organs in this case, so the patient underwent middle pancreatectomy as a limited surgery. The cut surface of the resected specimen demonstrated a noncapsulated solid mass, 3.4×3.1×2.8 cm in size, with a clear border (Figure 3). The resected pancreatic specimen was negative for surgical margin, vessel invasion, and lymph node metastasis. Further, it also had the same histological findings and immunohistochemical profile as the brain specimen (Figure 2a''-d''), thus, finally diagnosed as pancreatic metastasis of solitary fibrous tumor of the central nervous system.

Solitary fibrous tumor is a rare spindle neoplasm arising from pleura as first reported by Klemperer and Rabin in 1931 [10]. However, in the 1990s they were also found in various sites besides pleura, such as subcutaneous soft tissue, retroperitoneum, orbit, salivary gland, pharynx, thyroid gland, mammary gland, kidney, liver, pancreas, bladder, prostate, and adrenal gland. Although the histogenesis of solitary fibrous tumor is still controversial, it is assumed that solitary fibrous tumor is a mesenchymal neoplasm of probable fibroblastic origin [11].

As in the present case, some cases may have been misdiagnosed as intracranial meningioma because the features of solitary fibrous tumor of the central nervous system were not established until the seminal report by Carnerio *et al*. which was

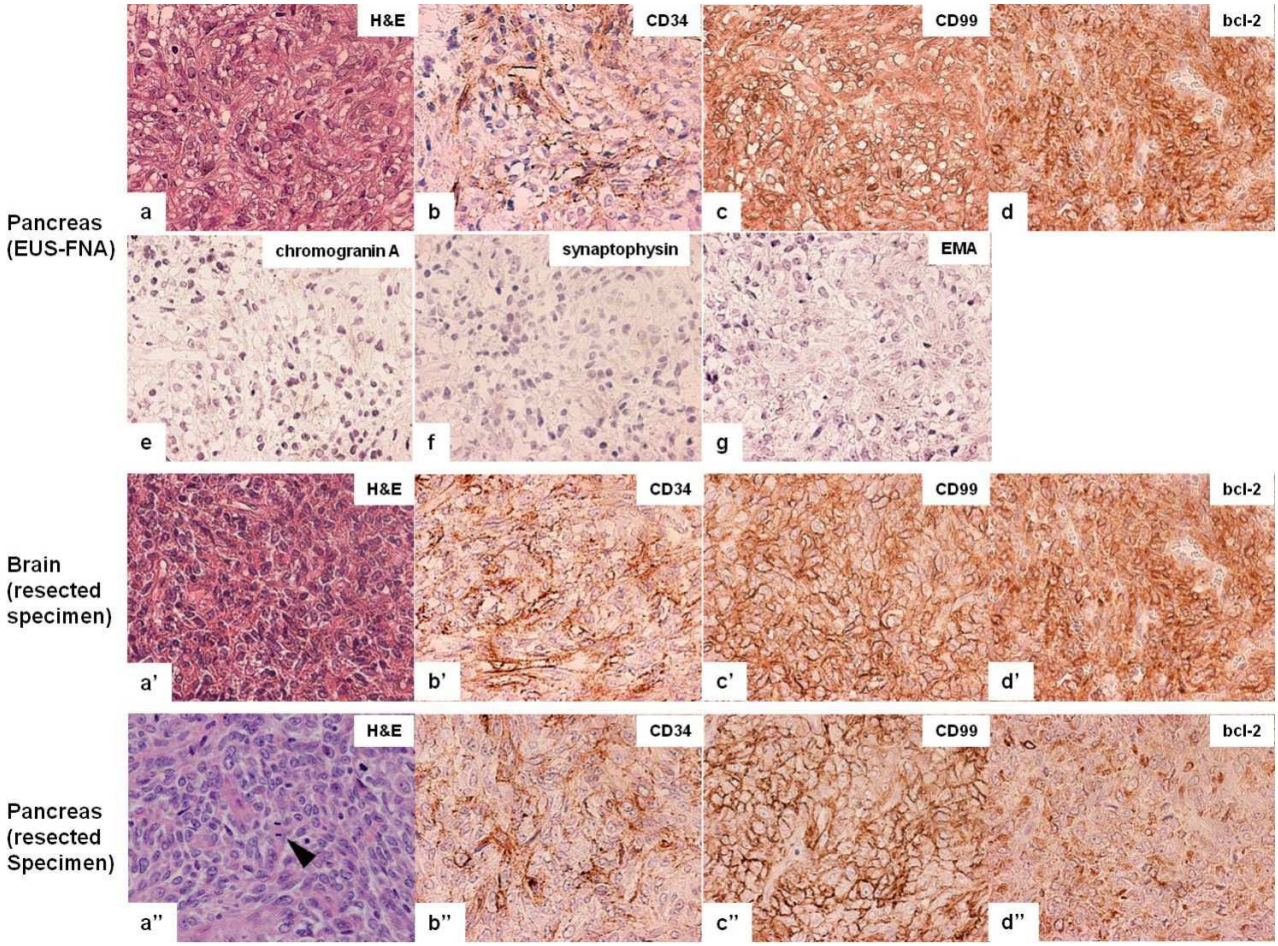

Figure 2. Histological findings and immunohistochemical profiles (400×). H&E staining of the EUS-FNA specimen showed presence of elongated cells containing oval or spindle shaped nuclei in a "patternless pattern" in EUS-FNA, surgically resected brain, and pancreatic specimen (**a**, **a'**, **a''**). The atypical feature of high mitotic rate (>4 mitoses/10 HPF) was found only in the resected pancreatic specimen (high magnification images of the boxed area from **a''**). Immunohistochemical profile of the EUS-FNA specimen demonstrated CD34, CD99, and bcl-2 positivity (**b**, **c**, **d**), but chromogranin A, synaptophysin, and EMA negativity (**e**, **f**, **g**,), which were identical in the surgically resected brain and pancreatic specimen (**b'**, **c'**, **d'**, **b''**, **c''**, **d''**).

published in 1996 [1]. Nowadays, this problem can be resolved by immunohistochemistry. Solitary fibrous tumor of the central nervous system is strongly positive for CD34, CD99, and bcl-2, but not epithelial membrane antigen (EMA), whereas meningioma demonstrates positivity for EMA, but not CD34 [12, 13, 14].

In general, biological behavior of solitary fibrous tumor of the central nervous system is indolent, with 33 recurrent cases, including 28 intracranial recurrences and 5 extracranial hematogenous metastases, of 220 cases in total documented in a survey conducted between August 1996 and July 2011 as reported by Bisceglia *et al*. [2]. Further, they concluded that the best predictors of an unfavorable outcome are incomplete surgical resection, brain infiltration, and atypical histological features, defined as the presence of nuclear atypia and/or cellular pleomorphism, hypercellularity, high mitotic rate (>4 mitoses/10 HPF), and necrosis. In order to better understand the background of the metastasis to the pancreas in the present case, we additionally reviewed the pathological findings of the brain and pancreatic specimens obtained by surgery. Atypical histological features and brain infiltration of the brain specimen were not seen, but its margin was evidently positive; therefore, first brain surgery was interpreted as an incomplete resection which may have been the main cause of the two intracranial recurrences and the pancreatic metastasis. Further, investigation of the pancreatic specimens revealed an atypical histological feature, that of high mitotic rate (>4 mitoses/10 HPF) (Figure 2a''), which indicated that this patient should be carefully observed due to high risk of recurrence.

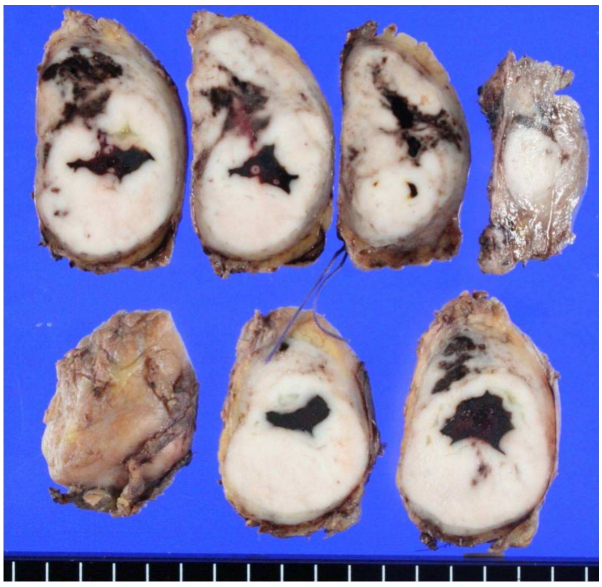

Figure 3. Photograph of the cut surface of the middle pancreatectomy specimen demonstrating a well-defined noncapsulated solid mass, 3.4×3.1×2.8 cm in size, with a central hemorrhage.

The pancreatic mass of present case showed a well-demarcated round shape with hypervascularity, considered as representative findings of NET of the pancreas, by contrast enhanced CT, which has high specificity, with a mean of 96% (range 83-100%) for diagnosis of NET [15]. However, these features may also indicate several other diseases such as metastatic pancreas tumor from renal cell carcinoma [16], pancreatic solitary fibrous tumor [17, 18], pancreatic schwannoma [19], and intra-pancreatic accessory spleen [20]. It is difficult to distinguish these diseases from NET by imaging examination alone. However, these issues can be resolved by EUS-FNA in some diseases, such as intra-pancreatic accessory spleen [20] and metastatic tumor of pancreas. Cause of which were confirmed by histology and/or immuno-histochemistry [21].

Meanwhile, NETs with larger size (>2cm in diameter) and main pancreatic stenosis indicate malignancy, thus necessitating typical pancreatic resection such as pancreaticoduodenectomy or distal pancreatectomy with lymph node dissection [22]. However, in our case the diagnosis obtained using image examination was overturned from NET of the pancreas to pancreatic metastasis of solitary fibrous tumor of the central nervous system by EUS-FNA. The manner of metastasis was hematogenous spread, so the patient underwent limited surgery instead. Selection of an adequate method of surgical resection is very important for patients with suspected low-grade malignancy or borderline malignancy, except for pancreatic ductal carcinoma, because pancreaticoduodenectomy is too invasive for such patients. Lately, limited surgery such as enucleation [23], middle pancreatectomy [24], and duodenum-preserving pancreas head resection [25] has been widely performed. Its advantages are preservation of duodenal function and both endocrine and exocrine functions of the pancreas.

CONCLUSION

We showed here the case of pancreatic metastasis from solitary fibrous tumor of the central nervous system. Even in such rare pancreatic masses, histological findings and immunohistochemical profile obtained by EUS-FNA are invaluable for the correct diagnosis to avoid excessive surgical procedures.

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Conflict of interests The authors declare no conflict of interest

Note The patient has provided permission to publish these features of her case, and the identity of the patient has been protected

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