CASE REPORT

Pancreatic metastasis from an osseous solitary fibrous tumour

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SUMMARY

A 73-year-old man presented with a right-sided chest wall mass that showed an epithelioid neoplasm with mild cytologic atypia on core needle biopsy. He underwent surgical resection, and final pathology revealed solitary fibrous tumour/hemangiopericytoma with negative margins. Three years after surgical resection, the patient presented with fatigue, abdominal pain, weight loss and mildly elevated liver function tests. Further workup revealed a 1.2 cm hypervascular mass in the neck of the pancreas. This case report summarises the surgical treatment and outcomes for a patient who presented with this rare tumour.

BACKGROUND

Solitary fibrous tumour (previously known as hemangiopericytoma or giant cell angiofibroma) is a rare neoplasm, which was first described by Stout and Murray in 1942.¹ It is a tumour of fibroblastic origin with a branching vascular pattern.² It can occur anywhere in the body where capillaries are found but usually occur in the soft tissues, thoracic cavity, pelvis, lower extremities or retroperitoneum.^{3,4} In most cases, solitary fibrous tumours behave in a benign fashion; however, it is a potentially aggressive neoplasm, with local recurrence and distant metastasis. The most common sites of metastasis are the bone, liver and lung.^{3,5} Pancreatic metastases are rare, and of the reported cases in literature, all were from primary central nervous system hemangiopericytoma. Here, we report a case of primary osseous solitary fibrous tumour with pancreatic metastasis.

CASE PRESENTATION

The patient initially presented in 2010 at the age of 73 years with a right-sided chest wall mass. A needle biopsy was performed, and the aspirate showed low-grade spindle cells. He subsequently underwent a core biopsy that revealed an epithelioid neoplasm with mild cytologic atypia of unclear nature. In view of the uncertain nature of the mass, the patient underwent resection including portions of three ribs and chest wall musculature, with reconstruction using a prolene mesh. Final histopathology, along with immunohistochemistry, was consistent with solitary fibrous tumour/hemangiopericytoma (figure 1A,B). All surgical margins were negative, and the two resected lymph nodes were negative for neoplasm.

The patient's interval history was unremarkable. However, 3 years later, he presented to his primary care physician with complaints of fatigue, vague abdominal pain and weight loss.

INVESTIGATIONS

Laboratory results were notable for mildly elevated liver function tests (alanine transaminase 58 U/L, aspartate transaminase 55 U/L), which prompted a right upper quadrant abdominal ultrasound. A 1 cm pancreatic mass was noted on ultrasound, which was further characterised by MRI. Abdominal MRI revealed a 1.2 cm mass in the neck of the pancreas with isointense signal of T2-weighted images (figure 2A), mild hypointense signal on T1-weighted images (figure 2B) and rim enhancement on contrast-enhanced, T1-weighted images (figure 2C). The mass was noted to be solid and hypervascular with progressive contrast enhancement. An endoscopic ultrasound revealed a well-circumscribed hypoechoic nodule in the pancreatic mass measuring 1.2 cm in diameter, which was closely abutting the pancreatic duct (figure 3). The remaining pancreas and pancreatic duct appeared normal. An endoscopic ultrasound-guided fine needle aspiration was performed at the time that showed spindle cell neoplasm consistent with solitary fibrous tumour/hemangiopericytoma.

TREATMENT

The patient underwent surgical resection by means of a laparoscopic robotic central pancreatectomy with Roux-en-Y pancreaticojejunostomy. A 3.6 cm segment of the pancreas was resected. The pancreas demonstrates a solitary well-circumscribed white-tan lesion that measures 1.5 × 1.2 × 1.1 cm. The lesion is composed of spindled cells arranged in fascicles with areas with a herringbone pattern. There is a prominent staghorn vessels throughout the lesion. The lesion compresses but does not infiltrate into the surrounding pancreatic parenchyma. A panel of immunohistochemical stains were performed, and the neoplastic cells were strongly positive for CD34, BCL2 and STAT6. The neoplastic cells were negative for pancytokeratin, EMA, CD31 and p63. The morphological and immunohistochemical features are consistent with a solitary fibrous tumour with negative margins (figure 1C–H).

OUTCOME AND FOLLOW-UP

The patient made an uncomplicated recovery from surgery. However, approximately 1 year

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Figure 1 Pathology. The patient's previous chest wall mass demonstrated destruction of the bone with involvement of the soft tissue. The cells are spindled with areas of staghorn vessels (A, B). In the pancreatic resection specimen, the lesion measured 0.5×1.2×1.1 cm. The lesion is well circumscribed compressing the background pancreas (C, D). The tumour is hypercellular consisting of spindled neoplastic cells arranged in fascicles and a herringbone pattern with prominent staghorn vessels (E). The neoplastic cells were positive for expression of CD34 (F) and STAT6 (H) but negative for CD31 (G).

after undergoing resection of the pancreatic mass, the patient developed recurrence in the left chest wall, which was resected. Despite negative margins at the time of left chest wall resection, the patient went on to develop multiple metastatic lesions that were not amenable to surgical resection.

DISCUSSION

Solitary fibrous tumour is a rare mesenchymal tumour of fibroblastic origin. They occur most commonly in middle-aged adults in the soft tissues, pelvis, lower extremities or retroperitoneum.²⁻⁴ It classically exists as grossly well-circumscribed or thinly encapsulated tumours,¹² with a yellow to tan-coloured cut surface.² On cross-sectional imaging, they appear as well-circumscribed hypervascular tumours, although these findings are non-specific.⁶ Microscopically, they have a thin-walled branching vascular pattern. Tumour cells are usually spindle shaped to round with varying

Figure 2 MRI abdomen, axial images. Axial magnetic resonance images show a 1.2 cm nodule in the neck of the pancreas with isointense signal on T2-weighted images (A), mild hypointense signal on T1-weighted images (B) and rim enhancement on contrast-enhanced T1-weighted images (C).

Figure 3 Endoscopic ultrasound image shows a 1.2 cm well-circumscribed hypoechoic nodule in the pancreas.

cellularity and variable mitotic rates and are often positive for CD34 and CD99.²

Approximately 10%–15% of solitary fibrous tumour/hemangiopericytomas behave aggressively, with local invasion occurring more commonly in hypercellular tumours with increased mitotic figures, necrosis and haemorrhage.^{2,3} In fact, necrosis and haemorrhage are twice as common in locally recurrent and metastasizing tumours.³ Metastasis to the liver, lung and bone is the most common, with rare reports of pancreatic metastasis.^{3,7,8} There are cases of pancreatic metastases from primary intracranial hemangiopericytoma reported in literature.^{8,9} In one case series, pancreatic metastasis occurred between 9 and 24 years after primary resection of primary intracranial hemangiopericytoma.⁸ However, there were no cases of pancreatic metastasis from primary osseous solitary fibrous tumour/hemangiopericytoma in the literature. This may be related to the low incidence of osseous solitary fibrous tumour/hemangiopericytoma, which comprises less than 0.1% of all primary bone tumours.¹⁰

The mainstay of the treatment of solitary fibrous tumour is surgical excision.^{3,7} Although there is some evidence for decreased local recurrence rates with adjuvant radiation therapy for intracranial hemangiopericytoma,⁹ there is little clinical evidence for the role of adjuvant chemotherapy or radiation.¹¹ There is, however, limited evidence for the use in pancreatic metastasis. Long-term outcome is unclear due to the rarity of the disease, although a 5-year and 10-year survival rate of 75% and 44%, respectively, for primary osseous hemangiopericytoma has been described in the literature.¹² The survival rate for pancreatic metastasis from primary intracranial hemangiopericytoma varied. However, complete resection of the metastatic lesions afforded better outcomes in the limited number of cases presented in the literature.⁸ On the basis of the literature available, complete resection of metastasis should be attempted when feasible. The role for chemoradiation, however, is unclear.

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Learning points

- ▶ Solitary fibrous tumour is a rare mesenchymal tumour of fibroblastic origin that usually occurs in the soft tissues, pelvis, lower extremities or retroperitoneum.
- ▶ Approximately 10%–15% of solitary fibrous tumours behave aggressively with metastasis to the liver, lung and bone being most common.
- ▶ Pancreatic metastasis of solitary fibrous tumour is rare, and there are no cases of pancreatic metastasis from primary osseous solitary fibrous tumour reported in the literature.
- ▶ Mainstay of the treatment of solitary fibrous tumour is surgical excision, and complete resection of metastasis should be attempted when feasible.

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