Rare disease

Solid-pseudopapillary carcinoma: a case study and literature review

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Summary

Solid-pseudopapillary tumour (SPT) is a rare exocrine tumour of the pancreas and is considered to have low malignant potential. Few morphological criteria are used to predict malignant behaviour such as equivocal perineural invasion, angioinvasion and invasion to surrounding tissue, and should be designated as solid-pseudopapillary carcinoma (SPC). We report a case of SPC. Clinical and radiological findings are typical for SPT with no metastatic disease. There is no tumour recurrence after 4 months postresection. Clinical history and radiological findings were retrieved from the patient's record sheet and Viarad system. H&E staining and few immunoperoxidase staining were reviewed by several pathologists. The histological findings are typical for SPT, with additional perineural invasion. There is no angioinvasion or capsular invasion identified. This is our first experience in diagnosing and managing SPC. We look forward to seeing the patient's disease status during her next routine follow-up. We expect good disease-free survival and very low risk of tumour recurrence, in view of only one risk factor (perineural invasion) and uninvolved surgical margins by the tumour.

BACKGROUND

Solid-pseudopapillary tumour of the pancreas (SPT) is a rare exocrine tumour of the pancreas, with 2–3% incidence rate among primary pancreatic tumour.¹ It was described by Franz in 1959,² and has several other names, such as solid and cystic tumour, solid and papillary neoplasm, Franz's tumour, papillary-cystic neoplasm and papillary epithelial neoplasm.^{1–3} SPT is the accepted terminology of this entity.

SPT is considered as low malignant potential, despite its benign morphology and it rarely metastasised.^{4–5} Even though exocrine tumours of pancreas have bad prognosis principally, SPT shows good prognosis, which makes the disease entity unique among this disease group. To predict malignant behaviour of SPT, few malignant morphological criteria are needed (angioinvasion, invasion to surrounding tissue or unequivocal perineural invasion) and should be designated as solid-pseudopapillary carcinoma (SPC).

We are reporting our first SPC case in this case study and we would like to share our experience, mainly in pathology point of view, in diagnosing and helping our clinician to manage the patient.

CASE PRESENTATION

A 14-year-old Malay woman, presented with a gradual onset of dull-left hypochondriac pain with radiation to her back. No medical history. She had history of early satiety with associated changes in appetite. However, she had no jaundice or vomiting. She denied history of abdominal trauma or pancreatitis. She had a history of intussusceptions which was operated during her neonatal

period. Family history was negative for pancreatic diseases. On examination there is a transverse surgical scar located at the right lumbar region, also is a smooth palpable mass arising from the left hypochondriac region.

INVESTIGATIONS

Blood investigation was normal.

CT abdomen scan revealed a large well-encapsulated mass arising from the tail of the pancreas (figures 1 and 2).

Histopathological finding revealed SPC with perineural invasion (figure 3).

DIFFERENTIAL DIAGNOSIS

Pancreatic tumour; most likely benign in nature.

TREATMENT

She underwent distal pancreatectomy with preservation of spleen.

OUTCOME AND FOLLOW-UP

Postoperative period was uneventful. After 4 months post-tumour resection, she had no evidence to suggest residual or tumour recurrence in routine Abdominal CT scan. There was no chemotherapy or radiotherapy started yet.

DISCUSSION

The diagnosis of SPT in this patient was expected from typical clinical presentation and imaging examination. However, the morphology revealed perineural invasion, an indicator for malignant behaviour, despite overall histological presentation of typical benign SPT.

Figure 1 Contrast-enhanced CT study of the abdomen.

SPT is defined by WHO as a benign tumour that is usually present in young girls and has pseudopapillary configuration and solid area histologically. Grossly, the tumour is usually encapsulated and shows a brown light-lobulated, solid cut surface with central haemorrhagic-cystic changes. But when the tumour has unequivocal perineural invasion, angioinvasion and deep invasion into the surrounding tissue, the tumour should be designated as SPC. There are many other factors such as venous invasion, degree of nuclear atypia, mitotic count and prominence of necrobiotic cell nest in association with malignant behaviour. However, the above-mentioned criteria are usually not present in metastasising tumour.⁶

The tumour can occur in both men and women, but more common in women (6:1–8.37:1).^{3 5 7–12} SPT has a wide range age at presentation from 6 to 75 years with median age of 27.2 years.^{3 7} Presentation at the first decade of life is rare. Abdominal discomfort and pain are the most common clinical presentations (36.8%) and are followed by palpable abdominal mass (12.3%).³ This similar clinical

presentation is also seen in other case series.⁵ In another review, they noted that nearly one-third of the patient were asymptomatic.^{7 13} The tumour is commonly seen involving the body or tail of the pancreas.^{3 5 7 9 12 14} Extrapaneatic SPT also had been reported.^{7 12 15 16}

Diagnosis of SPT by radiological imaging technique is variable. CT scan can accurately diagnose SPT in 80% of cases.⁴ In advance of the imaging technique, the SPT can be diagnosed even in a small lesion,⁴ with median tumour size being 7 cm (range from 4 to 16 cm).⁸ The tumour usually diagnosed by CT scan and has a well-circumscribed border with heterogeneously enhanced solid and cystic area. Areas of calcification at periphery are also seen.¹² Accuracy is improved when the patient is a young woman.^{3 11 14} However, few clinical reviews had shown low diagnostic specificity (23.7–50%).^{7 8} In our case, the patient was diagnosed as SPT after typical CT scan appearance with clinical history of a young girl. MRI is the best imaging technique for children due to reduce risk of radiation and better visualisation of central haemorrhage.¹⁷

Figure 2 Macroscopic examination. (A) The residual pancreatic tissue was unremarkable (red arrow). (B) Cut sections showed a solid, yellow-to-brownish tumour with central cystic degeneration and necrosis (red arrow).

Figure 3 Light microscopic examination under 200 × magnification. (A) Monotonous proliferation of round tumour cells in pseudopapillary architecture; (B) perineural invasion (red arrow).

Both central haemorrhage and peripheral fibrous pseudocapsule (heterogeneous signal intensity in T2-weighted image and low signal intensity in T1-weighted image, but usually enhance after gadolinium application) are important findings or correct diagnosis because the features are very unusual in other pancreatic tumours.¹⁷

Fine needle aspiration (FNA) cytology and endoscopic ultrasound role are controversial in diagnosing SPT and seldom performed.⁷ Less than half patients are correctly diagnosed by this technique in a study³ with few researchers thinking that the accuracy is high.¹⁴ Even though the usage of FNA can increase the likelihood of correct diagnosis, the procedure is associated with an increased risk of peritoneal seeding.^{3 12} Since the accuracy of radiological imaging is variable but usually high in typical cases, FNA should be reserved for doubtful cases, and sometimes clinical judgement and high index of suspicion is very important during the preoperative period since all investigations may be inconclusive.¹⁴

Surgical resection remains the main treatment for SPT,^{10 14} with local resection, distal pancreatectomy or pancreatoduodenectomy are the most common surgical procedures performed.^{7-9 16} The perioperative morbidity and mortality are low¹⁸ and curative complete resection is usually achievable even in huge tumours^{8 9} due to very thick fibrous pseudocapsule. Enucleation or limited pancreatectomy procedure may be adequate.¹⁸ However, a more aggressive surgical approach may be necessary to achieve complete resection.^{5 8} Metastasis to lymph node is rare but many centres agree that routine lymphadenectomy following surgical resection is not indicated.^{3 7 8 14}

The tumours are well circumscribed and encapsulated.^{8 12 14 16 19} Cross-section may show pure solid cut surface or almost total cystic with cystic degeneration and haemorrhage.^{1 5 8 14 20} The mean diameter of tumour is 7.87 cm and ranges from 1 to 25 cm.⁷ They are composed of monotonous,^{1 5 15 20} sheets of cells with ovals, regular nuclei and polygonal cytoplasm. When degenerative process takes place, the cells closest to the vessels are the only viable cells left, leaving perivascular cells, mimicking pseudopapilla or perivascular rosette.^{6 8 16} The degenerative process

includes foamy histiocytes, cholesterol cleft and vacuolated cells.²⁰

The differentiation between acinar cell carcinoma and islet cell neoplasm are based from immunochemistry profile.⁸ Few cases were mistaken as islet cell tumour before the diagnosis of SPT was given.^{1 14 15 21} The tumour is usually positive for α -1 antitrypsin (94.6%), vimentin (93.1%) and neurone-specific enolase (81%).⁷ Progesterone receptor (PR) and oestrogen receptor (PR) positivity are variable,⁵ with more PR positivity (56.7%).⁷ Cytokeratin, chromogranin A and synaptophysin positivity are usually focal.⁷ Ki67 positivity are increased in a descriptive study of three cases of very aggressive disease (two patients²² and one patient¹¹).

SPC is designated only if the tumour shows one or more malignant features (angioinvasion, unequivocal perineural invasion and invasion to surrounding tissue),⁶ with local invasion is the most common malignant feature.^{3 23} Tumour invasions can involve nearby structures such as duodenum, stomach, colon, left kidney, lung, spleen, portal vein, bile duct, liver, diaphragmatic muscle, omentum, peritoneum and superior mesenteric artery.^{3 7 9} A study done by Nishihara *et al* had compared three metastasising and 19 non-metastasising SPT and noted that few histological findings were associated with malignancy, which were venous invasion, degree of nuclear atypia, mitotic count and prominence of necrobiotic cell nests.⁶ However, these findings were not present in all metastasising SPT.⁶ In case series of 14 cases by Tipton *et al*,⁸ three cases who had nuclear pleomorphism and vascular invasion showed liver metastasis and another one had local intraperitoneal recurrence. Similar to two cases reported by Tang *et al*,²² who had unusual histologic features (diffuse growth pattern, extensive tumour necrosis, significant nuclear atypia, very high mitotic activity (35–70/50) and component of sarcomatoid changes), the patients had very aggressive tumour behaviour and rapidly fatal. These morphological findings are only seen in minority of all SPC, but important in managing and predicting malignant potential tumour, and should be considered as part of the criteria to diagnose SPC. Despite all these features and tumour metastasis,

overall long-term survival time is good, which gives a hard time to find a reliable predictive factor.¹⁰

The prognosis of patient with SPT is good, with overall 5-year survival rate of more than 95%.^{7 9 18 22} Tumour recurrence is rare, which account 3.5–6.6%,³ and all are seen in SPC.^{5 7} Even though all tumour recurrences are in the SPC group, only few will recur, make the WHO criteria for malignant potential is a very sensitive tool to predict tumour recurrence and metastasis.³ There are few factors associated with recurrence as proposed by Kim *et al*³ who studied 114 SPT cases, which are metastasis at first operation, invasion of adjacent vessels or organ, mass same or larger than 13 cm, young patient, tumour rupture and inadequate resection. However, few researchers found that tumour more than 5 cm had significant association with malignant potential.²³ Tumour recurrence is common within 5-year-post-tumour resection.⁷ Even after tumour recurrence, the prognosis is still excellent with long survival time, provided that the complete resection or appropriate treatment is given.^{3 7 8 23 24} Therefore, the presence of tumour recurrence is not a contraindication for surgical resection.^{8 10}

Even though she had a criterion for malignancy (perineural invasion), the chances of long survival are high. Since tumour recurrence always occurs in SPC, frequent long-term follow-up is advisable.

Learning points

- It is important to have definite criterion to differentiate between benign and malignant solid-pseudopapillary tumour.
- Surgical treatment is required; however, frequent long-term follow-up is advisable.

Competing interests None.

Patient consent Obtained.

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