Solid Pseudopapillary Neoplasm of the Pancreas

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Summary
Solid pseudopapillary neoplasm is a rare pancreatic tumour predominantly affecting young women. We present two cases in young female patients. Both tumours were surgically removed as abdominal masses, one from the pancreatic tail and the other posterior to the stomach with an unclear organ of origin. On gross examination, both tumours were encapsulated with solid and cystic cut surfaces that had hemorrhage and necrosis. On histological examination, both were composed of solid sheets and pseudopapillae lined by bland, monotonous and uniform round cells. One tumour had features of malignancy. Immunohistochemistry for both cases was consistent with solid pseudopapillary neoplasm. Solid pseudopapillary neoplasm should be considered in the differential in young women presenting with pancreatic masses.

Key Words: Pseudopapillary, Neoplasm, Pancreas

Introduction
Solid pseudopapillary neoplasm (SPN) of the pancreas is rare, accounting for 2-3 % of primary pancreatic tumours (1). It was first described by Frantz in 1959 and has since then been referred to as: solid and cystic tumour, solid and papillary neoplasm, Frantz's tumour, papillary-cystic neoplasm and papillary epithelial neoplasm. It was finally defined by the World Health Organization (WHO) in 2000 as a solid pseudopapillary neoplasm of the pancreas (2).

SPN is considered a low malignant potential neoplasm, due to its benign morphology and the fact that it rarely metastasizes (3). Even though pancreatic tumours generally have bad prognosis, SPN shows good prognosis, which makes the disease entity unique in this disease group. To predict malignant behavior of SPN, several morphological criteria are needed (angioinvasion, invasion to surrounding tissue or unequivocal perineural invasion) and if present, the tumour should be designated as solid pseudopapillary carcinoma (SPC) (4). We present two cases of SPN, highlighting histologic and immunohistochemical features.

Case 1
Distal pancreatectomy specimen was received from a 24 year-old female with an encapsulated tumour at the tail of the pancreas. On gross examination revealed encapsulated, ovoid tumour measuring 13 x 8 x 7.5 cm. There was attached normal pancreas measuring 4 x 2 x 2 cm. The cut surface had both solid and cystic areas with necrosis and haemorrhage (Figure 1a).

Histology showed a tumour with both solid and pseudopapillary foci composed of uniform cells with bland round nuclei and moderate eosinophilic cytoplasm. The pseudopapillae had hyalinised cores with mucinous change. Clusters of cholesterol clefts were also seen. Vascular invasion was identified (Figure 1b-f). The surgical resection margins were positive for tumour. Immunohistochemistry results were as follows: vimentin – positive, CD10 – positive, CD99 – paranuclear dot-like positivity, progesterone receptor – positive (Figure 1 g-i).

The tumour was diagnosed as a solid pseudopapillary carcinoma

Case 2
The second case was a biopsy received from a 15 year-old female patient and was described as a huge mass behind the stomach, the site of origin was queried at surgery as either posterior gastric wall or retroperitoneal origin. The liver and spleen were described as normal. A gastrointestinal

stromal tumour was suspected clinically. Gross examination, a grey encapsulated nodule measuring 3x2.5x2 cm was received. It weighed 80 grams. The cut surface was solid and white with a black, cystic focus. Histology showed an encapsulated tumour composed of solid sheets and pseudopapillae lined by bland cells with moderate cytoplasm and regular nuclei with even chromatin distribution. Focal areas of mucinous change were present (Figure 2 a, b). The surgical resection margins were positive. Immunohistochemistry results were as follows: cytokeratin AE1/AE3 – positive, vimentin – positive, CD10 – positive (Figure 2 c, d).

The tumour was diagnosed as a solid pseudopapillary tumour of pancreatic origin.

Figure 1: Specimens from case 1:

a) Gross appearance. Note the capsule as well as cystic, necrotic and haemorrhagic cut surface. b) Solid growth of monotonous round cells and perineural invasion. (Haematoxylin & Eosin stain, X4). c) Pseudopapillae lined my monotonous, uniform round cells. There is mucinous change of the cores. (Haematoxylin & Eosin stain, X10). d) Lymphovascular invasion (Haematoxylin & Eosin stain, X10). e) Cholesterol clefts. (Haematoxylin & Eosin stain, X10). f) Necrosis. (Haematoxylin & Eosin stain, X20). g) Vimentin immunostain, diffuse and intense staining. (X20). h) Progesterone receptor immunostain, strong nuclear staining. (X20). i) CD99 immunostain, paranuclear dot-like positivity. (X20)

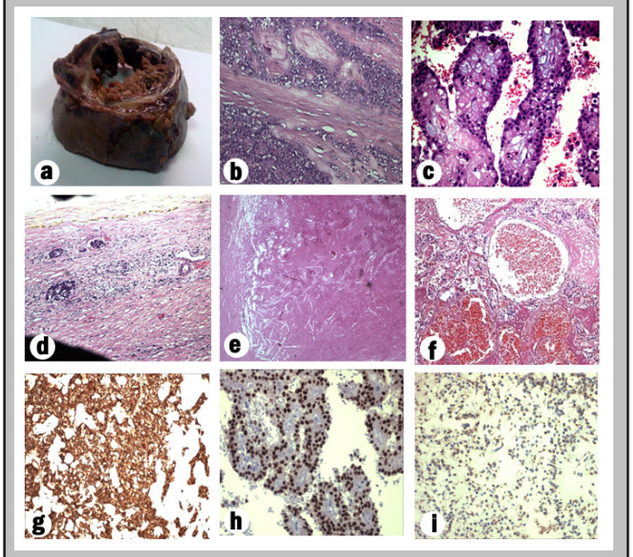
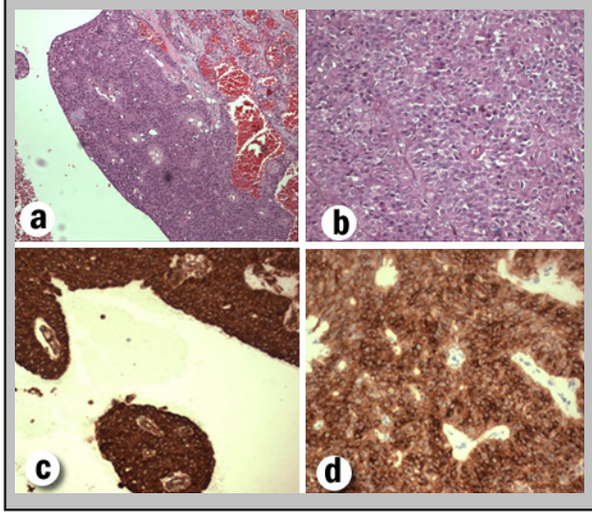


Figure 2: Specimen from case 2:

a) *Low power view of solid sheets and pseudopapillae composed of monotonous, uniform round cells. (Haematoxylin & Eosin stain, X4). b) High power view of the solid area of the tumour. (Haematoxylin & Eosin stain, X20). c) Vimentin immunostain, diffuse and intense staining. (X20). d) CD10 immunostain, diffuse and intense staining. (X20*



Discussion

SPN is defined by WHO as a usually benign tumour that occurs predominantly in young females, although it can occur in men as well. Mean age of presentation is 27.2 years (5). The tumours are discovered incidentally on routine examination, or they may cause abdominal discomfort and pain or a palpable abdominal mass (4). Diagnosis of SPN is possible by CT scan but specificity varies from 23.7–50% (6,7). Its appearance on CT scan is that of a well-circumscribed tumour with heterogeneously enhancing solid and cystic areas. MRI is preferred in younger patients to reduce radiation exposure (7). Surgical resection remains the main treatment for SPN (8), most commonly local resection, distal pancreatectomy or pancreatoduodenectomy (5,6,9). Grossly, they are large with a mean greatest dimension of 9cm and are usually well-encapsulated with a multicystic haemorrhagic and necrotic cut surface. They can be multifocal (10). They usually occur in the tail of the pancreas (8, 9). Histologically, they are composed of monotonous sheets of cells with regular oval nuclei and polygonal cytoplasm. There are also pseudopapillae with hyalinised fibrovascular cores lined by several layers of bland fragile epithelial cells with clear to eosinophilic cytoplasm, variable mucinous changes within the core and intracytoplasmic periodic acid Schiff

positive hyaline globules. The pseudopapillae are due to solid nests minus cells degenerating away from the small vessels; they resemble rosettes in cross section. Other histologic features include: round to oval nuclei, finely stippled chromatin, nuclear grooves, indistinct nucleoli and few mitoses. In addition there may be foam cells and clusters of lipid and cholesterol crystals surrounded by foreign-body giant cells. Pseudocystic areas can also occur. Tumour cells infiltrate without any stromal reaction. Literature shows that when the tumour has unequivocal perineural invasion, lymphangioinvasion and deep invasion into the surrounding tissue, the tumour should be designated as solid pseudopapillary carcinoma (11). Additionally, the degree of nuclear atypia, mitotic count and prominence of necrobiotic cell nests are associated with malignant behaviour. However, the latter criteria are usually not present in all metastatic tumours (6, 11). These morphological findings are only seen in a minority of SPC, but are important in predicting malignancy, and should be considered as part of the criteria to diagnose solid pseudopapillary carcinoma (SPC).

Immunohistochemically, the tumour is intensely and diffusely positive for vimentin, CD10 and CD56 (12). It is variably positive for oestrogen and progesterone receptors (with more PR positivity at 56.7%), and focally positive for neuroendocrine markers. Chymotrypsin and trypsin also stain positive. Others positive stains include: nuclear and cytoplasmic beta-catenin, cyclin D1, nuclear E-cadherin, and paranuclear dot-like CD99.

The cases presented above showed histologic features that are in keeping with SPN. Case 1 additionally had perineural invasion and angioinvasion, features of SPC. Both cases showed positivity for vimentin and CD10. Case 1 additionally showed positivity for progesterone receptor, and paranuclear dot-like positivity for CD99.

None of these markers have any therapeutic or prognostic significance at present. The prognosis of patient with SPN is good, with overall 5-year survival rate of more than 95% (9,13). Tumour recurrence is rare, and all are seen in SPC (5).

Conclusion

Solid pseudopapillary neoplasm is a rare pancreatic tumour that should be considered clinically in young, female patients with abdominal pain and distension. CT scan and MRI can be used for pre-operative diagnosis. Histological and immunohistochemical features are well-defined and sufficient for a definitive diagnosis. Specific microscopic malignant features should be sought and if present, the tumour should be designated solid pseudopapillary carcinoma. Unlike most pancreatic neoplasms, SPN has a good prognosis.

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