Frantz's tumour, solid pseudopapillary epithelial neoplasm (SPEN)

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DESCRIPTION

A 29-year-old woman from Myanmar underwent medical check-up before having a baby. She was healthy and had no familial history of pancreatic disease or cancer. Physical examination revealed an ill-defined palpable mass arising from the left hypochondriac region. Blood tests including tumour marker levels were normal. Her abdominal ultrasound scan revealed a 1.37×1.28 inches mixed echogenic solid mass at distal pancreas without main pancreatic duct dilation (figure 1). Abdominal MRI demonstrated a well-encapsulated mass of size 1.70×1.61 inches, hyposignal T1-weighted (T1W) and heterogeneous hypersignal T2W masses with cystic and solid component with enhancement solid component at the pancreatic tail, with no evidence of intra-abdominal metastasis (figure 2). Distal pancreatectomy was performed. No adjuvant therapy was given, nor was splenectomy performed. She has been followed up as an outpatient for 2 years and has remained healthy without any sign of local recurrent or distal metastasis.

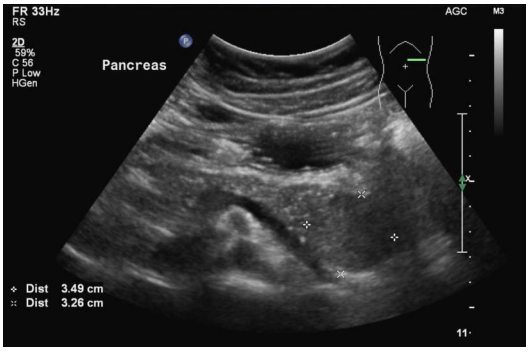

Figure 1 Ultrasound scan finding: A solid mass liked lesion at distal pancreas. No pancreatic duct dilation.

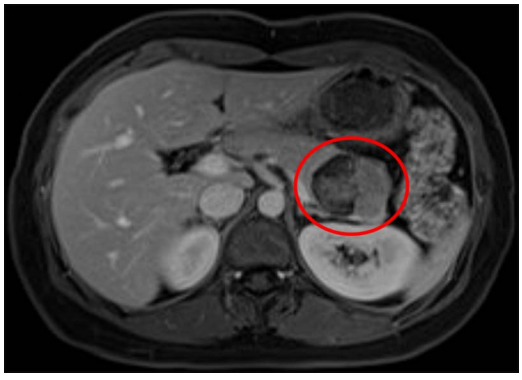

Figure 2 MRI finding: A lobulated hyposignal T1-weighted (T1W), heterogeneous hypersignal T2W masses with cystic and solid component with enhancement solid component at pancreatic tail.

The mass had typical pathological features of solid pseudopapillary epithelial neoplasm (SPEN) (figures 3–5)—a combination of solid and cystic components with a cellular degenerative change

Figure 3 Solid pseudopapillary neoplasm at distal pancreas.

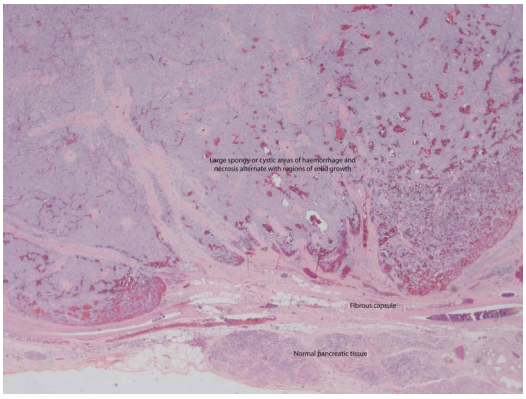

Figure 4 Pathological finding: Tumour tissue presenting cystic areas of haemorrhage and necrosis alternate with regions of solid growth, including notable encapsulation by pancreatic parenchyma (H&E, ×4).

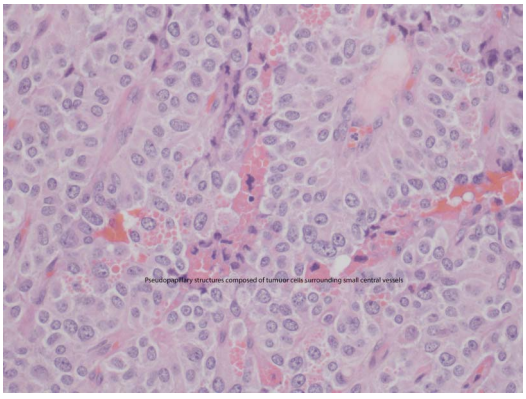

Figure 5 Pathological finding: Solid pseudo papillary structures composed of uniform cells surrounding small vessels (H&E, ×100).

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alternating with pseudopapillary formation along with uniform epithelioid cells with well-vascularised stroma and discohesive arrangement in the solid portions.[1] The differential diagnosis requires immunohistochemical staining. SPEN cells are usually absent when stains for endocrine markers are used and are characteristically positive for vimentin, CD10, CD56, and α-1-antitrypsin.[2] The commonly encountered cystic neoplasms of the pancreas can be classified into five categories: serous microcystic adenoma, mucinous cystic neoplasms (cystadenoma or cystadenocarcinoma), intraductal papillary mucinous neoplasm (IPMN), cystic neuroendocrine neoplasms, and SPEN. Among these uncommon pancreatic tumours, SPEN represents an exceedingly rare entity with low potential malignancy, usually found in asymptomatic young woman who undergo screening imaging studies.[1 2] It has many other names including Frantz tumour, as first described by Virginia Kneeland Frantz in 1959. Complete surgical resection is mandatory.[2 3]

Learning points

- Solid pseudopapillary epithelial neoplasm (SPEN) or Frantz's tumour is a rare benign condition, usually found in asymptomatic young woman who undergo screening imaging studies.
- Complete surgical resection is mandatory.

Contributors OT was the primary physician, made the diagnosis, managed the case and prepared the first draft of the manuscript. SK provided pathological results and helped to draft the manuscript. KP contributed to the concept, and helped to draft and revised the manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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