Solid pseudopapillary tumour of the pancreas in a child: case report
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Abstract
Introduction
Solid pseudopapillary tumour of the pancreas, though rare, is a low-grade malignancy with indolent behaviour. It is encountered predominantly in young females, although it has been seen in males and children. Most patients present with vague non-specific abdominal pain resulting in delayed diagnosis. Therefore diagnosis of this tumour may be an incidental finding during diagnostic imaging procedures or maybe assumed when a young woman presents with an asymptomatic palpable mass. Traditionally the presence of a large bulky pancreatic tumour in a child should raise suspicion of the diagnosis of pancreatoblastoma. This study reports a case of a solid pseudopapillary tumour of the pancreas in a child.

Case report
We present a case of an eight-year-old child presenting with pain and a lump in the epigastric region. Computerised tomography was subsequently performed to localise the mass accurately. Computed tomography showed a minimally enhancing solid mass in the head and proximal body of the pancreas compressing the second and third part of the duodenum and measuring 7.2 × 6.9 × 6.4 cm.

Conclusion
This case report highlights the fact that with characteristic imaging features the diagnosis of solid pseudopapillary tumour of the pancreas should be considered irrespective of the age profile of the patient.

Introduction
Solid pseudopapillary tumour (SPT) of the pancreas is rare and often suspected in young females who present with pancreatic mass accounting for less than 2% of exocrine pancreatic neoplasm1,2. SPT commonly occur in women in the second to fourth decade of life and is rare in children. It is thought to arise from cells of the exocrine pancreas, though some investigators have postulated their origin from the endocrine pancreas3. These tumours generally go undetected till they grow to a large size. Despite this presentation these tumours show low malignant potential and rarely metastasise. Therefore, surgical excision offers an excellent prognosis even in large or metastasising tumours4. Despite a few citations it is worthwhile to note that SPT may present on the odd occasion as an acute abdomen with sudden onset of pain due to compression upon the adjoining organs5. SPT shows different clinical features in childhood. High survival rates can be achieved in most cases, warranting aggressive treatments even in metastatic disease5. A meticulous radiological survey prior to any surgical intervention may aid in the timely diagnosis of this condition. This study is an attempt to highlight the importance of imaging modalities such as CT scan and ultrasound in particular, in the diagnosis of SPTs of the pancreas.

Case report
An eight-year-old female child presented to paediatric OPD with complaints of pain and a lump in the epigastric region for four months. Routine haematological and biochemical parameters were normal. On physical examination, patient was mildly cachexic. A non-tender slightly mobile epigastric mass, measuring 5 × 7 cm was palpable. On clinical test (decubitus, knee-elbow position) it was suspected to be of retroperitoneal origin. USG done on Philips 4000 showed a well-defined homogenously hypoechoic, smoothly marginated mass with a few necrotic areas in the epigastric region. The mass was inseparable from the pancreas. Adjacent visceral structures were normal with duodenum closely abutting the mass (Figure 1).

Figure 1: USG with colour Doppler showing avascular epigastric region solid mass with compression of splenic and portal vein and displacement of adjacent arteries.
Colour Doppler investigation revealed the mass to be avascular and displacing adjacent portal vein, SMA and splenic vein (Figure 2). Both plain and contrast CT abdomen were performed on Siemens Emotion 6 machine. It revealed a minimally enhancing solid mass in the head and proximal body of the pancreas compressing the second and third parts of the duodenum and measuring 7.2 × 6.9 × 6.4 cm. No calcification or haemorrhage was seen on plain scan, though a few necrotic areas and peripheral capsular enhancement was present. Splenic vein and superior mesenteric vein were compressed by the mass just short of their confluence. Inferior vena cava and aorta with their major branches were found to be normal. The common bile duct (CBD) and pancreatic duct were not dilated (Figure 3).

MRI on Philips 1.5 T Achieva unit was done for further anatomical and morphological characterisation of the lesion. It was hypointense on T1WI and hyperintense on T2WI compared to pancreatic parenchyma and showed a thin hypointense capsule (Figure 4). Postcontrast study showed minimal enhancement of the mass and moderate enhancement of the capsule. Magnetic resonance cholangiopancreatography revealed normal calibre of the CBD and MPD. The MPD was displaced superiorly by mass and was not visualised in its mid-course but seen normal distally. Therefore, it was inferred that the mass is displacing rather than encasing the MPD (Figures 5 and 6).

Pancreatoblastoma was kept as the first diagnosis in view of the solid nature of mass and age of the patient. SPT was kept as second possibility even though the age of the patient was not in favour, based on CT and MRI imaging features like presence of true capsule, paucity of necrosis relative to tumour size, minimal enhancement of the mass with good capsular enhancement and patchy foci of haemorrhage are also seen within the mass.

Figure 2: CECT abdomen showing minimally enhancing pancreatic mass with no evidence of calcification. Peripheral capsular enhancement is seen. Adjacent visceral structure and vessels are displaced by the tumour.

Figure 3: Axial T2W SPIR image showing hyperintense mass with hypointense capsule. Minimal area of patchy cystic degeneration seen. Adjacent bowel loops are displaced by the mass. CBD shows normal calibre.

Figure 4: Precontrast T1W image showing shows well-defined, smoothly margined pancreatic mass with hypointense capsular rim. Hyperintense foci of haemorrhage are also seen within the mass.
SPT is most commonly diagnosed in adolescent girls and young women, and a predilection for Blacks and East Asians has been suggested. Most patients with SPT are brought to medical attention in the second decade of life. Female patients account for 83%-98.5% of the reported cases. Controversies regarding the role of genetic or hormonal factors in the causation of the tumour to explain the strong female predilection are present. Some authors in the past have denied it whereas others have agreed to it strongly.

Zhou et al. have suggested that SPT may represent the most common pancreatic tumour in Asian children.

SPT is a slow-growing tumour and hence the majority of patients present with vague abdominal symptoms, resulting in a delay in presentation and diagnosis. It is commonly large and circumscribed or encapsulated, with haemorrhagic changes. The tumour is usually round to ovoid and solitary and can occur in any part of the pancreas, although some investigators have observed a predilection for the tail.

The tumour is thought to be ductal or acinar in origin. The tumour exhibits typical features on light microscopy. The cystic appearance on gross examination may present a diagnostic dilemma with cystic neoplasm of the pancreas. Immunophenotyping has been used to differentiate these tumours from other pancreatic neoplasms. SPTs show positive tests for vimentin, neuron-specific enolase, α1-antitrypsin and α1-antichymotrypsin, and are negative for chromogranin, epithelial membrane antigen, and cytokeratin, insulin and glucagon. Cytological smears are highly cellular and show monotonous population of small cells arranged in aggregates and papillae with fibrovascular cores.

There are no clear histological features, which establish the clinical behaviour of these neoplasms. The tumours exhibit low-grade foci of T1W hyperintensity within the mass suggesting haemorrhage.

Surgical exploration of the tumour was performed which revealed a 7 cm diameter pancreatic mass arising from the head and proximal body of the pancreas displacing the duodenum anterolaterally. The major blood vessels were displaced. The tumour was in close relationship with MPD, so along with tumour resection a Whipples procedure was carried out (Figure 6). Postoperative course was uneventful. Histopathology confirmed the diagnosis of SPT of the pancreas.

**Discussion**

SPT of the pancreas is a rare tumour and only 700 reports have been reviewed in the literature since it was first described by Frantz in 1959. The tumour has been known by a number of synonyms including Frantz’s tumour, solid and cystic acinar tumour, papillary epithelial neoplasm, and solid and papillary epithelial tumour or neoplasm. In 1996, the World Health Organization recognised the designation ‘solid pseudopapillary tumour’ as a distinct tumour of the exocrine pancreas.

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malignant potential and metastasise infrequently\textsuperscript{13}. The imaging features of SPT reflect the pathologic findings of cystic and solid components, intratumoural haemorrhage, a fibrous capsule and, less commonly, calcification. When present, the fibrous capsule and internal haemorrhage are the features that distinguish SPT from other pancreatic tumours\textsuperscript{10,12}.

USG and CT show a large well-circumscribed mass with quite variable appearance depending on its composition. Tumours compress adjacent structures rather than invading them. CT shows solid portions of the mass to be iso to hypovascular and displaces the pancreas on T1-weighted images and are small in size), acinar cell tumour, ductal adenocarcinoma (patients are in the older age group, lymphoma (homogenous enhancing mass with bulky lymph nodes), lymphangioma, dermoid cyst and pseudopancreatic cyst (appears as a multiseptated cystic lesion on USG with none to minimal peripheral enhancement), primitive neuroectodermal tumour and mesenchymal tumours (heterogeneous and infiltrating margins)\textsuperscript{10,12}.

### Conclusion
The possibility of SPT should always be kept in a child who presents with large bulky pancreatic mass and characteristic imaging features. Because of the indolent nature of these tumours and the low malignant potential, aggressive attempts at complete surgical resection are warranted. Large tumours are usually resectable and size does not predict the outcome. Surgical bypass may be the only feasible option in patients with large tumours where the risks of attempting resection or debulking may be associated with overwhelming morbidity.

### Consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

### References

### Table 1 Comparison of the pseudopapillary tumour with pancreaticoblastoma

<table>
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<th>Features</th>
<th>SPT</th>
<th>Pancreaticoblastoma</th>
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<tr>
<td>Age</td>
<td>Second decade</td>
<td>First decade of life</td>
</tr>
<tr>
<td>Sex</td>
<td>Female</td>
<td>Male predominance</td>
</tr>
<tr>
<td>Biochemical</td>
<td>No such association</td>
<td>Associated with elevated fetoprotein in one-third cases</td>
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<td>Location</td>
<td>Tail</td>
<td>Head of the pancreas</td>
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<td>Radiological appearance</td>
<td>Capsule with internal haemorrhage</td>
<td>The tumour appears multiloculated with enhancing septa</td>
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