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Extremely rare presentation of Frantz's tumour: synchronous localisation in the pancreatic head and tail

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Abstract

Introduction: Solid pseudopapillary tumour of the pancreas is a rare lesion with low malignant potential occurring predominantly in young women. This is a report of an extremely rare occurrence of synchronous presentation of pseudopapillary tumour in the pancreatic head and tail of a 16-year-old female patient.

Case presentation: The patient presented with a three-month intermittent upper abdominal pain and swelling. Computed tomography scan showed two separate masses, involving the pancreatic head and tail. The patient underwent surgery, where successful tumour enucleation of both tumours was performed. Histological report confirmed solid pseudopapillary tumour of the pancreas with the low malignant potential.

Conclusion: To the best of our knowledge, this is the first report of synchronous presentation of pseudopapillary tumour of the pancreas.

Keywords

Solid pseudopapillary neoplasm, Frantz's tumour, pancreas, synchronous neoplasms, children

Introduction

Solid pseudopapillary tumours of the pancreas, first reported by Frantz in 1959, are rare and occur most frequently in young women.[1,2] These tumours are composed of poorly cohesive, monomorphic cells forming solid and pseudopapillary structures with frequent hemorrhagic-cystic degeneration.[2] Different names of this tumour were reported until it was defined by the World Health Organization in 1996 as a 'solid pseudopapillary tumour' of the pancreas.[2] These tumours rarely cause symptoms, and can be located anywhere in the pancreas.[3] Symptoms of solid pseudopapillary tumours of the pancreas are often nonspecific and include abdominal pain, dyspepsia, early satiety, nausea and vomiting.[1,4]

Although solid pseudopapillary tumour is considered an indolent lesion with a low malignant potential and a favourable prognosis after surgical resection, 10–15% of the patients will develop metastases.[5] These metastases are often amenable to resection, and complete extirpation is associated with long-term survival.[3,5] The occurrence of infiltrating varieties of solid pseudopapillary tumour is very rare.[3–5] To the best of our knowledge, this is the first report of synchronous presentation of pseudopapillary tumour of the pancreas, localised in pancreatic head and tail.

Case report

Medical history

A 16-year-old female presented to our department with a three-month intermittent upper abdominal pain and swelling.

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Clinical features

On physical examination, an 8 cm, well-defined, non-tender, non-pulsatile mass was palpable in the epigastrium. Hematological and biochemical studies were unremarkable. The tumour markers (CA19-9, CEA and AFP) were normal.

Abdominal ultrasound (US) and computed tomography (CT) showed two circumscribed encapsulated heterogeneous masses with solid and cystic areas and calcifications, the first one arising in the head of pancreas, measuring $10.2 \times 7.1 \times 6$ cm (Figure 1(a)), and the second arising in the tail of pancreas, measuring $6.9 \times 6.8 \times 6.5$ cm (Figure 1(b)). No lymphadenopathy or other pathology was found.

Operative findings

At laparotomy, large encapsulated tumours of pancreatic head and tail were found with no evidence of intra-abdominal metastasis. Successful tumour enucleation for both tumours was performed. Both tumours were encapsulated and well-demarcated from the surrounding pancreas (Figure 2).

Pathologic examination

On macroscopic examination, both tumours were round and solid, the first from pancreatic head measured $10 \times 9 \times 5.5$ cm and the second from pancreatic tail measured $7 \times 6.8 \times 6.5$ cm. The tumour was composed of solid cellular nests with poorly supported tiny vessels resulting in perivascular pseudopapillary pattern (Figure 3(a)). The tumour cells were uniform, small- to medium-sized with ovoid nuclei. Eosinophilic periodic acid-Schiff (PAS)-positive 'hyaline globules' and vacuoles were found in cytoplasm (Figure 3(b)). Lipid crystals were surrounded by foreign-body giant cells (Figure 3(c)). Immunohistochemical staining with Ki-67 antibody reveals low proliferative activity (Figure 3(d)). Final histological report confirmed solid pseudopapillary tumour of the pancreas.

Differential diagnosis

The differential diagnosis included other tumours that share the microscopic appearance, mainly acinar cell carcinoma, mixed acinar-endocrine carcinoma, pancreatic endocrine neoplasms and pancreatoblastoma.

Outcome, prognosis and follow-up

The postoperative course was complicated by acute pancreatitis and pseudocyst formation three weeks after surgery. Pancreatic pseudocyst was treated by

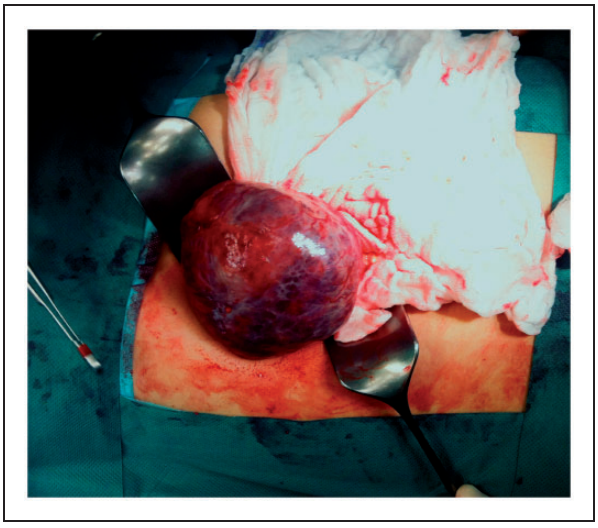

Figure 2. Intraoperative finding: large and solid encapsulated tumour, well demarcated from the surrounding pancreas.

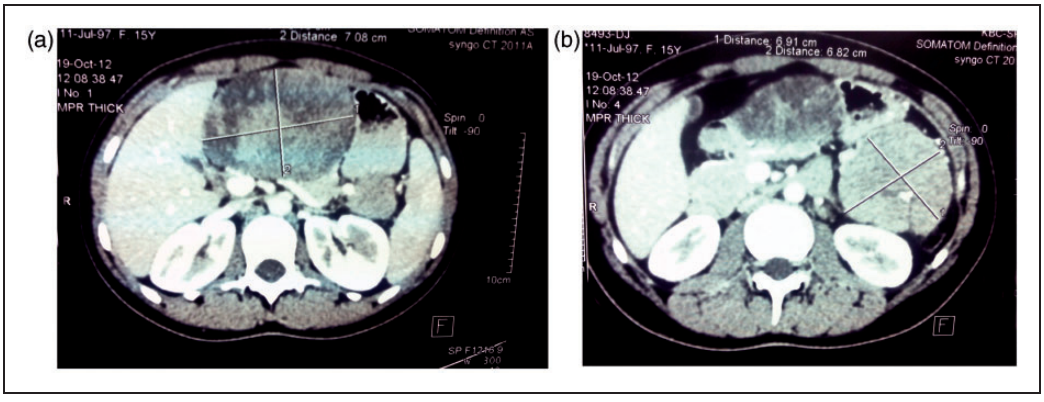

Figure 1. Computed tomography showed two circumscribed encapsulated heterogeneous masses with solid and cystic areas and calcifications. (a) Tumour arising in the head of pancreas and (b) tumour arising in the tail of pancreas.

the administration of CT-assisted external percutaneous drainage. The cyst was drained percutaneously for six weeks. After that period, CT imaging showed total cyst regression. The patient is currently disease-free 24 months after surgery.

Discussion

Solid pseudopapillary tumour was first observed in a 19-year-old girl in 1927. However, Frantz first described it in 1959, and he proposed that four cases with non-functional island cell tumours were actually a new entity called 'papillary pancreas tumour'.[4,6] In general, solid pseudopapillary tumour occurs predominantly in young women and is rare in men.[6,7] Tien et al.[8] reported no significant differences in patient age, size, neoplasm location or malignancy rate between the genders in solid pseudopapillary tumour. By contrast, Machado et al.[9] reported that cases of solid pseudopapillary tumour in men are more aggressive and that the patients are older. Solid pseudopapillary tumours account for 1–2% of exocrine pancreas neoplasms and for 1% of all pancreas tumours and 3% of all cystic pancreas neoplasms.[4–7] There are about 800 cases reported in the English literature, and 553 cases were reported in Chinese literature studies.[2,4,6] The increasing frequency of solid pseudopapillary tumours in the last 10 years has caught much attention. As patients present with no specific symptoms, including abdominal pain or discomfort, poor appetite and nausea, which are related to tumour compression on adjacent organs, tumours are usually identified incidentally on routine radiological examination as ultrasound or CT. Consequently, the tumours reach large proportions at the time of identification.[6,7] Our patient had no specific symptomatology. Rare cases presenting with spontaneous rupture, intra-abdominal bleeding and gastric outlet obstruction have also been reported.[6,10] In patients with solid pseudopapillary tumours, as in our case, tumour markers are generally within normal limits.

Most of the tumours are localised in pancreatic tail, second most common localisation is pancreatic head while pancreatic corpus is rare localisation.[7] All reports in literature are single-tumour presentation. Only two cases of synchronous solid pseudopapillary neoplasm and intraductal papillary mucinous neoplasm of the pancreas were reported in English literature.[7,11]

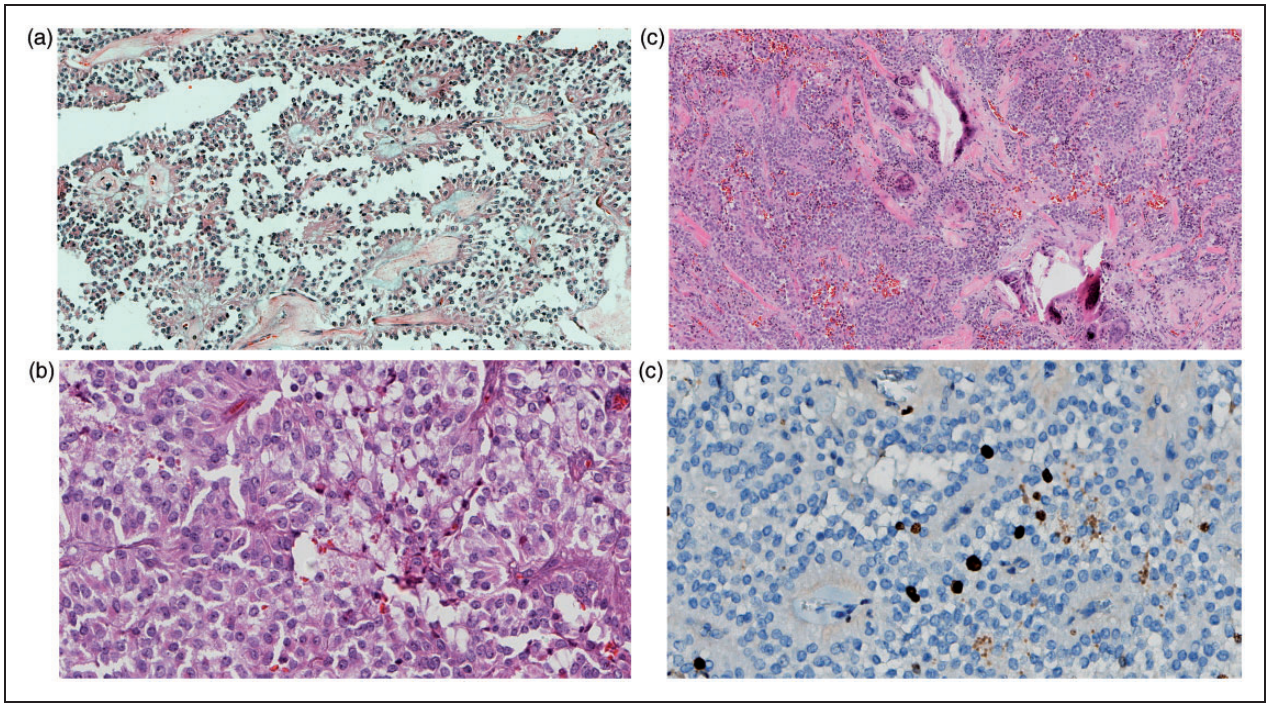

Figure 3. Pathohistological findings: (a) the tumour was composed of solid cellular nests with poorly supported tiny vessels resulting in perivascular pseudopapillary pattern (HE, ×200), (b) the tumour cells were uniform, small- to medium-sized with ovoid nuclei. Eosinophilic PAS-positive 'hyaline globules' and vacuoles were found in cytoplasm (HE, ×400), (c) lipid crystals were surrounded by foreign-body giant cells (HE, ×100) and (d) Immunohistochemical staining with Ki-67 antibody reveals low proliferative activity (immunohistochemistry Ki-67 DAKO, Denmark, ×400).

To the best of our knowledge, this is the first report of synchronous tumour presentation of solid pseudopapillary tumour localised in pancreatic head and tail.

Although the malignant potential of solid pseudopapillary tumours is low, up to 15% of the patients develop metastasis. The most common sites of metastasis are the liver, regional lymph nodes, mesentery, omentum and peritoneum. Local invasion of the duodenum, stomach, spleen or major blood vessels may also occur, as reported by Yu et al.[2] Washington et al.[12] in their study showed that the clinical and pathologic features of solid pseudopapillary tumours, including diffuse growth, venous invasion, nuclear pleomorphism, mitotic rate, necrosis and dedifferentiation, are related to its aggressive behaviour or metastatic potential. Sun et al.[13] reported that 62.5% of solid pseudopapillary tumour patients are infected with hepatitis B virus (HBV), which can induce over-expression of ß-catenin in tumour cells, indicating that HBV infection may be involved in the pathogenesis of solid pseudopapillary tumour, which, however, has not been confirmed. Microscopically, the growth pattern of the tumour cells is remarkably uniform, with a combination of solid, pseudopapillary or hemorrhagic pseudocystic structures in various proportions. The tumour contains a mixture of solid, cystic and pseudopapillary patterns in various proportions. Immunohistochemically, the tumour cells are diffusely positive for vimentin; most cases express diffuse positive staining for neuron-specific enolase, some of them are focally positive for creatin kinase (CK) and synuclein (SYN), and few for S-100 protein.[13]

Yu et al.[2] reported that patients positive for Ki-67 immunoreactivity were confirmed to have malignant solid pseudopapillary tumour, suggesting that positive staining of Ki-67 may correlate with the malignant potential and poor outcome of solid pseudopapillary tumours. Some studies also reported high proliferation index for Ki-67 (>25%) is an indication of malignancy.[7] In our case, the Ki-67 proliferation index was low.

The prognosis of solid pseudopapillary tumour even in the presence of local recurrence, invasion or metastases is favourable. The five-year survival rate is more than 95%.[7,14] Local recurrence is less than 10% and usually occurs four years after surgery.[2,7,9,13] Even with metastases, the survival rate is reported to be more than 10 years.[7]

Surgical resection has been considered the treatment of choice although few cases of spontaneous tumour regression have been reported.[2,4] Most cases of solid pseudopapillary tumour have previously undergone the standard pancreatic resections such as pancreatoduodenectomy and distal pancreatectomy because the tumours were relatively large and misdiagnosed as malignant disease.[14] Newer studies recommended a minimised resection such as enucleation and partial resection for solid pseudopapillary tumour, because there were no significant differences in the incidence of postoperative complications and the prognosis between the minimised resection and the standard resection.[1,2,6,15,16]

Conclusion

Solid pseudopapillary neoplasm of the pancreas is a rare low-grade malignant tumour that typically affects young women without significant symptoms. Its behaviour is relatively indolent. Surgical enucleation alone is the best treatment and it should be pursued regardless of distant metastasis and size. The prognosis of patients with solid pseudopapillary tumours even with unresectable metastasis is good.

Learning points

- Solid pseudopapillary tumours of the pancreas are composed of poorly cohesive, monomorphic cells forming solid and pseudopapillary structures with frequent hemorrhagic-cystic degeneration.
- In general, solid pseudopapillary tumours occur predominantly in young women and are rare in men.
- These tumours rarely cause symptoms, and can be located anywhere in the pancreas. Symptoms of solid pseudopapillary tumours of the pancreas are often nonspecific and include abdominal pain, dyspepsia, early satiety, nausea and vomiting.
- The prognosis of solid pseudopapillary tumour even in the presence of local recurrence, invasion or metastases is favourable. The five-year survival rate is more than 95%.

Authors' contributions

The work presented here was carried out in collaboration among all authors. IJ and ZP performed operation. IKP performed pathohistological analysis and report. JSG and ZP defined the research theme, performed literature review and wrote the paper. IJ, IKP and ZP have been involved in drafting the manuscript or revising it critically for important intellectual content. JSG and IKP have given final approval of the version to be published. All authors have contributed to, seen and approved the manuscript.

Declaration of conflicting interests

None declared.

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