Rare disease

Solid pseudopapillary tumour of the pancreas in an 11-year-old Iranian girl

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Summary

Solid pseudopapillary tumour (SPT) of the pancreas is a low-malignancy-potential tumour of the pancreas. It mostly occurs in young female adults. It is uncommon in childhood. Here, we present the case of an 11-year-old girl with SPT.

BACKGROUND

Solid pseudopapillary tumour (SPT) of the pancreas accounts for 1–3% of the primary pancreatic tumour cases.¹ Most of the patients are young women. The female to male ratio is approximately 10:1.² The female predilection encouraged sex hormone receptor studies, but only a small proportion of patients had oestrogen- or progesterone-positive tumours. Therefore, it seems that sex hormones are more likely to play a role in tumour growth than in the pathogenesis of SPT.¹ These tumours have an unclear pathogenesis and a low malignancy potential. Surgical resection offers a very good chance of achieving long-term survival.² There are only a few case reports on SPT in children. The disease has different clinical features in adults and children. Tumours in children are larger and more symptomatic.³ Because long-term survival can be achieved even with the synchronous or metachronous metastatic lesions, solid pseudopapillary neoplasm (SPN) should be treated aggressively, with complete resection, even though this requires metastatectomy. There is no clear family history of cancer in these patients. In this paper, the case of an 11-year-old girl with SPT and a family history of breast cancer in her mother is presented.

CASE PRESENTATION

Our patient is an 11-year-old Iranian girl who had been healthy with no serious medical problem until 6 months ago when she had been referred to the emergency ward because of severe sudden onset of abdominal pain. The location of pain had been mostly in the left upper quadrant radiating to the epigastric area and her back. The pain had been relieved in the sitting position and had become worse when lying down. Although the patient had been suffering from nausea and vomiting, her appetite had not been affected. Liver function tests, carcinoembryonic antigen (CEA) and alpha fetoprotein were normal. Abdominal sonography had already been carried out for the patient in another clinic, and at referral she presented the results to us. An isoechoic cystic mass with 50 mm in the greatest diameter was seen in the tail of pancreas; however, the images were poor and therefore

not presentable in the paper. Abdominal CT scanning (figure 1) showed a well-defined 80×65 mm hypodense mass in the tail of the pancreas with faint rim enhancement. The other findings were normal. One noticeable finding was a history of infiltrating hormone-positive ductal carcinoma of the breast in her mother.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis included non-functional pancreas endocrine tumour (PEN), acinar cell carcinoma and in its clear-cell variant of pancreatic endocrine neoplasm a ductal adenocarcinoma and metastatic renal cell carcinoma.^{4–6} Ductal adenocarcinomas are mostly seen in elderly men. They are much smaller than SPTs. Microscopically, the tumour is formed of ductal and glandular structures. They grow in an infiltrative manner with capsular circumscription. Neuroendocrine tumours usually present with a solid or microacinar pattern. Haemorrhage and pseudocystic areas are not usually observed. Nuclei are small, round, smoothly contoured and possess fine

Figure 1 Abdominal CT scan shows a well-defined 80×65 mm hypodense mass in the tail of the pancreas with faint rim enhancement.

chromatin structures. SPT may be confused macroscopically with pseudocyst unless sufficient samples are taken.

Characteristic microscopic features of SPT allow easy differential diagnosis. Immunohistochemical staining can be helpful. Negative chromogranin A and synaptophysin staining essentially rules out PEN, whereas negative keratin stain excludes acinar carcinoma.⁴ Clear cells in SPN do not contain mucin, glycogen or lipid, substances found in PEN.⁷ Clear-cell ductal adenocarcinoma is cytokeratin 7 (CK7) positive and CD10, CD56 and vimentin negative and only membranous β -catenin is positive.⁷

TREATMENT

The patient underwent distal pancreatectomy and the resected brownish-coloured mass, approximately 100×60×20 mm, was sent to the pathology department. Microscopic evaluation revealed small- to medium-sized and polygonal to elongated cells, with ovoid nuclei often grooved or indented (figure 2). Some rare cells presented enlarged or irregular nuclei. Nucleoli were inconspicuous. Mitotic cells were rare. The cytoplasm ranged from clear to eosinophilic and lacked glycogen and mucin. These cells made papillary structures around the vessels. The cells adjacent to the tiny blood vessels were attached to the stroma and to one another; however, those farther away were detached from each other. Slit-like spaces separated the pseudopapillae. Foci of cystic and myxoid degeneration and necrosis and foamy macrophages were seen. Diagnosis was solid pseudopapillary neoplasm of the pancreas.

OUTCOME AND FOLLOW-UP

The patient showed an uneventful postoperative course. CT scanning performed 2 months later was normal. She is well now.

DISCUSSION

For the first time VK Frantz in 1959 described this neoplasm in four cases mistakably diagnosed as non-functioning islet cell tumours. She named this tumour as ‘papillary tumour of the pancreas—benign or malignant’. In 1970, Hamoudi *et al*⁴ described the distinctive histological characteristics of this tumour. In 1996, WHO described this tumour and named it

Figure 2 Microscopic evaluation revealed small- to medium-sized and polygonal to elongated cells, with ovoid nuclei that often were grooved or indented.

as ‘solid pseudopapillary tumour’, leading to the various phrases that have been used to refer to this tumour. The histogenesis of these tumours is unknown but they possibly originate from the primordial cells and lack definite endocrine and exocrine differentiation.⁸ This tumour affects mainly young women and its peak incidence is in the second and third decades, and it is rare in children.³ Oestrogen and progesterone receptors were found in some of the cases. It seems that sex hormones are more likely to play a role in tumour growth than in the pathogenesis of this neoplasm.¹ This theory may explain tumour growth during pregnancy and the effectiveness of tamoxifen observed in some patients.¹ Our patient’s mother had oestrogen and progesterone receptor-positive breast cancer. The tumour mostly presents with abdominal discomfort or pain. Sometimes there is a palpable mass. Less common symptoms include: nausea, vomiting, fever, jaundice, weight loss² and anaemia as a result of tumour rupture.¹ Our patient had nausea, vomiting and abdominal pain. SPT often remains asymptomatic until the tumour has enlarged considerably and therefore it is sometimes detected incidentally on abdominal sonography for another reason.¹ The mean diameter of the tumour is 6 cm in adults^{2 3} and 8 cm in children³ and maybe this is the reason why the tumour is more symptomatic in children. The location of the tumour is different in children and adults. The tail and body of the pancreas is the most common site of disease in adults;^{1 2 9} however, the head of the pancreas is the most common location of the tumour in children.^{1 3} However, in our patient the tumour location was the tail. There are reports of extrapancreatic tumours as a result of ectopic pancreas.¹ Generally, there is no evidence for pancreatic insufficiency, either endocrine or exocrine. Liver function tests, pancreatic enzymes and tumour markers such as CEA and carbohydrate antigen 19-9 (CA 19-9) are usually normal^{1 10} as in our patient. CA 19-9 serum level increases in 71–93% of patient with adenocarcinoma of pancreas and is a useful tumour marker for evaluation of exocrine pancreatic carcinoma; however it remains normal in most of SPT patients. Although sonography is a helpful procedure, contrast-enhancing CT scanning is more sensitive and specific, showing a large hypervascular encapsulated mass in contact with the pancreatic tissue, with peripheral contrast enhancement. It may show either a solid or a cystic aspect, or it may be heterogeneous in appearance. The existence of various layers of cystic tissues will correspond to the solid tissue necrosis found in the pathological study. Occasionally, calcifications and bleeding signs may be seen inside the tumour.¹⁰ MRI can be diagnostic. Typically, a large, well-defined, encapsulated lesion with heterogeneous high or low signal intensity on T1-weighted, heterogeneous high signal intensity on T2-weighted and early peripheral heterogeneous enhancement with progressive fill-in is found on gadolinium-enhanced dynamic MRI. These features help differentiate this rare tumour from other pancreatic neoplasms.⁸ Endoscopic ultrasound-guided fine-needle aspiration can have an important role and provide an accurate preoperative diagnosis.^{1 11}

Surgery should be considered the therapy of choice. The localisation and presence of local invasion affect the surgical management: from simple excision for a mass surrounding with dense fibrous tissue in the tumours of the body and tail as in our patient to classic pancreatic oduodenectomy (Whipple) for the tumours of the head of the

pancreas.^{1 2 4 9 12} Invasion to the portal vein or superior mesenteric artery should not be included as a criterion for non-resectability of these pancreatic neoplasms.² Extensive lymphadenectomy is not necessary.¹⁰ Complete surgical removal of the tumour even in the case of metastases or local invasion offers an excellent prognosis.¹²

Although resection of the tumour provides a 5-year survival rate of 97%, local recurrence or distant metastases can occur in a significant number of patients. In a minority of patients there is a locally advanced or metastatic disease at presentation.¹³ Metastases involved one or more of the following organs: lymph nodes, liver, spleen, colon, mesocolon and generalised carcinomatosis.¹² Although there are no established clinical or histological criteria to predict the biological behaviour of SPT, invasion of blood vessels, perineural infiltration, invasion of adjacent structures, high degree of cellular polymorphism and elevated mitotic rate are suggested to be associated with metastases and recurrence. The absence of these features does not preclude malignant behaviour.¹³ Metastasis in children is rare. Recurrent and metastatic disease must be managed surgically as much as possible.^{1 2 4 8 12 14} Although the role of chemotherapy is unknown, there are a few reports of experimental chemotherapy with various chemotherapy drugs (fluorouracil, cisplatin, ifosfamide, etoposide and gemcitabine) for metastatic disease.^{1 4 10 12 13} It was helpful in some cases. Neoadjuvant chemotherapy for increasing the respectability of advanced disease with invasion to portal vein or superior mesenteric vein was done with good response.¹² There are a few case reports showing a survival benefit with radiotherapy in unresectable diseases; however, there is no role for radiation therapy in an adjuvant setting or incomplete resection.

Learning points

- Solid pseudopapillary tumour (SPT) is a rare pancreatic tumour that typically affects women and occasionally children.
- The typical site of SPT in children is the head of the pancreas; however, as in adults it may occur in the pancreatic body or tail.
- It seems that surgical resection can be helpful for SPT in spite of mesenteric artery and/or portal vein involvement and even metastasis, contrary to adenocarcinoma of the pancreas.

Competing interests None.

Patient consent Obtained.

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