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Laparoscopic distal pancreatectomy of a solid pseudopapillary tumour in a teenager after size reduction: A case report

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ABSTRACT

A solid pseudopapillary tumour (SPT) of the pancreas is very rare in children. Despite its malignancy potential, SPT is known to have a good prognosis. No prior report has described the use of neoadjuvant chemoradiation with retinoid therapy for size reduction of an SPT before surgical excision.

An 11-year-old girl had intermittent abdominal pain and a huge mass occupying the whole abdomen. The plain abdominal radiograph and abdominal computed tomography scan showed a 16-cm, huge pancreatic mass across the midline. An ultrasonography-guided biopsy was performed, and pathological examination confirmed an SPT of the pancreas. As the mass was adjacent to large vessels, chemoradiotherapy was preferred to surgical resection. After 1 year of chemoradiotherapy, the tumour decreased to 14 cm. Three years of immunotherapy with retinoic acid reduced the tumour to 10 cm. Laparoscopic distal pancreatectomy was performed after the size of the tumour had decreased. The tumour was successfully excised, and the pathological examination confirmed an SPT of the pancreas without malignant characteristics. The patient's postoperative clinical course was uneventful; there was no evidence of recurrence or metastasis during the 16-monthfollow-up.

Surgical resection is a reliable treatment for SPTs; however, a second option needs to be used when operation is inappropriate. In our case, subsequent laparoscopic distal pancreatectomy was successful.

A solid pseudopapillary tumour (SPT) of the pancreas is very rare and accounts for about 1–2% of all solid pancreatic tumours [1]. It is known to have low-grade malignant potential, and its prognosis is good after surgical resection [2]. Here, we describe a patient with a huge SPT located across the midline and abutted on large vessels. Our report illustrates how neoadjuvant chemoradiotherapy with retinoid therapy can reduce the size of a large SPT, enabling complete excision.

1. Case report

An 11-year-old girl was referred to our services because of intermittent epigastric pain and a huge abdominal mass. Her epigastric pain persisted for 2 weeks and was vague and dull. She had previously been healthy. On physical examination, the mass was round, firm, and smooth-surfaced, but immovable and across the midline. The patient's vital status was stable, and her laboratory findings were within normal ranges. Normal values were also observed for tumour markers, including α-fetoprotein, the carcinoembryonic antigen, and carbohydrate antigen 19-9. On the abdominal plain radiograph, a huge mass with calcification occupied the whole abdomen, and the bowels were shifted in the lower abdomen (Fig. 1). The computed tomography (CT) scan showed a 165 × 140 × 105-mm pancreatic tumour with mixed components adjacent to the superior mesenteric vein (SMV), celiac trunk, and great curvature of the stomach (Fig. 2A–B). An ultrasonography-guided biopsy was performed, and an SPT of the pancreas was confirmed pathologically. Immunohistochemistry staining showed that the tumour was positive for α-1 antitrypsin, the progesterone receptor, neuron-specific enolase, and vimentin, and it was negative for S-100 and the cytokeratin pan monoclonal antibody. The huge tumour abutted on vital structures, including the SMV and celiac trunk, so that the risk of bleeding, injury of SMV and celiac trunk and incomplete resection were concerned; therefore, chemoradiotherapy was performed. Six cycles of gemcitabine

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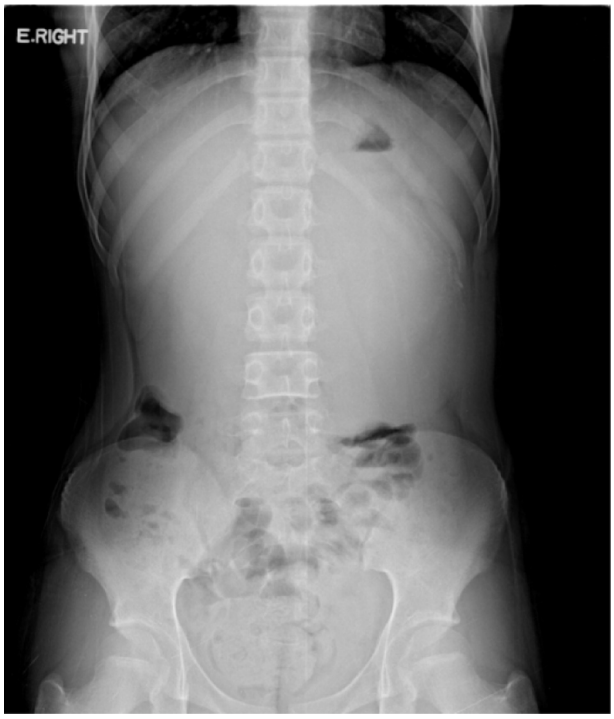

Fig. 1. Abdominal radiograph. Plain abdominal radiograph delineating a huge, space-occupying lesion with calcification in the whole abdomen and peripheral shift of the bowels.

chemotherapy were administered over 6 months. Subsequently, the abdominal CT scan showed a subtle reduction in the tumour size (from 165 × 65 m 105 mm to 154 × 139 × 112 mm). Radiotherapy was performed during the following 2 months (total 3600 cGy[cGy]: 180 cGy by 20 times over 4weeks). After radiotherapy, the abdominal CT scan showed a large reduction in the tumour size (from 154 × 139 × 112 mm to 139 × 120 × 89 mm). Three more cycles of chemotherapy were administered over 3 months, using a multidrug regimen of vincristine, ifosfamide, etoposide, and cisplatin. However, there was no significant change in the tumour on the follow-up abdominal CT scan. Retinoid therapy (Roaccutan®, Roche, Basel, Switzerland) of 100 mg m^{-2} $day^{-1}$was administered to the patient for 1 year. The size of the tumour reduced significantly, shrinking from 139 × 120 × 89mmto 108 × 102 × 155 mm. Regular follow-up visits with ultrasonography and retinoic acid therapy were continued for 3 years, and further size reductions were observed (from 108 × 102 × 155 mm to 88 × 77 × 96 mm; Fig. 3).

Laparoscopic exploration was performed. The tumour was in the body and tail of the pancreas (Fig. 4A–B). There was neither adjacent organ invasion nor enlarged lymph nodes. Laparoscopic distal pancreatectomy was performed successfully. The pathological examination demonstrated that the tumour was an SPT with extensive haemorrhagic necrosis and dystrophic calcifications (Fig. 5A–B). It showed positive immunohistochemistry for CD56, chromogranin, synaptophysin, and β-catenin, and it was negative for E-cadherin.

The patient's postoperative clinical course was uneventful. On postoperative day 4, she was discharged from the hospital. During the 16-month follow-up, ultrasonography and abdominal CT showed no evidence of residual mass or recurrence in pancreas.

2. Discussion

An SPT is most commonly detected because of symptomatic presentation with abdominal pain [3]. However, the next most common reason for SPT detection is incidental identification [3], e.g. imaging performed for unrelated purposes. Lee et al. [4] reported that the most common location of an SPT was the head of the pancreas. However, Kim et al. [3] reported that the most common location was the body and tail of the pancreas. Our patient complained of abdominal pain and had a palpable abdominal mass. The location of the tumour was the body and tail of the pancreas, and it occupied the entire middle of the abdomen because of its huge size. A previous report described the traumatic rupture of an SPT of the pancreas [5], but this seemed unlikely to occur in our patient.

Surgical resection is suitable treatment for an SPT, and it has excellent results. The surgical strategy depends on the location of the tumour: pancreaticoduodenectomy is performed for tumours in the head of the pancreas, central pancreatectomy is performed for tumours located in the body of the pancreas, and distal pancreatectomy is performed for tumours located in the tail of the pancreas. Although some reports have described good long-term outcomes despite a remnant SPT [6], other reports have described destructive progression of an SPT [7]. Tumours that appear predominantly solid on a CT scan have greater malignant potential, and a histologically malignant SPT is associated with recurrence [3]. For unknown reasons, an SPT behaves indolently in general, despite having malignant potential.

Adjuvant or palliative therapy would generally be considered when surgical approaches are inappropriate or hazardous. The

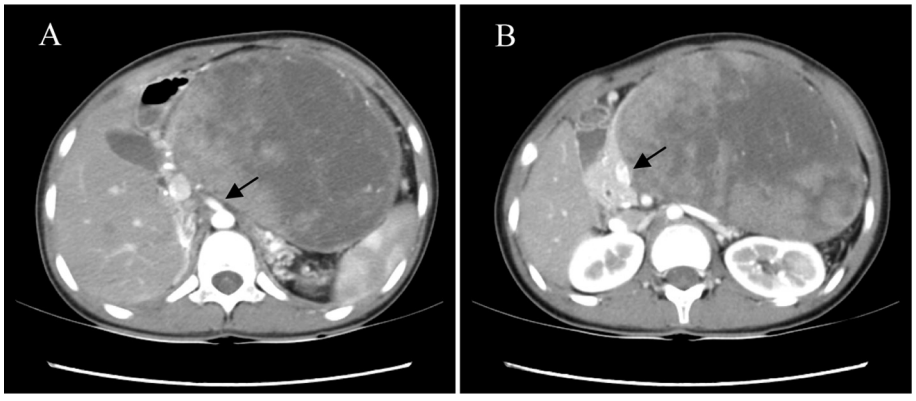

Fig. 2. Abdominal computed tomography scans. A large, solid pseudopapillary tumour (165 × 105 mm) abuts the celiac trunk (A, arrow) and superior mesenteric vein (B, arrow).