

Endoscopic linear stapler-assisted resection of a giant solid pseudopapillary pancreatic tumor with concurrent regional portal hypertension: a case report

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Abstract

Solid pseudopapillary tumor of the pancreas (SPTP) is a rare neoplasm with a low incidence and low rate of malignancy. We herein report a rare case of SPTP concurrent with regional portal hypertension (RPH) that was successfully treated by distal pancreatectomy and splenectomy. A 22-year-old woman presented with a left upper abdominal apophysis and normal liver function. She was diagnosed with an SPTP and RPH by abdominal ultrasound and computed tomography, and she subsequently underwent distal pancreatectomy and splenectomy. Noticeably, varicose vein plexus with wide range appeared on the upper edge of the pancreatic body and posterior gastric wall of the patient. Therefore, we created a path to avoid touching the varicose veins and took advantage of the endoscopic linear stapler to staple the veins. We herein report our surgical experience on SPTP assisted with the endoscopic linear stapler, which will be very realistic for the management of this rare clinical entity.

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Keywords

Solid pseudopapillary tumor of the pancreas, regional portal hypertension, endoscopic linear stapler, distal pancreatectomy, splenectomy, apophysis

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Background

Regional portal hypertension (RPH) can be classified into pancreatic, splenic, or retroperitoneal according to its origin; among these, pancreatic portal hypertension (PPH) is the most common.¹ PPH is primarily caused by retardation of the venous flow in patients with pancreatic-related diseases, especially chronic pancreatitis, and it mainly presents as splenomegaly and varicose veins in the spleen and stomach accompanied by normal liver function and portal veins.¹ Pancreatic tumors and pseudocysts can also result in portal hypertension. Solid pseudopapillary tumor of the pancreas (SPTP) is a rare pancreatic neoplasm with a low incidence. It was first described by Franz² in 1959 and accounts for less than 1% to 3% of pancreatic tumors.^{2,3} The clinical manifestations of SPTP are nonspecific. Surgical resection is currently the most effective treatment method for preventing and curing SPTP. Pancreatic surgery remains challenging in many cases because of its anatomical location and proximity to important vascular structures. In the present case study, distal pancreatectomy with splenectomy using an endoscopic linear stapler was successfully performed on a patient with concurrent SPTP and RPH. Our surgical experience in treating SPTP with RPH assisted with an endoscopic linear stapler will be helpful in the management of this rare clinical entity.

Case presentation

All protocols were performed in accordance with the Helsinki Declaration and approved

by the ethics committee of our university (YKD2016138). Written informed consent was obtained from our patient for publication of this case report and all accompanying images.

A 22-year-old woman was admitted to our hospital on 23 September 2016 with a 2-month history of a left upper abdominal apophysis. The abdominal bulge moved when the patient turned around in the lying position. B-Mode ultrasound examination at a local clinic showed a huge mass in the left upper abdomen; no treatment was performed at that time. The patient experienced occasional nausea but no vomiting, no obvious abdominal distension or pain, no chills or fever, and no jaundice. Additionally, her mental activities and appetite were normal with no notable changes in her body weight. She reported no history of surgical trauma and no family history of cancer or genetic diseases. After admission to our hospital, physical examination showed normal vital signs and no obvious tenderness, muscle tension, or rebound tenderness. A left upper abdominal bulge was found with an estimated size of about 15 × 14 cm; it was palpable with a hard, clear boundary. Laboratory examination showed an alanine transaminase level of 6.4 U/L (reference range, 7–40 U/L), direct bilirubin level of 7.2 μmol/L (reference range, 0–6.8 μmol/L), uric acid level of 375 μmol/L (reference range, 150–350 μmol/L), alkaline phosphatase level of 22 U/L (reference range, 35–104 U/L), cancer antigen 125 (CA125) level of 40.70 U/ml (reference range, 1.0–35 U/ml), and hemoglobin level of

102 g/L (reference range, 115–150 g/L). B-Mode ultrasound showed a 44- × 122- × 128-mm cystic mass in the left abdomen with a clear boundary that was closely associated with the tail of the pancreas. The images indicated that the cystic mass in the left middle abdomen probably originated from the tail of the pancreas. In addition, a computed tomography (CT) scan showed a quasi-circular shadow on the left side of the upper abdomen, and the size of this shadow was about 11 × 13.6 cm (CT value, 37–52 HU) (Figure 1). An enhanced CT scan showed that the solid content on the margin of the upper and lower left abdominal mass manifested as moderate continuous reinforcement, and linear intensification was present within the mass

(Figure 2). The portal vein was thickened and the splenic vein was varicose (Figure 3). The examination results indicated the presence of a solid pancreatic mass in the left upper and lower abdomen, suggesting an SPTP and portal hypertension. An endoscopic examination was not performed because the patient declined it. Additionally, the CT scan revealed no varicose vein plexus in the stomach (Figures 1–3). Moreover, an endoscopic examination would have been associated with a bleeding risk in this patient. For these reasons, an endoscopic examination was not performed.

We considered that complete removal of the tumor was the best option for this patient because she was quite young and the low-grade malignant tumor was very

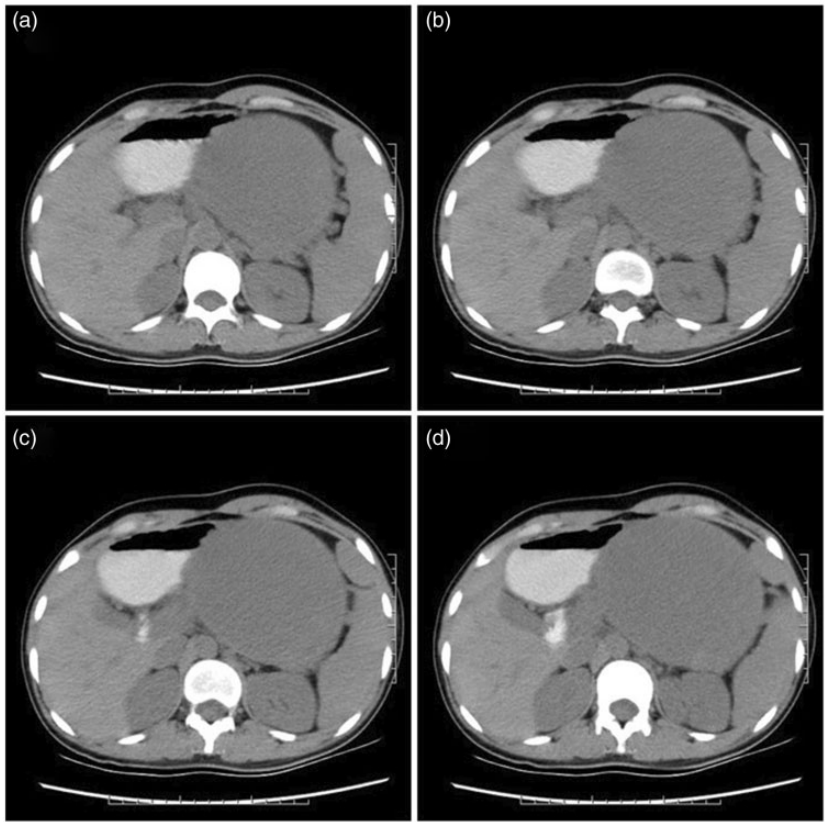


Figure 1. Preoperative non-contrast computed tomography scan showed a huge tumor.

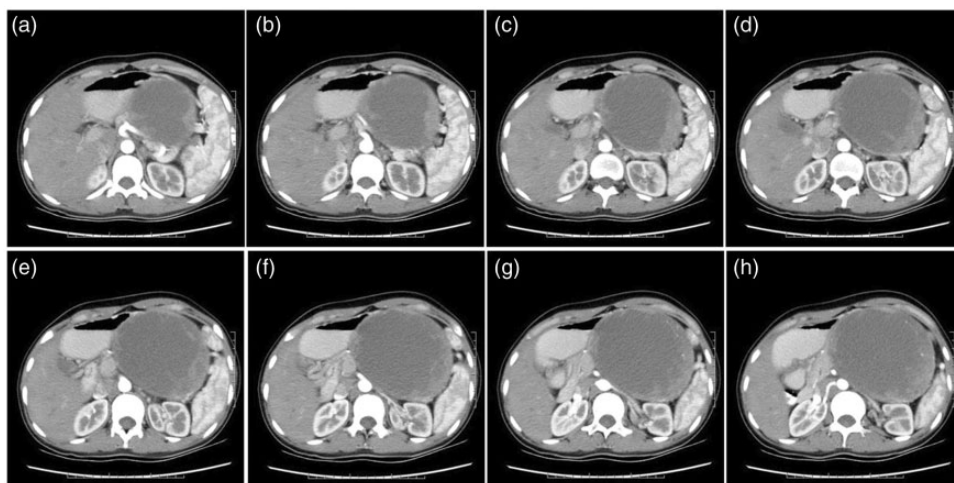


Figure 2. Preoperative enhanced computed tomography scan of the solid component of the neoplasm showed uneven enhancement in the arterial phase. No varicosity was observed in the stomach.

large. However, the adhesion between the splenic veins and tumor was tight. Distal pancreatectomy with spleen preservation would have been very difficult, possibly resulting in catastrophic bleeding. Therefore, the patient underwent resection of the giant tumor mass and pancreatic body and tail under intravenous anesthesia on 27 September 2016. Intraoperatively, we found that the spleen was enlarged and that a tumor measuring $14 \times 13 \times 13$ cm was present in the pancreas body with a hard texture and clear border. At the beginning of the operation, the surgeon opened the tumor membrane and dissected the lower part of the tumor until the lower edge of the pancreas and postpancreatic rarefaction interstitial, then opened the greater omentum of the gastric body until the spleen and stomach ligament could be ligated and resected. A venous plexus with an obvious varix in a large area was subsequently observed on the upper edge of the pancreatic body and posterior gastric wall. While avoiding touching the venous plexus, the surgeon cut the pancreatic membrane and opened it from the right side of the tumor to

the upper edge of the pancreas, and the unconsolidated space after the pancreas was then disassociated from the upper and lower edges of the pancreas at the neck. The pancreatic neck tissue and peritoneum were disassociated completely; therefore, the splenic artery and vein became free. The splenic arteries and veins were cut and closed with an endoscopic linear stapler (ECR60W, Echelon Flex 60; Ethicon Endo-Surgery, LLC, Cincinnati, OH, USA) at the pancreatic neck, 2 cm away from the tumor. The proximal end was sutured using a continuous suture line to ligate the blood vessels. The distal end of the pancreas and the tumor were raised up to the left of the rear of the dissociated pancreas. Here, the varicose veins of the upper edge of the pancreas and the posterior gastric wall were not touched. An artificial tunnel was formed through the right-back side of the varicose veins to the left side (incision site of spleen and stomach ligament). The endoscopic linear stapler was dipped into the tunnel and the varicose veins were cut and stapled. The entire pancreatic body and tail and spleen were

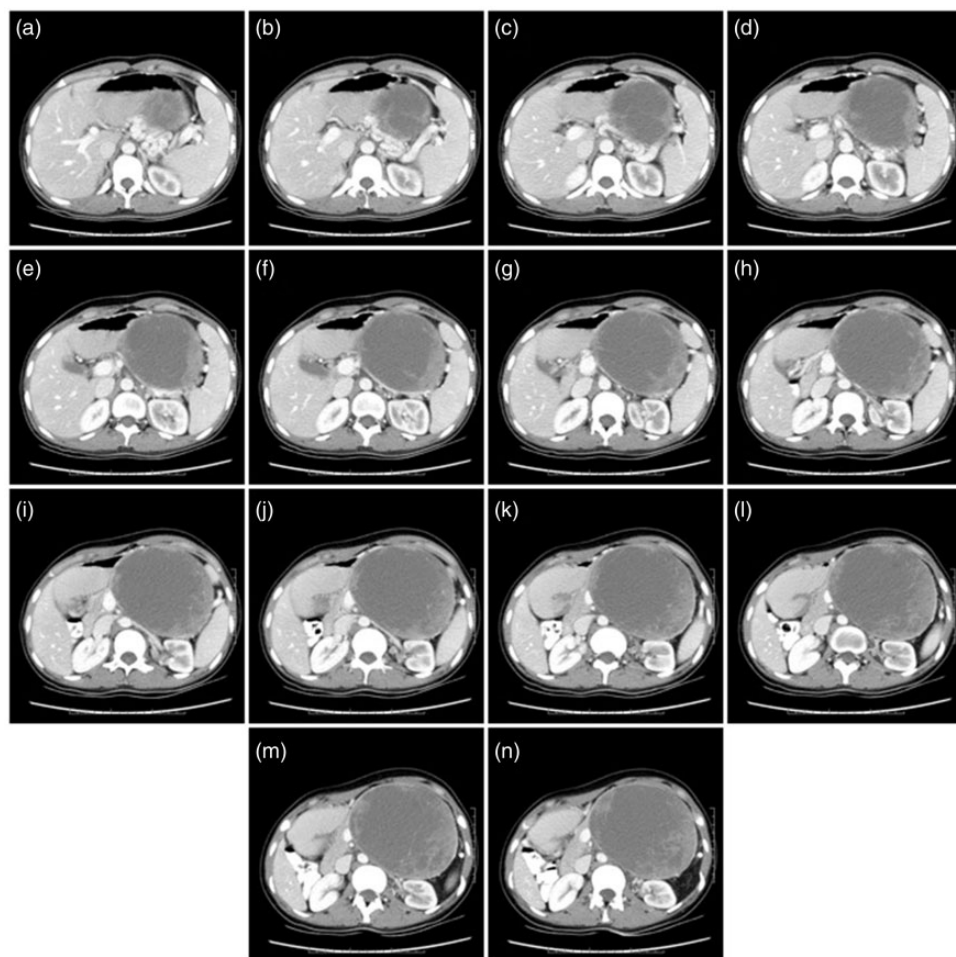


Figure 3. Preoperative enhanced computed tomography scan showed splenic varices and splenomegaly. A varicose vein plexus was present on the upper edge of the pancreatic body and posterior gastric wall.

removed. The tumor was then easily removed (Figure 4).

The removed tumor was a solid, pseudopapillary, and cystic mass. A large volume of bloody fluid was present in the cystic area (Figure 4). Hematoxylin and eosin staining indicated a solid pseudopapillary tumor (Figure 5). Immunohistochemically, SPTP generally exhibits expression of CD10, neuron-specific enolase, vimentin, progesterone receptor, α 1-antitrypsin, chymotrypsin- α 1, synaptophysin, and β -catenin as well

as local expression of other neuroendocrine markers.⁴ In the present case, most markers were positive except Ki67 and E-cadherin (data not shown). Synaptophysin- and chromogranin A-positive cells were occasionally found. Taken together, these results indicated the presence of SPTP. All antibodies were obtained from Fuzhou Maixin Biotech Co., Ltd. (Fuzhou, China). The immunohistochemical protocols were performed in strict accordance with the manufacturer's instructions.

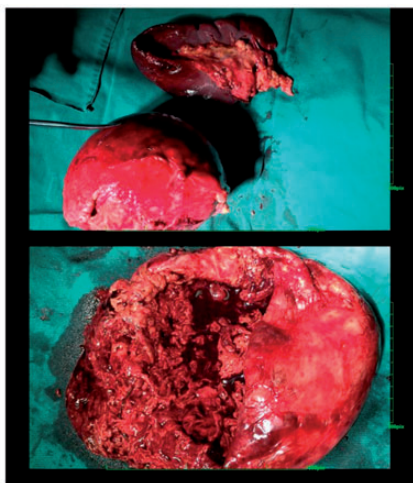


Figure 4. Photograph of the SPTP specimen after distal pancreatectomy and splenectomy showed the solid and cystic components.

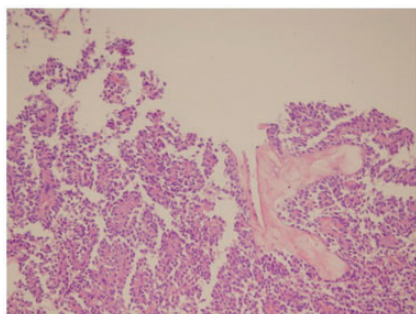


Figure 5. Representative hematoxylin and eosin staining of the tumor, indicating SPTP.

The patient's postoperative recovery was smooth. Postoperative diabetes is not uncommon due to the presence of a small amount of pancreatic tissue after the operation. In this case, the patient's fasting blood glucose level after resuming a normal diet was within the reference range. Six months and 1 year postoperatively, CT examinations were performed, and the findings are shown in Figures 6 and 7. No varicose vein was observed. At the 2-month follow-up, the patient reported

no discomfort, and her blood glucose level was normal.

Discussion and conclusions

This patient had an obvious pancreatic neoplasm on the upper edge of the pancreas, and a varicose vein plexus was clearly observed on the back of the neoplasm. The spleen was enlarged, but the function of the liver and the levels of hepatitis-related biomarkers were normal. Therefore, the patient was diagnosed with PPH. The patient was either asymptomatic or showed only symptoms of gastrointestinal tract compression when tumor enlarged in the early stage of SPTP. Serum tumor markers are often negative in patients with SPTP. In our patient, the CA125 level was slightly increased, which is consistent with the higher expression of CA125 in some pancreatic adenocarcinomas.⁵ The solid and cystic components of the SPTP were shown by CT and magnetic resonance imaging. Enhanced CT showed reinforcement in the solid mass and no reinforcement in the cystic mass, which is consistent with a previous report.⁶ Paz Soldán Mesta *et al.*⁷ described a 9-year-old child with an SPT localized in the pancreatic head who was treated by pancreaticoduodenectomy. SPTP with concurrent RPH is rare in the clinical setting. Li *et al.*⁸ reported 1 case of SPTP with RPH in a retrospective review of 34 cases of PPH. Because surgical resection is the better option for SPTP, the patient underwent surgery to remove the tumor. In general, surgical resection techniques include pancreaticoduodenectomy (Whipple resection), pancreatectomy combined with pancreaticojejunostomy, pancreatic resection combined with splenectomy, tumor topectomy, and others. In the present case, the tumor was huge and regional varicose veins were observable with a wide range. Therefore, a thorough understanding of unusual

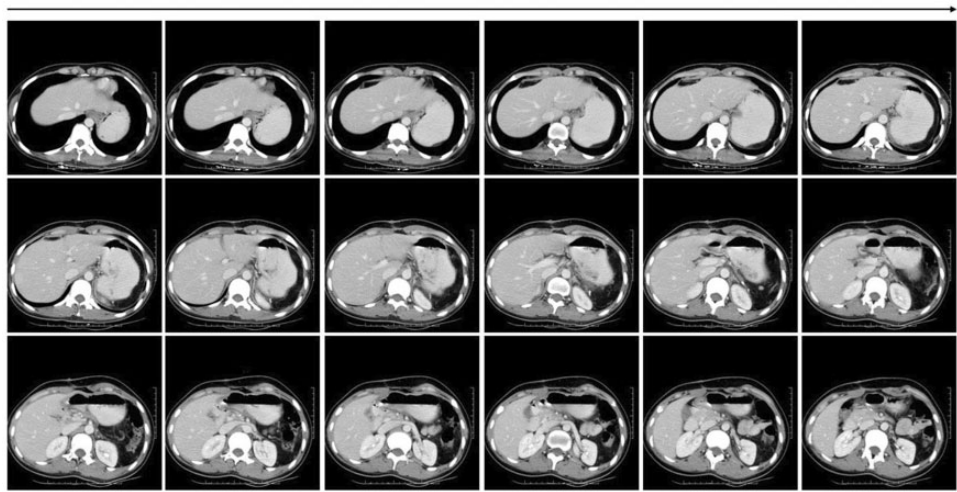


Figure 6. Six-month postoperative computed tomography images.

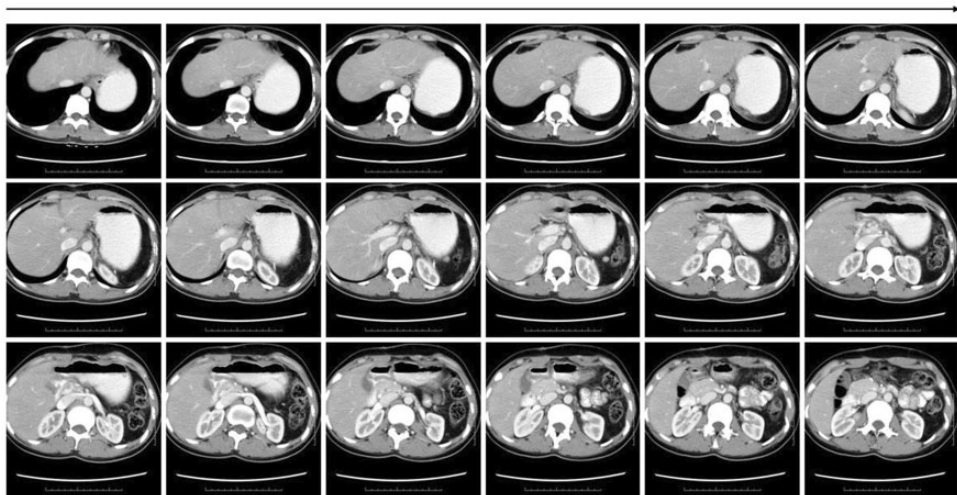


Figure 7. One-year postoperative computed tomography images.

presentations is important before the operation so that preparations can be made accordingly. Splenectomy is very difficult in patients with PPH because the peripancreatic adhesion is severe and the splenic hilum and surrounding parts are full of varicose veins, making the splenic artery, vein, and pedicle difficult to see.⁹ Such an operation is very dangerous because of the

risk of bleeding. Before the surgery, the surgeon should predict the difficulty of the procedure and make an emergency plan. In the present case, the patient's tumor was located in the tail of the pancreas. The lesions were present in the region of the splenic vein and the greater curvature of the stomach. The gastric fundus vein showed tortuous expansion. Considering

the facts that the patient had no gastrointestinal bleeding before surgery, preoperative CT showed clear expansion of the regional portal vein, the malignancy was low-grade, and the splenic veins were tightly adhered to the tumor, we decided to perform distal pancreatectomy and splenectomy in this patient. Preoperative vaccination for infectious complications after splenectomy was not performed in accordance with an expert consensus on the technical specifications of pericardial devascularization (2013 edition).¹⁰ Preoperative vaccination for an adult patient who undergoes splenectomy is optional in the clinical setting. To avoid bleeding when the varicose vein plexus was resected, we first completely dissociated the tissues at the pancreatic neck and posterior peritoneum, and the splenic artery and vein were suspended. They were then cut and closed with the endoscopic linear stapler at the pancreatic neck, which was 2 cm away from the tumor. Using this approach, the tumor and pancreatic body were easily raised for convenient resection. Initially, we tried to precisely separate and ligate the veins for resection. However, the operation was too difficult, and bleeding occurred with gentle tissue manipulation. Therefore, we switched the starting point and began from the left upper side of the tumor and moved inward gradually. An artificial tunnel was concurrently formed from the back side of the varicose vein plexus before separating it from the huge neoplasm and pancreas. The endoscopic stapler was then utilized to cut and close the varicose veins through the tunnel.

In conclusion, surgery in patients with RPH is considered difficult, and rigorous preoperative preparation is necessary. Endoscopic linear staplers are now widely used in diverse types of gastrointestinal surgery and show superiority in cutting the pancreas. In the present study, use of an endoscopic linear stapler was very useful

for tumor resection in a patient with RPH with a wide area of severe varicose veins.

Availability of data and materials

The data and materials used during the current study are presented in the main paper, and the immunohistochemistry data are available from the authors.

Authors' contributions

JY partly designed and performed the surgery; RX summarized the process and wrote the final manuscript. JZ assisted with the surgery and wrote a draft of the manuscript; ZW interpreted the computed tomography images; JR designed and performed the surgery. All authors carefully read and approved the final manuscript.

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Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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