CASE REPORT

Long-term Follow-up after Pancreatoduodenectomy with Portal Vein Resection for a Huge Solid Pseudopapillary Neoplasm in an Adolescent Girl

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A 16-year-old Japanese girl with a huge 13-cm-diameter tumor in the pancreas head presented with life-threatening symptoms and findings including severe anemia, obstructive jaundice, duodenal stenosis, and serious portal vein compression. She underwent a pancreatoduodenectomy with combined resection of the portal vein. Reconstruction of the portal vein was successfully performed using an external iliac vein graft and postoperative anticoagulant therapy. Pathological examination revealed a solid pseudopapillary neoplasm of the pancreas. The patient's postoperative course was uneventful, but her menstruation ceased for 14 months. She is now alive with no evidence of recurrence 100 months postoperatively and she suffers no impairments in daily activities of life. As a treatment of solid pseudopapillary neoplasms of the pancreas, pancreatoduodenectomy combined with portal vein resection is rarely performed in adolescent patients, but is reportedly successful, with patients tolerating the operation and surviving without recurrence. An aggressive surgical attitude is recommended when dealing with this tumor type with curative resection, even in adolescent patients. (doi: 10.2302/kjm.2014-0003-CR; Keio J Med 63 (3) : 52–59, September 2014)

Keywords: solid pseudopapillary neoplasm, pancreatoduodenectomy, portal vein resection, adolescent

Introduction

Solid pseudopapillary neoplasms (SPNs) of the pancreas are well known to have a benign character and to arise most often in young women.1,2 This tumor type is also described as a solid and cystic disease of the pancreas because of the variety of features of the macroscopic appearance of the tumor.2 We present herein a successfully resected case of a huge tumor arising at the head of the pancreas with life-threatening effects that was treated with pancreatoduodenectomy (PD) with combined resection of the portal vein and its reconstruction. We also discuss the technical aspects of the reconstruction of the portal vein and major invasive surgery in adolescent patients.

Case

A 16-year-old Japanese girl presented with vomiting in September 2005 and was admitted to our hospital. Physical examination revealed a huge mass in her upper abdomen with mild tenderness and anemic conjunctiva. Laboratory tests showed severe anemia with a hemoglobin level of 4.3 g/dl, leucocytosis (19300 /mm3), and mild elevation of aspartate aminotransferase (AST) (86 IU/l) and alanine aminotransferase (ALT) (108 IU/l). Compu-
Computed tomography identified a huge 13 × 11-cm solid tumor originating from the head of the pancreas (Fig. 1A). The tumor was round, well circumscribed, and showed inhomogeneous enhancement. Magnetic resonance imaging also visualized the huge tumor and identified a mixture of solid and cystic components. T2-weighted imaging showed a high-intensity area, suggestive of hemorrhagic pathology of the tumor. The portal vein and common bile duct were compressed and deviated ventrally. Hypotonic duodenography showed a compressed duodenum and poor passage. Angiography revealed a huge hypervascular tumor fed by both the superior and inferior pancreaticoduodenal arteries. Portography revealed severe compression, stretching, and narrowing of the portal vein, with the formation of collateral vessels (Fig. 1B). Endoscopic retrograde cholangiopancreatography showed the main pancreatic duct with a normal diameter at the head, but failed to enhance the distal part of the duct. The common bile duct could not be cannulated.

Jaundice developed on day 3 after admission and the level of total bilirubin increased to 3.3 mg/dl. The imaging findings strongly suggested a SPN of the pancreas, presenting with severe clinical problems of anemia and passage disturbances of the duodenum, common bile duct, portal vein, and pancreatic duct. The resection of the tumor was necessitated because of these life-threatening conditions. Open laparotomy was carried out in October 2005. A huge soft tumor, 13 cm in diameter, was located at the head of the pancreas (Fig. 2). The tumor was round and encapsulated with expanding growth characteristics. The duodenum, common bile duct, and portal vein were riding on the tumor and all were compressed and deviated ventrally, as visualized preoperatively. The portal and superior mesenteric veins were involved and were flattened on the tumor for a distance of about 8 cm. Portal collateral vessels had developed at the liver hilum and hepatoduodenal ligament. No distant metastasis was found. PD with combined resection of the portal vein and Child’s reconstruction were performed. The procedure took 18 h and involved 2970 mL of blood loss. Portal vein reconstruction was achieved using a right external iliac vein graft under portal clamping for 36 min. The pancreatic duct was intubated with a 6-French tube and totally drained, and a pancreaticojejunostomy was performed with the invagination method.

Heparin was continuously infused as postoperative anticoagulant therapy with a daily dose of 10000 units starting just after the operation and continuing for 16 days; heparin was followed by warfarin administration for a further 38 days. Pathological examination revealed a huge encapsulated tumor, 11 × 10 cm in size; the solid cut surface demonstrated hemorrhage and necrosis (Fig. 3A). The duodenum, common bile duct, and portal vein were markedly compressed and were involved, but were not directly invaded by the tumor. Microscopically, the tumor consisted of eosinophilic polygonal neoplastic cells arranged in a solid and pseudopapillary pattern (Fig. 3B). Immunohistochemically, the neoplastic cells were positive for alpha-1-antitrypsin, neuron-specific enolase, vimentin (Fig. 3C), and nuclear accumulation of beta-catenin (Fig. 3D). No lymph node metastasis was detected microscopically. The final pathological diag-

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**Fig. 1** (A) Enhanced computed tomography before surgery: a huge (13 × 11 cm), round, solid tumor is demonstrated at the pancreas head. The tumor has a clear boundary and expanding growth, markedly compressing the portal vein and adjacent organs. (B) Portal phase of superior mesenteric arteriography. The portal vein and superior mesenteric vein are markedly stenotic as a result of compression by the tumor.
PD with Portal Vein Resection for SPN

Diagnosis was SPN. The patient’s postoperative course was mostly good, but minimal right leg edema occurred for a few days postoperatively. She was able to return to school soon after discharge from the hospital on postoperative day 35.

The patient’s body weight recovered to the preoperative level at 9 months. Amenorrhea developed postoperatively, but her menstruation reappeared first at 14 months. Follow-up computed tomography showed no finding of recurrence and good patency of the portal vein (Fig. 4). The serum levels of fasting blood glucose (86 mg/dL) and glycosylated hemoglobin A1c (5.6%) were normal. Currently, at 100 months after surgery, she is well and works in an office.

Discussion

SPN was first described as a papillary tumor of the pancreas by Frantz in 1959. It has a definitive pathological description, namely, “a benign or low-grade malignant epithelial tumor which occurs predominantly in young women and is composed of monomorphous cells variably expressing epithelial, mesenchymal, and endocrine markers.” Many pathologists have expressed a great deal of interest in this type of tumor, mainly because of its morphologic characteristic features of a mixture of solid and cystic components macroscopically and a solid and papillary pattern microscopically. Because of these characteristics, SPN has many synonyms, such as “solid-cystic tumor,” “papillary-cystic tumor,” and “solid and papillary epithelial neoplasm.” Another point of pathological interest is that the tumors characteristically demonstrate multiple expressions of acinar cell and neuroendocrine differentiation, and therefore, it has been suggested that the origin of the tumor is a pluripotent stem cell. Clinically, the disease often affects young adolescence girls and has a good prognosis in resected cases. These clinical characteristics thus encourage surgeons to treat SPN aggressively.

The SPN in our case was pathologically typical, but had several specific characteristics: (1) it was accompanied by severe anemia, (2) huge tumor size, (3) presentation with vomiting, (4) development of jaundice, and (5) severe portal vein involvement.

First, severe anemia was noted. SPNs tend to exhibit tumor necrosis and hemorrhage, and the pathological causes of anemia associated with SPN may be hemorrhage within the tumor or duodenal bleeding via the pan-

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**Fig. 2** Operational view. The tumor at the pancreas head is huge and is compressing the surrounding organs. The portal vein (PV) is stretched, flattened, and severely narrowed for 8 cm of its length. The pancreas has already been cut at the body and the pancreatic duct cannulated. SMV, superior mesenteric vein.
creatic duct or the papilla of Vater. Duodenal mucosal bleeding was not likely in our case because of the lack of direct invasion of the duodenum. The severe anemia was life-threatening and was an important factor in the indication of resection.

Second, the mean sizes of SPNs are reportedly in the range 8–10 cm² and 6.08 cm. Therefore, the size of 13 cm in our case was large compared with other reported cases and meant a great deal of tumor pressure against adjacent organs, which resulted in the subsequently mentioned compressive morbidities. Third and fourth, as a result of compression by the tumor, the patient presented with vomiting as the primary symptom, resulting from duodenal stenosis, and preoperatively developed obstructive jaundice due to choledochal stenosis. These problems would have worsened if an operation had not been carried out; therefore, they supported the indication of surgical intervention.

Finally, compression of the portal venous system was the most significant and potentially lethal problem in the present patient. The compression was serious because the portal vein and superior mesenteric veins were flattened and involved within the tumor capsule for a distance of 8 cm, and collateral vessel formation was identified. This condition also resulted from the large size of the tumor. Portal vein obstruction is well known in pancreatic cancer and is often managed with combined resection and reconstruction. However, collateral vessel formation in the portal system in pancreatic cancer is a sign of poor prognosis and suggested that our patient was an inappropriate candidate for tumor resection. For SPNs, portal

Fig. 3 (A) Gross appearance of the tumor. The tumor is encapsulated, 11 × 10 cm in size, solid, and inhomogeneous at the cut surface, suggesting the presence of hemorrhage and necrotic regions. The defect in the capsule at the left upper side is an artifact from processing the resected specimen. (B) Microscopic appearance of the tumor (hematoxylin and eosin stain, ×100). In the solid tumor, the epithelial neoplastic cells with low-grade atypia are arranged in a pseudopapillary pattern with a fibrovascular core. This is a typical finding for a solid pseudopapillary neoplasm of the pancreas. (C, D) Immunohistochemically, the cells stain positively for vimentin (C, ×400) and show nuclear accumulation of beta-catenin (D, ×400).
vein resection has been reported in only a few cases. In 1983, Dales et al. reported a case of SPN in the pancreas body treated with total pancreatectomy and combined resection of the portal vein. Jeng et al. reported in 1993 the first SPN patient (a 28-year-old woman with a 10-cm tumor) who underwent PD and combined resection of the portal vein; the patient died of unrelated hepatic encephalopathy 66 months after the initial surgery. Chen et al. reported the second such case, which involved an 11-year-old girl with liver metastasis who underwent PD with portal vein resection and resection of metastatic lesions of the liver; the patient survived 3 years without tumor recurrence. Chen et al. recommended aggressive surgery including portal vein reconstruction and metastasectomy. Thus, ours was the third case of PD with portal vein resection for SPN.

The excellent prognosis for patients with SPN has been mainly achieved by tumor resection, even in patients with portal vein involvement. Our patient had an excellent outcome, being alive without recurrence 100 months after surgery. In SPN, the mechanism of portal vein involvement seems to be mainly compression, whereas pancreatic cancer tends to invade the portal vein wall. Thus, the reason for the favorable prognosis of resected cases of SPN with portal vein involvement is thought to be that portal vein involvement does not mean extensive spreading of tumor cells. We emphasize that because most SPNs have a relatively benign character of tumor growth, aggressive surgery is recommended, despite the involvement of major vessels. Even under metastatic conditions of SPN, aggressive hepatic resections have been reported and recommended with successful results.

For portal vein reconstruction, the safety of the operation should be thoroughly established during the perioperative period. We performed external iliac vein grafting during portal vein reconstruction because direct anastomosis of the portal trunk and superior mesenteric vein would have been risky because of resulting tension caused by the 8-cm defect in the length of the vessel. We selected an autograft to achieve a tension-free anastomosis and to obtain long-term functionality. Because the vein graft was substantially long and there were two anastomotic sites, we used anticoagulant therapy postoperatively in the present case. However, it remains controversial whether therapeutic anticoagulant reduces the rate of thrombotic complications after venous reconstruction.

Operative indications and postoperative characteristics concerning PD in adolescent patients with SPN are not well known. There are few papers analyzing the effect or harm of invasive PD on teenage patients. We have summarized reported cases of PD for SPN in adolescent (i.e.,

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Fig. 4 Follow-up computed tomography at 88 months demonstrating no evidence of recurrence and good patency of the portal vein.
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<th>No.</th>
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ND, not described; IVC, inferior vena cava; PD, pancreatoduodenectomy; PPPD, pylorus-preserving pancreatoduodenectomy; Htx, hepatectomy; PV, portal vein; QOL, quality of life; IDDM, insulin-dependent diabetes mellitus.
teenage) patients in Table 1.5,14–28 Almost half of the cases were reported in Asian countries. Some patients had serious preoperative morbidities other than abdominal mass, e.g., choledochal stenosis, duodenal stenosis, and hemorrhage, similar to our case, and were thus obligated to undergo surgery. Only two cases (No.12 and our patient) underwent combined resection of the portal vein. In-hospital postoperative courses were uneventful in the majority of cases, a fact that endorses the aggressive approach to the treatment of SPN. All patients survived, mostly without recurrence. While the median follow-up period of the reported cases is 48 months, the present case has the second longest time of observation (100 months).

Less invasive surgery should also be considered, especially in adolescent patients. Akiyama et al. reported a successful case of duodenum-preserving pancreas head resection for SPN.28 In a Japanese case (No. 8), pancreatic insufficiency and insulin dependent diabetes mellitus occurred after adjuvant chemotherapy for 2 years.21 Adverse reactions to anticancer drugs were likely involved in the pathogenesis of these pancreatic disorders. In the present report, we described the complication of transient amenorrhea for 14 months after PD. Menstruation disturbance probably resulted from nutritional impairment after surgery, and the reappearance of regular menstruation may be a sign of full recovery after major surgery in adolescent girls. Looking at Table 1, we can see that adolescent patients were often obligated to undergo major surgery, i.e., PD, but tolerated it well and recovered from SPN.

The outcome after surgical resection for SPN is excellent, with more than 90% enjoying long-term survival. Recurrence is reported in approximately 5% of cases, and typical recurrence sites are the liver, lymph nodes, and peritoneum.5,29 There is general consensus that surgical debulking should be recommended for these metastases, in contrast to other pancreatic malignancies.

In conclusion, we reported a very rare case of combined resection of the pancreas head and portal vein for SPN in an adolescent girl. The operation was necessary because of life-threatening conditions caused by severe anemia due to tumor hemorrhage and by the huge mass of the tumor resulting in compressive morbidities including duodenal stenosis, obstructive jaundice, and severe portal vein stenosis. The present patient is now alive with no evidence of recurrence 100 months postoperatively; she has no impairments in daily activities of life. Aggressive major surgery, including PD and portal vein reconstruction, should be performed even in adolescent patients if it is necessary to relieve-life-threatening conditions or to cure neoplastic conditions, especially for tumors having a better prognosis such as SPNs.

References


