Pediatr Surg Int (2009) 25:617–621
DOI 10.1007/s00383-009-2388-9

ORIGINAL ARTICLE

Is laparoscopic management suitable for solid pseudo-papillary tumors of the pancreas?

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Accepted: 15 May 2009 / Published online: 29 May 2009

Abstract
Purpose Solid pseudo-papillary tumors (SPT) are rare pancreatic neoplasms of low-malignant potential occurring mainly in young women. The purpose of this report is to describe our experience with laparoscopic management of these tumors with 4-year follow-up.
Methods Three children with SPT were admitted to two hospitals in Paris, France, between February 2000 and December 2006. Diagnosis or treatment was carried out using laparoscopic techniques (biopsy and resection in one case and biopsy only in two). Long-term follow-up data were collected.
Results All three patients presented recurrences within 3 years after resection, i.e., disseminated peritoneal recurrence in two patients and local recurrence in one. The two patients with peritoneal recurrences were treated by surgical resection and chemotherapy. The patient with local recurrence could not be treated due to contraindicating local factors. All three patients were alive at the time of this writing.
Conclusion This is the first report describing long-term follow-up after laparoscopic management of SPT. All three patients developed recurrences. These poor results contrast sharply with the low risk of local or disseminated recurrence after open laparotomy without chemotherapy that has been considered as the treatment of choice up to now. Recurrences after laparoscopic management may have been due to diffusion of tumor cells caused by gas insufflation especially during biopsy. Laparoscopic biopsy should not be performed in patients presenting SPT.

Keywords Frantz's tumour · Pancreas · Cysto-papillary tumour · Laparoscopy · Child · Reccurence · Peritoneal carcinosis

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Introduction

Solid pseudo-papillary tumors (SPT) are rare pancreatic tumors of low malignant potential occurring mainly in women before the age of 35 [1, 2]. The first author to describe this tumor was Frantz [3] in 1959. However, it was only recently, i.e., in 1996, that the World Health Organization board proposed the name SPT for the international histological classification of tumors of the exocrine pancreas. Since adenocarcinoma of the pancreas rarely affects children, differential diagnosis of a pancreatic mass in young patients includes pancreatic cyst (or pseudo-cyst), endocrine pancreatic tumor, pancreatoblastoma, and SPT. Histological diagnosis can be difficult due to the inconsistent features of SPT [4–7]. Histogenesis of

this tumor is still unclear with acinar, ductal, endocrine, and primordial cell origins having been proposed [6]. Several studies have demonstrated the low malignant potential of these tumors, and surgical excision is considered as the treatment of choice [8–10]. Specific mortality is lower than 2%, and only 10–15% of patients present local recurrence or metastases after resection. Recently several teams have reported good immediate results using laparoscopic techniques for either diagnosis (biopsy) or treatment of SPT [12, 13]. The purpose of this report is to describe long-term outcome of laparoscopic management of SPT in three young patients between February 2000 and December 2006.

Patients and methods

This study includes all patients in whom laparoscopy was used for diagnosis and/or treatment of SPT in the pediatric surgical departments of the Robert Debré Hospital and Necker Enfants-Malades Hospital in Paris, France, between February 2000 and December 2006. The following data were collected: age at time of diagnosis, tumor location, preoperative biopsy findings if available, surgical procedures, and follow-up findings including mode, nature, treatment and outcome (e.g., recurrence or not).

Results

Between February 2000 and December 2006, laparoscopic techniques were used for diagnosis or treatment of SPT in three patients operated on by two surgeons (PDL and SS). In all three cases clinical and laboratory work-up failed to achieve differential diagnosis and pre-resection biopsy was performed.

Patient no. 1

A 9-year-old boy was admitted for abdominal pain. Ultrasonography revealed a 2-cm tumor in the pancreatic tail. Diagnostic laparoscopy was performed with grasping forceps to obtain a specimen for extemporaneous frozen section biopsy. Since results failed to determine if the lesion was benign or malignant, laparoscopic resection using an endobag extractor was performed in the same procedure. Conservation of the spleen was possible since there was no spleen vessel involvement. Histological examination of the surgical specimen demonstrated a partially encapsulated 2.2 cm × 1.7 cm SPT tumor infiltrating the healthy pancreas. Surgical margins were negative and lymph nodes included in the specimen were not metastatic (Fig. 1).

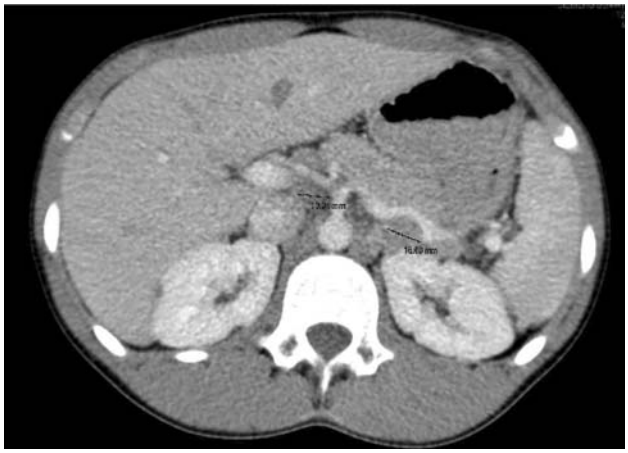

Fig. 1 CT scan showing two coelio-mesenteric nodules due to recurrent SPT

Forty-three months after resection, routine ultrasonography detected a recurrence. Computed tomography confirmed a disseminated peritoneal recurrence and abdominal nodes. PET scan showed local uptake. Four courses of chemotherapy (5FU + oxaliplatine + irinotecan) were administered with no effect on tumor volume (stable disease) or local uptake on PET scans. Laparotomy was performed with resection of all nodules (no macroscopic residual tumor). Histology confirmed recurrent SPT. The patient was disease-free on follow-up examination 1 year after resection of the recurrence.

Patient no. 2

A 11-year-old girl was admitted for a pancreatic tumor discovered incidentally during abdominal ultrasound examination performed 2 months after renal transplantation for congenital renal hypoplasia. Examination of a biopsy specimen obtained by diagnostic laparoscopy using grasping forceps confirmed SPT. Twenty days after diagnosis distal pancreatectomy was performed via open laparotomy. As in patient no. 1, the spleen showed no vessel involvement and was conserved.

The final histology report indicated a 4.5-cm SPT tumor that despite being well-encapsulated had infiltrated the healthy pancreas as well as peri-pancreatic fatty tissue. Resection was considered as complete since surgical margins were negative, and lymph nodes included in the specimen were not metastatic.

Forty months later numerous peritoneal nodes were detected during routine follow-up ultrasonography. Diagnostic laparoscopy confirmed peritoneal recurrence. Open laparotomy was performed with removal of most lesions and the transverse colon that was involved (R0 surgery). Surgery was followed by six courses of chemotherapy

(5FU + oxaliplatine + irinotecan, protocol folfirinox) [14]. The patient was in complete remission at follow-up examination 43 months after resection of the recurrence.

Patient no. 3

A 12-year-old girl was admitted for haematemesis and melaena. Endoscopy found a duodenal ulcer and abdominal ultrasonography revealed a heterogeneous mass in the head and body of the pancreas. Examination of a biopsy specimen obtained by laparoscopy led to diagnosis of SPT. Ten days after diagnosis subtotal resection of the tumor was performed via surgical laparotomy. Due to the closeness of the lesion to the duodenal wall, mesenteric superior vein and spleen vessels, the posterior part of the pseudo-capsule was not removed to avoid having to perform complete duodenopancreatectomy. Neither the spleen nor tail of the pancreas sparing could be conserved due to the extensive vessel involvement. The pylorus could not be spared due to involvement of the duodenal wall. Histological examination confirmed diagnosis of SPT. After multidisciplinary case review, it was decided not to perform chemotherapy.

Thirty-two months after resection, the patient presented anorexia and asthenia. Abdominal CT-scan revealed a 60 × 45 × 45 mm recurrence of the tumor in the head of the pancreas. Thrombosis of the splenic and inferior mesenteric vein was also observed. Laboratory tests demonstrated exocrine and endocrine pancreas insufficiency. Cephalic pancreatoduodenectomy was planned but could not be performed because loco-regional extension led to complete thrombosis of the portal vein. Liver transplantation was also contraindicated. Total excision of the tumor was not possible because of adherence to major blood vessels. Two years after the last surgical procedure, the patient was still alive and had a normal active lifestyle despite intermittent abdominal pain.

Discussion

Although SPT is usually regarded as a low-malignant tumor, our three cases and perusal of the literature underline the potential aggressiveness of SPT and the necessity for careful surgical management. The treatment of choice for SPT is surgical resection since local recurrence is rare (10%) after complete removal [10, 15]. This low recurrence rate contrasts sharply with the 100% rate observed in our three patients in whom laparoscopy-assisted management was used. These poor results have led us to reconsider our initial advocacy of laparoscopic management of SPT based on experience in patient 1 [11]. To obtain a better idea about these issues, we reviewed the literature regarding predictors of peritoneal recurrence of SPT, methods used for pre-operative biopsy of SPT, and risks associated with laparoscopic management of other tumors.

Complete resection of SPT is considered as mandatory especially in presence of metastases [16], but this attitude can be tempered in light of the tumor's low malignant potential [10]. If the lesion is located in the pancreatic tail or body, distal pancreatectomy can easily be performed and the need for splenectomy is debatable. However, for tumors located in the head of the pancreas as in our patient 3, the decision to perform duodenopancreatectomy can be problematic. In these cases the potential risk of diabetes or pancreatic insufficiency [18] must be carefully evaluated based on pre-operative assessment of pancreatic function and of the quantity of residual healthy tissue after resection.

Recurrences or metastasis of SPT occur preferentially in the liver and peritoneum [16]. Some teams report that prognosis is worsened by male gender, advanced age and histological findings such as tumor necrosis and presence of a sarcoid component or undifferentiated tissue [6]. Tumor size and infiltration of the healthy pancreas [17] have also been reported as risk factors for recurrence after incomplete resection as was the case in one of our patient 3.

Patients 1 and 2 in our series presented disseminated peritoneal recurrences with multiple masses. Since peritoneal carcinosis of SPT is considered as uncommon [19–21], this finding is puzzling. Three possible explanations can be proposed, i.e., highly aggressive tumor behavior, incomplete resection, or laparoscopic-related causes. The first two explanations seem unlikely. The pathologic features of the lesions in our patients were not different from those of previously described SPT and no histological factor could account for the aggressive behavior observed in patients 1 and 2. Although both patients had infiltration of healthy tissue, operative outcome was considered as satisfactory and surgical margins were negative. Thus, the third explanation, i.e., laparoscopic-related causes, appears to be the most plausible.

As all three patients in this series underwent diagnostic laparoscopy, it seems reasonable to implicate this procedure in recurrence. In this regard, it is interesting to note that several teams have reported their experience using other biopsy techniques for diagnosis of SPT. No complications including recurrence have been observed using ultrasound-guided fine needle aspiration and percutaneous biopsy [14, 23]. Similarly no recurrence has been reported after surgical biopsy [24].

The deleterious role of pancreatic trauma in SPLT was underlined in a report by Levy et al. [19]. Cancer dissemination during laparoscopic surgery has been reported previously [22]. Both surgical and laparoscopic techniques require breaking of the tumor capsule, but only laparoscopy

involves insufflation that could lead dissemination and spreading as in the case of traumatic injury. This difference could account for the recurrences observed after laparoscopic management in patient 1 (diagnosis and resection) and patient 2 (diagnosis only). Patient 3 who underwent laparoscopic biopsy followed by resection with positive surgical margins via laparotomy developed local recurrence rather than peritoneal recurrence. This would rule out the possibility that tumor cell seeding due to dissemination during surgical biopsy under laparoscopy.

In a previous report, we presented a pediatric case of recurrence after laparoscopic management in a patient presenting a huge ovarian teratoma with a large cystic component. Preoperative findings were favorable (αFP normal and no calcification on U.S.) and laparoscopy allowed drainage and removal of the tumor. Nevertheless, the child developed a recurrence in association with peritoneal carcinosis (article published in French).Similar recurrences have been reported in adults [32]. Based on these findings laparoscopy is currently not indicated in case of mixed ovarian tumor (cystic and solid components).

Laparoscopy-assisted techniques have been used for management of various pancreatic tumors. Several studies have established laparoscopy as the state of the art technique for pancreatic adenocarcinoma in adults [28]. Laparoscopic surgery is now recognized as safe for both diagnosis and treatment of adrenal neuroblastoma at all stages in adults and recent evaluation in children indicates that laparoscopy is feasible especially at stage I that has been treated exclusively by surgery up to now [29]. The value of pancreatic laparoscopic surgery has also been documented for pediatric management of endocrine tumors [25], pancreatic cysts [26], and pseudo cysts [27]. Little data have been published on laparoscopy for management of pancreatoblastoma and SPT in children.

Laparoscopy may be useful for management of cystic or endocrine pancreatic tumors in children. A larger series will be needed to evaluate the safety and efficiency of laparoscopy for management of tumors like SPT without chemotherapy. However, based on our experience we cannot recommend laparoscopy biopsy for diagnosis of SPT in pediatric patients. Since disseminated recurrence has never observed after biopsy via the open surgical technique, we think that laparoscopic approach was responsible for the complications observed in two of our three cases. This procedure may increase the risk of peritoneal dissemination. Percutaneous biopsy by a posterior approach or endoscopic ultrasound-guided fine-needle aspiration may be considered as an alternative to primary resection. With regard to fine needle aspiration, it should be noted that a quantity sufficient for diagnosis is obtained in 75% of cases [23].

Use of a new technique must not worsen the prognosis of diseases that can be successfully managed using conventional techniques. For this reason, careful assessment is necessary to compare the outcome of laparoscopic surgery to that of open surgery that considered as the gold standard for many pancreatic tumors. A good illustration of the lengthy validation process for new techniques involves laparoscopy versus laparotomy for colonic cancer surgery in adults. It is only now that a consensus is emerging about the benefit of laparoscopy in expert hands [30, 31]. Defining the utility of laparoscopy for pancreatic tumors in children is a slower process due to the infrequency and pathologic variability of pediatric pancreatic tumors. This underlines the need for sharing experience in this field.

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