Solid pseudopapillary tumor of the pancreas: a rare entity

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Solid pseudopapillary tumor (SPT) of the pancreas is a rare neoplasm in children that mainly occurs in young females. We herein report a rare case of SPT arising from the tail of the pancreas. A 13-year-old girl was admitted to our clinic with abdominal pain and anorexia. A mass was palpated on the physical examination. A 90x72 mm, encapsulated, heterogeneous mass with solid and cystic components was defined on computerized tomography (CT). Distal pancreatectomy was performed during the operation. Histopathological examination revealed that the tumor was a SPT with negative surgical margins. A six-month follow-up after surgical resection showed no evidence of recurrent disease. SPT should always be considered in the differential diagnosis in a young female with a palpable mass.

Key words: solid pseudopapillary tumor, pediatric, pancreatic tumors.

Pancreatic neoplasms are exceedingly rare in children. It has been reported that they account for less than 0.2% of deaths from cancer in children[1]. Solid pseudopapillary tumor (SPT) is a histologic subtype of pancreatic neoplasms (2-3% of primary pancreatic tumors occurring in all ages)[2]. The first case of SPT was described by Frantz in 1959[3]. Patients with SPT usually present with nonspecific clinical symptoms and without abnormalities in clinical laboratory tests, and SPTs are more common in young females[2]. Even though there are possible histological findings of malignancy, SPT typically shows a benign clinical course and a low malignant potential. We herein report a rare case of SPT that was treated with complete surgical resection.

Case Report

A 13-year-old girl, previously well, presented with a one-month history of abdominal pain and anorexia. The physical examination showed a firm, well-palpable, painless mass in the left upper quadrant of the abdomen. Blood laboratory tests, including tumor markers, were normal (α-fetoprotein, carcinoembryonic antigen, β-human chorionic gonadotropin). An abdominal ultrasound (US) showed a 90x72 mm diameter solid tumor. An abdominal computerized tomography (CT) demonstrated a well-defined, encapsulated, heterogeneous mass with solid and cystic components, as well as hemorrhagic areas (Fig. 1). The mass was located between the tail of the pancreas, left kidney and spleen. A mass originating from the tail of the pancreas and a mesenteric cyst were considered in the differential diagnosis. During the operation, a solid mass originating from the tail of the pancreas was found (Fig. 2). The mass was lobule and there were areas of hemorrhage; it was resected completely with distal pancreatectomy. Histopathologically, the tumor was defined as SPT. The tumor was resected en bloc, was encapsulated, and the specimens demonstrated the characteristic areas of hemorrhage. No vascular invasion was found. Surgical margins were negative (Figs. 3a, 3b). The patient showed an uneventful postoperative course. During our four-month follow-up, no evidence of disease was seen.

Discussion

Malignant pancreatic neoplasms are extremely rare in children, with less than 20 patients reported, even in the reports from large referral centers spanning 20 years of experience[4]. Malignant pancreatic tumors in pediatric patients include pancreatoblastoma, acinar cell carcinoma, ductal adenocarcinoma, SPT, pancreatic neuroendocrine tumor, sarcoma,

lymphoma, and cystadenocarcinoma[5].

Our patient presented with a palpable mass, abdominal pain and anorexia. Park et al.[4] reviewed the clinical features and outcomes of children with pancreatic neoplasms who were treated at their center, and they found that most of the patients with pancreatic tumors presented with abdominal pain or a palpable abdominal mass. Serum tumor markers were all within normal values in our case. Similarly, in previous studies, serum tumor markers were not found to be predictive of SPT[3]. SPT was located in the tail of the pancreas in our case, which is the most common location mentioned in the literature (35.9%-44%)[3]. Aggressive resection is advocated in the treatment of SPT, although it is relatively indolent compared to other malignant pancreatic tumors. Complete resection of the tumor with distal pancreatectomy was performed in our case. In the literature, distal pancreatectomy was performed in SPT localized in the tail of the pancreas, while pancreaticoduodenectomy was performed when the SPT was in the head of the pancreas[3]. Zampieri et al.[6] performed a duodenum-preserving pancreatic head resection (Berger procedure) for a SPT located in the head of the pancreas and splenopancreatectomy using a Roux-en-Y pancreatic jejunostomy for the body/tail tumors. Furthermore, Uchida et al.[7] performed laparoscopic spleen-preserving distal pancreatectomy with conservation of splenic vessels in a child with SPT. In contrast, Campanile et al.[8] advocated that laparoscopy is not indicated for the treatment of SPTs because of possible spray effect and consequent peritoneal carcinosis risk. They suggested complete surgical resection in locally invasive SPT, even if it requires difficult and mutilating surgery.

Park et al.[4] found that primary tumor resection resulted in a 100% cure rate, regardless of tumor size, even in patients positive for tumor at the resection margin. Their findings indicate that, despite the large tumor size and its ability to extend locally, complete excision can be beneficial in most patients. Metastases are uncommon in SPT (10-15% of patients with advanced disease)[5]. Although complete resection was more frequently performed on patients with SPT, Rojas et al.[5] reported that positive margins did not affect the outcome of

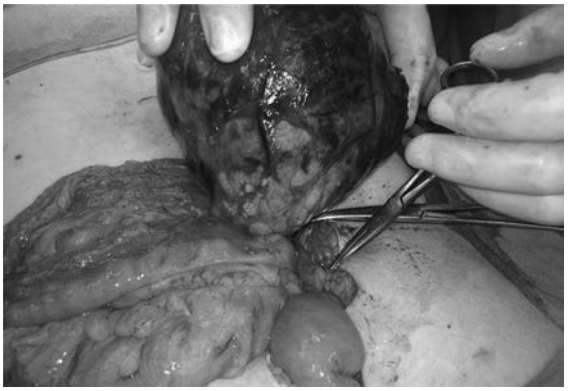

Fig. 1. Enhanced coronal CT images show a well-defined, heterogeneous, minimally enhanced mass in the tail of the pancreas.

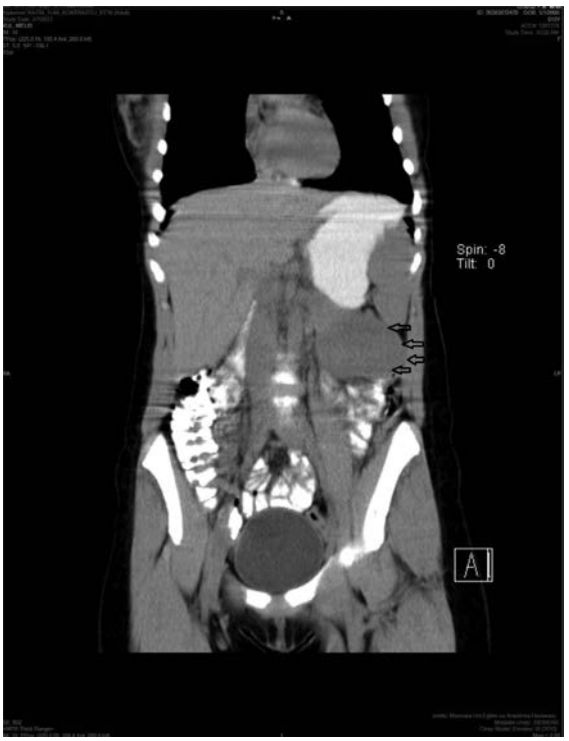

Fig. 2. Solid pseudopapillary tumor in the tail of the pancreas.

patients with SPT, suggesting that enucleation when feasible, rather than radical resection, is likely sufficient to achieve long-term survival. The role of chemotherapy and radiation is controversial, but it has been used in some cases with aggressive disease[5].

Solid pseudopapillary tumor (SPT) (Frantz tumor) is a papillar-cystic tumor. Histologically, no acinar or ductal structures appear to be present. Degenerative changes result in the

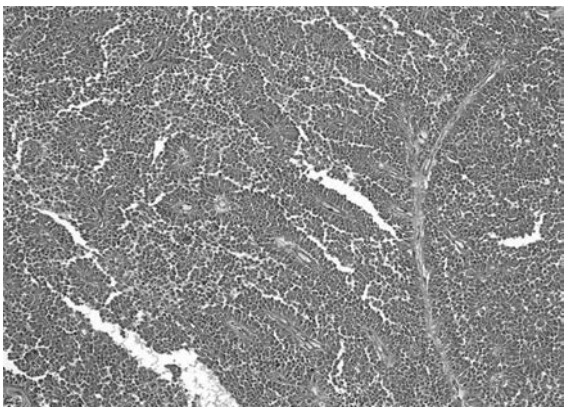

Fig. 3a. Perivascular pseudopalisading pattern with fibrovascular cores and septae, typical of solid pseudopapillary tumor (hematoxylin & eosin (H&E) stain, X100).

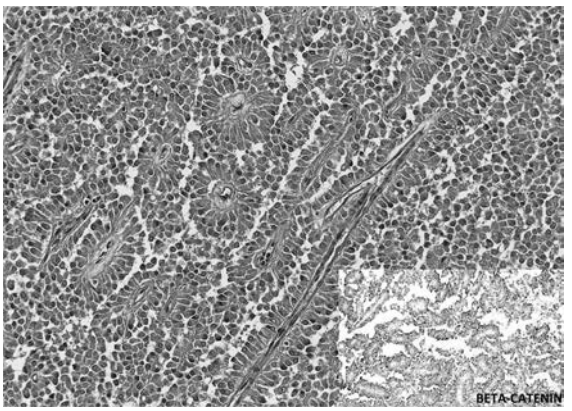

Fig. 3b. Detaching tumor cells revealing the pseudopapilla (H&E stain, X200) and strong nuclear and cytoplasmic beta-catenin positivity through the tumor (anti-beta catenin antibody, immunoperoxidase, X100).

formation of pseudopapilla, and a fibrous capsule is usually seen. Rojas et al.[5] also reported that the overall survival of patients with SPT was significantly better than of patients with other histologic tumors of the pancreas. Their estimated five-year survival rate was 100% for patients with SPT.

Solid pseudopapillary tumor (SPT) is a slow-growing, low-grade malignancy that occurs more frequently in Asians[4]. The pathogenesis of SPT is still unknown. Limited data have shown that patients with SPT have the best prognosis among pediatric patients with pancreatic malignances[4]. Due to the rarity of the pediatric SPT, only a few single institution series and case reports with a limited number of patients have been published in the pediatric literature[2-4,6,9]. Therefore, our understanding of pediatric pancreatic tumors is incomplete[4]. Since pancreatic tumors in children are extremely rare, there is no consensus about the optimal preoperative evaluation. The diagnosis of SPT is not considered until the operation is performed because while radiographic studies are helpful, the findings are not reliable, and specific laboratory tests are not currently available. Nevertheless, the diagnosis of SPT should always be considered when a palpable mass is found in a young female adolescent with normal blood laboratory tests and a large, well-circumscribed mass located in the pancreas in radiological studies.

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