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Solid pseudopapillary neoplasm of the pancreas

Ishna Sharma[a,*], Woo Cheal Cho[b], Shefali Thaker[a], Todd Jensen[a], Fabiola Balarezo[b], Richard Weiss[a], Christine Finck[a]

[a] Connecticut Children's Medical Center, Department of Surgery, 282 Washington Street, Hartford, CT, 06106, USA
[b] Connecticut Children's Medical Center, Department of Pathology, 282 Washington Street, Hartford, CT, 06106, USA

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ABSTRACT

Solid pseudopapillary neoplasm of the pancreas (SPN) is a very rare tumor with a low malignant potential. Although most commonly presenting in females in the second to fourth decade of life, it has been reported in the pediatric population with an incidence of 8–16.6%. SPN was first described by Virginia Kneeland Frantz in 1959 as a papillary cystic tumor of the pancreas in a 2 year old male patient. Herein we report 2 cases of SPN and review the pathophysiology, diagnosis, and management.

Solid pseudopapillary neoplasm (SPN) is a type of pancreatic neoplasm. It is extremely rare and has low malignant potential [1,2]. It was first described by Dr. Virginia Kneeland Frantz in 1959 as a papillary cystic tumor of the pancreas in a 2 year old male patient [3–5]. It makes up 1–3% of exocrine pancreatic tumors, 5% of cystic pancreatic tumors, and 2–3% of all primary pancreatic tumors occurring at all ages [6–17]. Interestingly, metastases can develop many years after the initial complete resection [18,19]. Prognosis is favorable even in the presence of distal metastases [1]. Due to the rarity of this diagnosis in the pediatric population, there is no clear algorithm for diagnosis and management. Herein we report 2 cases of SPN, review the literature, and propose an algorithm for management.

1. Case

Patient A is a 17 year old female who presented with a four day history of right upper quadrant abdominal pain with nausea and vomiting. On initial exam she was febrile with systolic blood pressure in the low nineties. Her abdominal exam was soft, nondistended, with tenderness to palpation at the right upper quadrant, with a negative Murphy's sign, and no rebound or guarding. Her past medical history included multiple sclerosis, right eye retrobulbar optic neuritis, and acute disseminated encephalomyelitis at age 6 years old. An abdominal ultrasound showed intrahepatic biliary ductal dilatation, a normal common bile duct, and a 5.5 × 3.2 centimeter (cm) nonvascular pancreatic head mass. Computerized tomography (CT) of the abdomen with contrast showed a solid low attenuation minimally enhancing mass in the pancreatic head measuring 4 × 3.2 × 4.7 cm, with no pancreatic or ductal dilatation, and no peripancreatic inflammatory changes (Fig. A). Labs are detailed in > Table A.

Pathology from endoscopic ultrasound-guided biopsy revealed a solid pseudopapillary neoplasm of the pancreas. The patient subsequently underwent a Whipple procedure. Surgical pathology of the specimen confirmed solid pseudopapillary tumor, with 1 cm negative resection margins. This mass measured 5.4 × 3.5 × 3.4 cm and was tan-dark brown, rubbery, lobulated, focally hemorrhagic and well-encapsulated. Histologically, the mass showed a proliferation of relatively uniform polygonal cells admixed with numerous capillary-sized blood vessels, forming solid and discohesive pseudopapillary architectural patterns (Fig. B1 and B2). The cytoplasm of the neoplastic cells varied from eosinophilic to clear to foamy, and the nuclei were round to oval with occasional grooves. Some areas showed prominent PAS-positive diastase-resistant intracytoplasmic eosinophilic globules (Fig. B3).

Immunohistochemically, the tumor cells were diffusely and strongly positive for vimentin (Fig. C1), CD10 (Fig. C2), PR (Fig. C3), and beta-catenin (nuclear and cytoplasmic) (Fig. C4), focally and weakly positive for synaptophysin (Fig. D1), and negative for chromogranin (Fig. D2).

These morphologic features and immunophenotypic profiles were consistent with SPN of the pancreas. Postoperatively the patient has done well and her follow-up CT scan 2.5 years postoperatively was negative for recurrence.

Patient B is a 15 year old female who presented with intermittent

* Corresponding author. Connecticut Children's Medical Center, Department of Surgery, 282 Washington Street, Hartford, CT, 06106, USA.
E-mail address: isharma@uchc.edu (I. Sharma).

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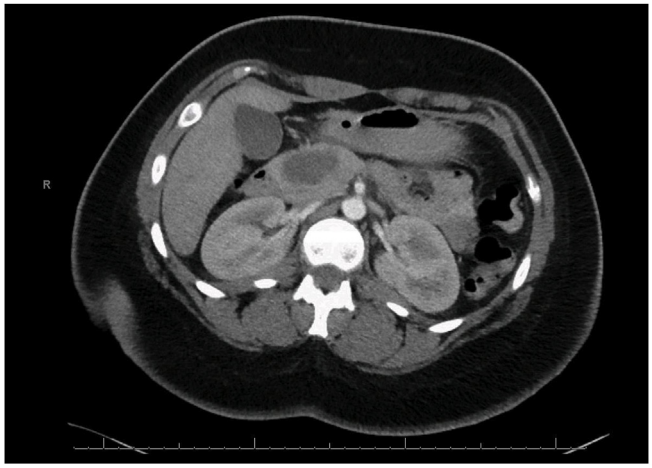

Fig. A. CT abdomen with contrast for patient A.

Table A
Laboratory values for patient A and B.

Labs	Patient A	Patient B
Lipase	Elevated	Normal
Amylase	Normal	Normal
LDH	Elevated	Normal
AST	Elevated	Normal
ALT	Initially normal, Elevated on hospital day 2	Normal
WBC	Normal	Normal
CRP	Elevated	Not measured
5-HIAA	Normal	Not measured
Chromogranin-A	Normal	Not measured
Vasoactive Intestinal Peptide	Normal	Not measured
Glucagon	Normal	Normal
Somatostatin	Normal	Normal
Pancreatic Polypeptide	Normal	Normal
Urine Vanillylmandelic Acid	Normal	Not measured
Homovanillic Acid	Normal	Not measured
Gastrin	Normal	Normal
Total Insulin	Normal	Normal
Ca 19-9	Normal	Not measured
CEA	Normal	Not measured

abdominal pain for 5 months, which had worsened in the week prior to presentation. The patient also complained of fatigue, occasional nausea and vomiting, but without any weight loss. The patient had a history of mononucleosis over six months ago. She underwent an abdominal ultrasound, which showed a pancreatic mass with splenic involvement. CT of the abdomen with contrast showed an 11–12 cm mass in the pancreatic tail, extending into the splenic hilum (Fig. E).

She subsequently underwent a CT-guided biopsy, which demonstrated SPN of the pancreas. Patient B had laboratory values notable for elevated lipase, LDH, AST, ALT, and CRP (Table A). The patient underwent a distal partial pancreatectomy with en-bloc splenectomy. Pathology revealed a mass that was well-circumscribed, tan-red, fleshy and partially hemorrhagic mass, measuring 11.3 × 10.4 × 5.2 cm, in the pancreatic tail (Fig. F).

The morphology and immunoprofile of the tumor was essentially identical to that of Patient A, displaying a pseudopapillary growth pattern with rosette-like growth around fibrovascular cores and a dropout of the loosely cohesive cells in between septa and characteristic immunoreactivities for vimentin, CD10, PR, and beta-catenin, confirming the diagnosis of solid pseudopapillary neoplasm of the pancreas (Figs. G1-G5). Patient B had an uncomplicated postoperative course.

2. Discussion

Malignant pancreatic neoplasms are extremely rare in pediatric patients, although the incidence has been rising since 1987, perhaps due to increasing recognition of the tumor [20–22]. In pediatric patients, the incidence rate is 8–16.6% with an average age at diagnosis of 13 years old [23–25]. It is the most frequent pancreatic tumor in the second decade of life, and mainly occurs in the second to fourth decade in young women [2,26–29]. The incidence of the tumor is 10 times greater in females compared to males and accounts for less than 0.2% of deaths from cancer in pediatric patients [30,31]. Some studies suggest no known ethnic predilection, however others suggest a predilection for Asian and African-American women [20,26,28]. 85% of patients have SPN limited to the pancreas, whereas 10–15% have metastases at time of presentation [1,32,33]. SPN tends to be fairly benign in young females, and more aggressive in older males [28].

The clinical presentation of SPN in children is usually nonspecific [1]. Palpable abdominal mass is the most common presentation, followed by abdominal pain, vomiting, and jaundice [23,34]. In pediatric patients, SPN is most commonly in the head of pancreas, and rarely occurs in extrapancreatic locations [1,26,35,36]. There are no known SPN-specific serum markers [26]. Common serum markers like alpha-fetoprotein, CEA, beta-human chorionic gonadotropin, CA 19-9, and CA 125 are consistently negative in SPN patients [1,26]. There is usually no evidence of pancreatic insufficiency, abnormal liver function tests, cholestasis, elevated pancreatic enzymes, or an endocrine syndrome.

Most SPN cases grossly appear as well-circumscribed lesions surrounded by a pseudocapsule, often with a mix of cystic and solid components [1]. Preoperative workup can include an abdominal ultrasound and CT which show a well-encapsulated, complex mass, with both solid and cystic components [1]. An MRI is very helpful in identifying complex cystic masses in the pancreas [37]. A T1-weighted MRI shows areas of hemorrhagic degeneration as high-signal intensity, and a surrounding hypointense fibrous capsule and these two features are distinguishing of SPN [28,38–40]. A similar dark rim is also seen on T2-weighted images corresponding to the pseudocapsule [38]. MRI is also beneficial for pediatric patients because it does not use radiation [41]. Compared to MRI, CT is limited in showing hemorrhage, cystic degeneration, or capsule presence [42]. Less commonly, positron emission tomography (PET) scan can also be used, which usually shows increased fluorodeoxyglucose uptake in SPN [43]. In the two cases described in this paper, CT scan provided enough diagnostic data to take the next appropriate management step for SPN.

Preoperative diagnosis of SPN continues to be difficult because of the similarity of findings among cystic lesions [1]. The differential diagnoses for a pediatric pancreatic mass include SPN, inflammatory myofibroblastic tumor, rhabdomyosarcoma, pancreatoblastoma, pancreatic endocrine neoplasm, congenital pancreatic cysts/pseudocysts, and chronic pancreatitis [44–46]. Some studies suggest preoperative endoscopic ultrasound-guided fine needle aspiration biopsy, although there is risk of uncertainty in the diagnosis and possibility of tumor spread [1]. The authors of this paper, propose that cytologic confirmation can be helpful to distinguish a lesion from transient forms of pancreatitis, which would be self-limiting. Histologically SPN has cystic areas and solid pseudopapillary arranged cells, however the origin of these tumors is controversial [1,7]. Some suggestions include ductal epithelial, neuroendocrine, multipotent primordial, or even an extrapancreatic genital ridge angle-related cell origin [7,47]. However, SPN