Case Report

Solid Pseudopapillary Tumor of Pancreas in a 15-year-old Female: A Case Report

Xiaobing Jin, MD, PhD; Abdur R Khan, MD*

Department of Pathology, State University of New York at Buffalo, Buffalo, NY

Solid pseudopapillary tumor (SPT) is a very rare tumor accounting for only 1% of all pancreatic exocrine tumors. In this case, patient is a 15-year-old female with history of obesity and oligomenorrhea. She was admitted with a six-day history of severe upper abdominal pain, non-bloody vomiting and occasional diarrhea with no history of fever or sick contact. MRI abdominal examination with contrast showed a cystic 3.4 x 2.2 x 2.0 cm mass in the tail of the pancreas. Patient then underwent the distal pancreatectomy and splenectomy. Grossly, the tumor mass is well-circumscribed and has a tan/yellow cystic cut surface. Microscopically, most of the tumor tissue is necrotic. Sheets of cells demonstrate pseudopapillary arrangement in the preserved area. The nuclei are uniform without apparent mitotic figures and cytoplasm is moderate and eosinophilic. Immunohistochemistry study revealed that tumor cells are positive for CD10, progesterone receptor, synaptophysin and nuclear beta-catenin staining. Diagnosis of this case is challenging because extensive necrosis of the tumor tissue, however, the small areas of residual tumor still retain the pseudopapillary architecture and nested pattern. Individual tumor cells have monotonous low grade character. Immunoprofile also supports the diagnosis of solid pseudopapillary tumor.
[N A J Med Sci. 2019;12(1):021-023. **DOI:** *10.7156/najms.2019.1201021]*

Key Words: *Solid pseudopapillary tumor, pancreas, necrosis*

INTRODUCTION

Solid pseudopapillary tumor (SPT) of pancreas was first reported by Frantz in 1959.[1] It is a rare tumor accounting for only 1% of all pancreatic exocrine tumors and often affects young females in their third and fourth decades of life.[2] The most common location of the tumor is the tail, then the head, body and neck of the pancreas.[3] Patient with SPT usually have abdominal pain, nausea, vomiting and palpable abdominal mass. We report a case of SPT in a 15-year-old female presented with abdominal pain and vomiting, follow up radiologic study showed a cystic mass in the pancreas.

CASE REPORT

Patient is a 15-year-old female with history of obesity and oligomenorrhea. She was admitted with a six-day history of severe upper abdominal pain, non-bloody vomiting and occasional diarrhea with no history of fever or sick contact. Lab results revealed normal amylase and CEA level, and slightly elevated lipase and CA 19-9. Screening abdominal ultrasound showed an intra-pancreatic cystic lesion, follow-up MRI revealed a 3.4 x 2.2 x 2.0 cm cystic mass in the tail of the pancreas with wall enhancement (**Figure 1, A**). Patient then underwent the distal pancreatectomy and splenectomy.

Grossly, the tumor mass is well-circumscribed and has a tan/yellow cystic cut surface with scattered hemorrhage (**Figure 1, B**). Microscopically **(Figure 2**), most of the tumor tissue is necrotic (**A**). Sheets of cells demonstrate pseudopapillary arrangement in the preserved area (**B**). The nuclei of the tumor cells are uniform without apparent mitotic figures and the cytoplasm is moderate (**C**) and focal eosinophilic globules are present (**D**). Immunostaining revealed that tumor cells are positive for CD10 (**E**), and nuclear beta-catenin staining (**F**). Tumor cells are also weakly positive for synaptophysin (**G**) progesterone receptor (**H**).

DISCUSSION

Solid pseudopapillary tumor of pancreas is a rare tumor with low malignant potential. It usually affects young women with a M:F ratio of 1: 7 to 11.[3,4] SPT is quite uncommon in children, in a review of 718 cases reported in English literature, 22% of them were younger than 19 years old.[3] Clinical symptoms of pancreatic SPT are usually non-specific and may be incidentally diagnosed during routine checkup. Abdominal pain, discomfort and palpable non tender mass are most common symptoms, patient may also complain of nausea, vomiting and weight loss et al.[5] In this presented case, patient is a 15-year-old female, who was admitted with severe upper abdominal pain, non-bloody vomiting and occasional diarrhea. A follow-up MRI revealed a 3.4 x 2.2 x 2.0 cm cystic mass in the tail of the pancreas. Lab results revealed normal amylase and CEA level and slightly elevated lipase and CA 19-9. However, these serum markers are not specific for SPT and serum value is not an indicator of malignancy.[4,6]

Received: 08/23/2019; Revised: 09/17/2019; Accepted: 09/25/2019
***Corresponding Author:** Department of Pathology, Oishei Children's Hospital, 818 Ellicott St, Buffalo, NY 14203. Tel: 716-878-1481, Fax: 716-888-3802. (Email: arkhan2@buffalo.edu)

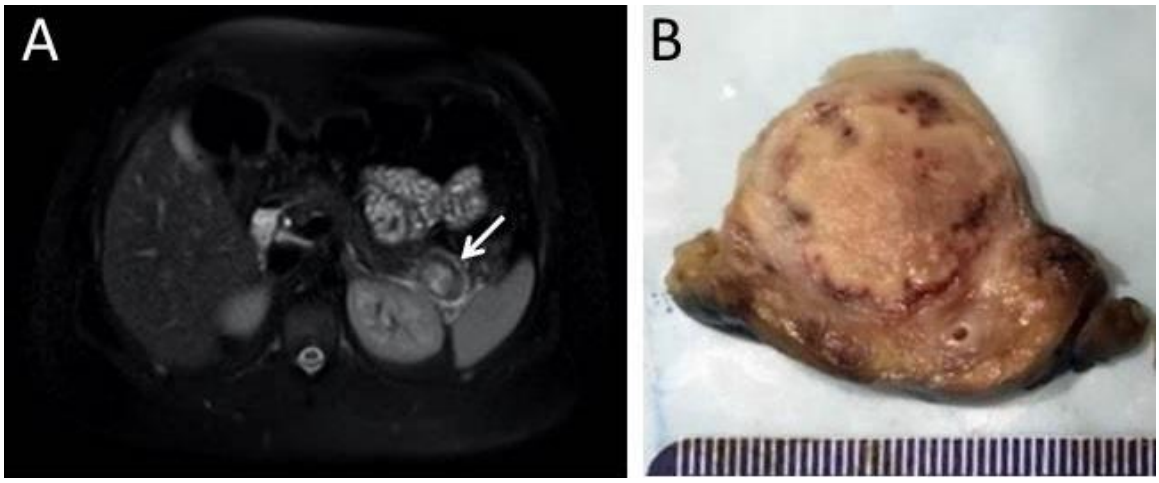

Figure 1. Radiology and gross examination of the tumor. Abdominal MRI demonstrated a cystic mass in the tail of the pancreas with wall enhancement (**A**), Grossly, the tumor mass is well-circumscribed and cut surface appears tan/yellow and cystic cut with scattered hemorrhage (**B**).

SPT are regarded as the low malignant tumor and have favorable long term outcome,[7] it is crucial to differentiate it from other malignant pancreatic tumors. Precise diagnosis of SPT on radiological images is difficult, especially the non-cystic mass may commonly mimic other pancreatic endocrine tumor, serous cystadenoma and mucinous cystadenocarcinoma et al.[8] It is recommended that an EUS-guided FNA is necessary to make a preoperative diagnosis.[9] Histology examination typically shows solid nests of poorly cohesive cells forming a pseudopapillary architecture. Tumor cells usually have a moderate amount of eosinophilic cytoplasm with large intracytoplasmic hyaline globules and perinuclear vacuoles. The tumor cells demonstrate relatively uniform nuclei with finely textured chromatin, inconspicuous nucleoli and characteristic longitudinal grooves. Immunohistochemically, the tumor cells are usually positive for CD 10, CD 56, beta catenin, progesterone receptor, synaptophysin and negative for chromogranin A and estrogen receptor. Careful interpretation of unique histology/cytology features and specific immuoprofile usually allow pathologists to make the diagnosis of this uncommon tumor and differentiate it from neuroendocrine tumor, intraductal papillary carcinoma, acinar cell carcinoma and pancreatoblastoma et al, however, pathologic diagnosis in this case is challenging because extensive necrosis of the tumor tissue. Only small areas of residual tumor still retain the pseudopapillary architecture and nested pattern. Individual tumor cells have monotonous low-grade character and immunoprofile also supports the diagnosis of SPT. No biopsy was performed before surgery in this patient, the extensive necrosis might be due to insufficient blood supply because of rapid growth rate of the tumor.

SPT has excellent prognosis after surgical resection and close follow up is necessary for the early detection of the recurrence and metastasis.[6] Chemotherapy or intra-arterial chemo-embolization is indicated in metastatic cases.[3] In this presented case, no local recurrence or distant metastasis is identified during first year follow up examination after surgical resection.

In conclusion, pancreatic solid pseudopapillary tumor is a rare tumor, it is even more uncommon in children. Pathologic diagnosis may become challenging if tumor demonstrates extensive necrosis.

CONFLICT OF INTEREST

The authors have no conflict of interest to disclose.

REFERENCES

1. VK F. Tumors of the pancreas, DC: Armed Forces Institute of Pathology. Atlas of tumor pathology. 1959; section VII, fascicles 27 and 28.
2. Butte JM, Brennan MF, Gonen M, et al. Solid pseudopapillary tumors of the pancreas. Clinical features, surgical outcomes, and long-term survival in 45 consecutive patients from a single center. J Gastrointest Surg. 2011;15:350-357.
3. Papavramidis T, Papavramidis S. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. J Am Coll Surg. 2005;200:965-972.
4. Yu PF, Hu ZH, Wang XB, et al. Solid pseudopapillary tumor of the pancreas: a review of 553 cases in Chinese literature. World J Gastroenterol. 2010;16:1209-1214.
5. Antoniou EA, Damaskos C, Garmpis N, et al. Solid Pseudopapillary Tumor of the Pancreas: A Single-center Experience and Review of the Literature. In Vivo. 2017;31:501-510.
6. Song H, Dong M, Zhou J, Sheng W, et al. Solid Pseudopapillary Neoplasm of the Pancreas: Clinicopathologic Feature, Risk Factors of Malignancy, and Survival Analysis of 53 Cases from a Single Center. Biomed Res Int. 2017;2017:5465261.
7. El Imad T, Haddad FG, Kesavan M, Deeb L, Andrawes S. Solid Pseudopapillary Tumor of the Pancreas: An Unusual Cause of Abdominal Pain. Cureus. 2017;9:e1252.
8. Uchimi K, Fujita N, Noda Y, et al. Solid cystic tumor of the pancreas: report of six cases and a review of the Japanese literature. J Gastroenterol. 2002;37:972-980.
9. Bardales RH, Centeno B, Mallery JS, et al. Endoscopic ultrasound-guided fine-needle aspiration cytology diagnosis of solid-pseudopapillary tumor of the pancreas: a rare neoplasm of elusive origin but characteristic cytomorphologic features. Am J Clin Pathol. 2004;121:654-662.

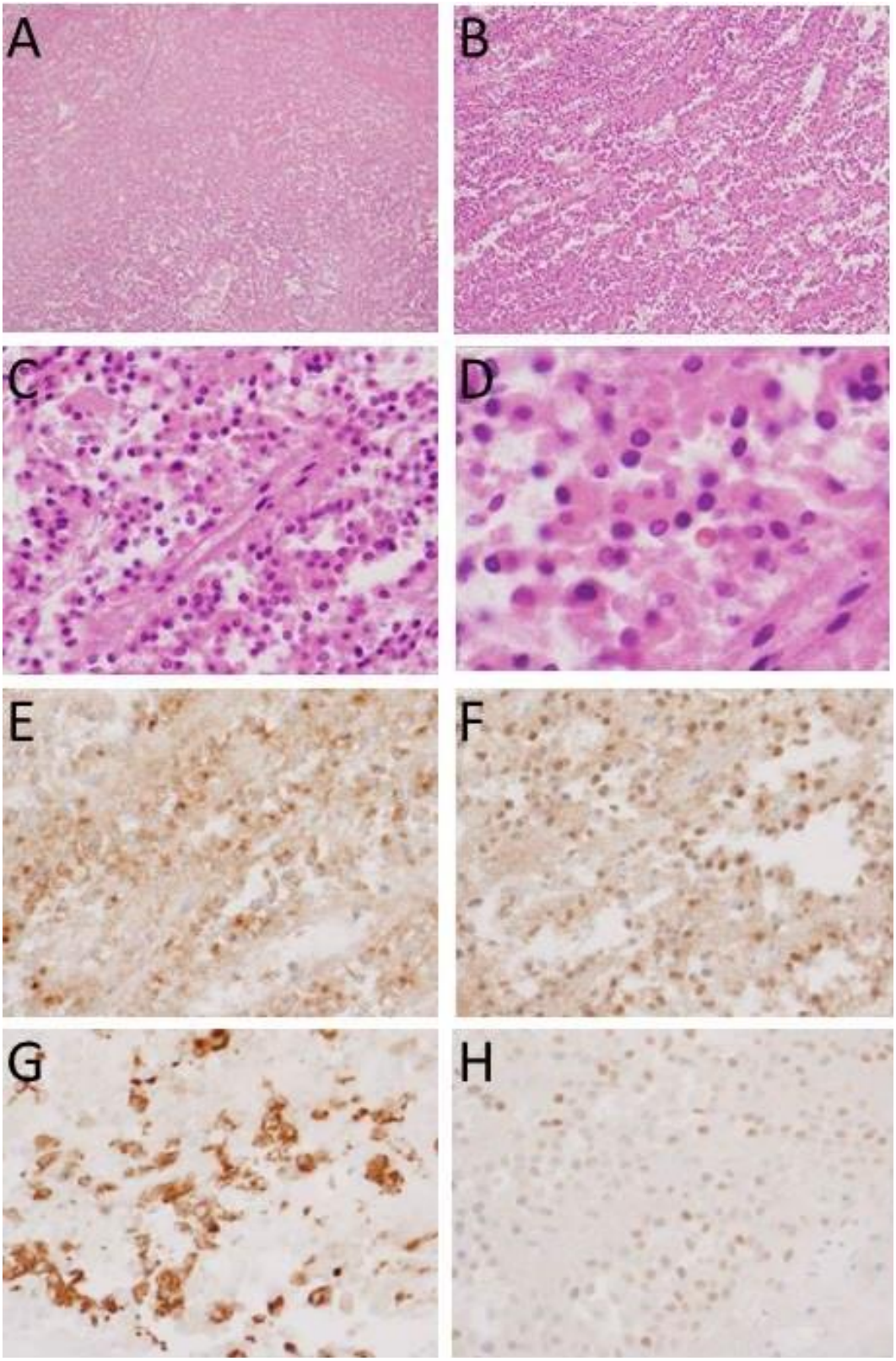

Figure 2. Histology and immunohistochemistry examination. "HE" staining shows most of the tumor tissue is necrotic (**A**, x10), cells demonstrate pseudopapillary arrangement in the small preserved area (**B** x10), the nuclei of the tumor cells are uniform without apparent mitotic figures, cytoplasm is moderate (**C**, x40) and focal eosinophili globules are present (**D**, x100). Immunostaining revealed that tumor cells are positive for CD10 (**E** x40), nuclear beta-catenin staining (**F**, x40). And weakly positive for synaptophysin (**G**, x40) progesterone receptor (**H**, x40).