

Case Report

Solid pseudopapillary neoplasm of pancreas: a rare presentation

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ABSTRACT

Pancreatic neoplasms are rare in children and have a different histo-logic spectrum and prognosis than those in adults. Pancreatoblastoma is the most common pancreatic neoplasm in young children. Solid pseudopapillary neoplasm occurs in adolescent girls. It is heterogeneous in internal architecture, with a mixture of solid and cystic hemorrhagic and necrotic elements. All pancreatic neoplasms in children are capable of producing metastases, usually to the liver and lymph nodes; however, on the whole, these tumors have a better clinical outcome than most pancreatic tumors in adults. We present a case of solid pseudopapillary neoplasm with a liver metastasis in a 13 year old male patient.

Keywords: Pancreatic neoplasm, Solid pseudopapillary neoplasm, Adolescent girls

INTRODUCTION

Pancreatic tumors are quite rare in children; causing less than 0.2% of malignant pediatric deaths.¹ The scarcity of cases limits our ability to study these tumors.

In addition, confusing and evolving nomenclature makes it difficult to compare current cases to more remote cases in the literature.² Pancreatic neoplasms are divided into epithelial and nonepithelial types.³

*Pancreatic tumors that may occur in children*³

Epithelial tumors

- Ductal cell origin
- Ductal adenocarcinoma (exceedingly rare)
- Acinar cell origin
- Pancreatoblastoma
- Acinar cell carcinoma

Uncertain origin

- Solid-pseudopapillary tumor

Endocrine cell origin

Non-epithelial tumors (exceedingly rare)

- Lymphoma, particularly Burkitt
- Sarcomas, particularly rhabdomyosarcoma
- Dermoid cyst
- Lymphangioma
- Hemangioendothelioma

First described by Franz in 1959, SPT of the pancreas is a rare, low-grade malignant tumor of unknown etiology accounting for 0.2-2.7% of all primary pancreatic tumors.⁴⁻⁶ Clinically, solid pseudopapillary neoplasms are significantly more common in women (male to female ratio, 1: 9).⁷ It occurs predominantly in adolescent girls and young women (mean 35 years; range 8-67 years). It is rare in men (mean, 35 years; range 25-72 years).⁸ Occurrence in first decade, however, is rare.⁷

CASE REPORT

A 13 year old male child presented in surgical OPD with dull aching pain and swelling over upper abdomen since 3 months. His biochemical findings were normal.

Ultrasonographically, pancreas was normal in size and showed a well-defined hypoechoic mass of size 2.96x4.08 cm involving the tail of pancreas, pancreatic duct not dilated. On MRI, a well-defined heterogeneously enhancing mass lesion of size 3.8x2.7x 3.5 cm in tail of pancreas with mass effect extended to splenic vein inferiorly, the lesion is hypointense on T1 and hyperintense on T2, with heterogenous post contrast enhancement. Likely, suggestive of neoplastic etiology.

Liver showed two small well defined rounded lesions seen in right lobe of liver in segment VII which is hypointense on T1 and hyperintense on T2, with heterogenous post contrast enhancement, likely hepatic metastasis.

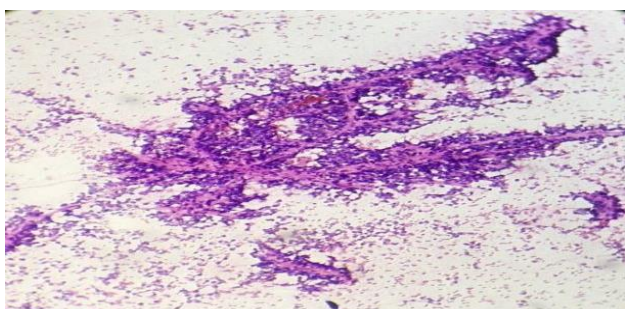


Figure 1: Cytology smear microphotograph showing branching papillary fronds with myxoid stroma (10x, MGG).



Figure 2: Cut section of distal pancreatectomy specimen showing solid-grey white tumor with central necrosis.

Ultrasound guided fine needle aspiration cytology was performed from lesions identified in liver. Highly cellular smears showed tumor cells with papillary architecture having monomorphic round to oval nuclei with moderate amount of pale cytoplasm. Diagnosis of metastatic pseudopapillary neoplasm, probably of pancreatic origin was given (Figure 1). The patient underwent distal pancreatectomy with splenectomy. Gross examination showed a well-encapsulated tumor mass of size 7x4x2 cm, cut section revealed solid grey-white mass with central area of necrosis (Figure 2).

Microscopically tumor cells were loosely arranged in pseudopapillary pattern with hyalinised stroma. The cells

were small to medium sized having ovoid nuclei and eosinophilic cytoplasm. Mitotic figures were rare. Immunohistochemistry showed strong positivity for vimentin and S-100 along with CK-20 negative (Figure 3-5).

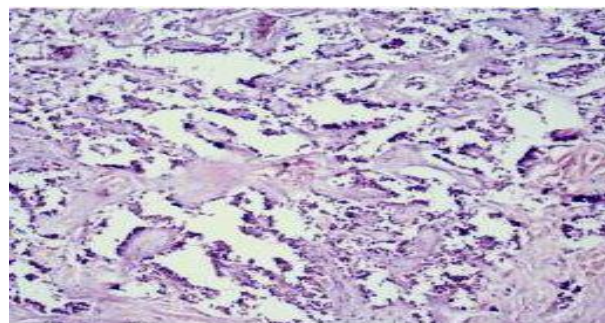


Figure 3: Microphotograph of histopathology section showing pseudopapillary pattern of tumor cells having hyalinised stroma; (10x, H&E).

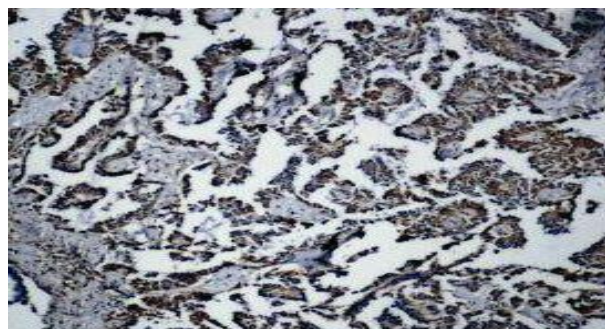


Figure 4: Tumor cells showing strong immunoreactivity for vimentin in cytoplasm.

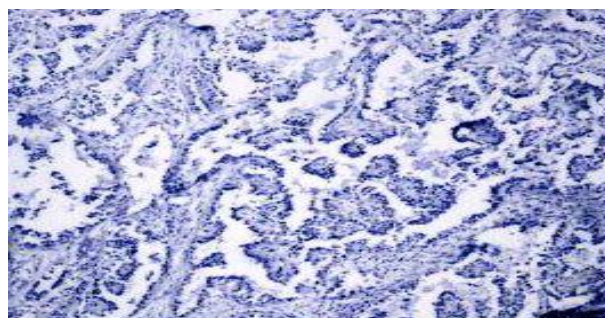


Figure 5: Tumor cells negative for CK 20.

DISCUSSION

Solid pseudopapillary neoplasm (SPN) is a tumor of uncertain cellular differentiation.⁷ Kosmahl et al hypothesized that SPN may actually be derived from genital ridge/ovarian anlage-related cells which were attached to the pancreatic tissue during early embryogenesis.⁹ However, the fact that SPN can occur in males without sex hormone defects as well as the inability to identify and ovarian cell which exactly corresponds to the immunohistochemical pattern of SPN

cells coupled with the apparent lack of ovarian tumors having a strong similarity to SPN has led to serious doubts about this concept.¹⁰ In support of this view point, we also present a case of 13 year old male child.

Symptoms are non-specific, and some cases are detected only incidentally after trauma or during gynaecologic or obstetric examinations. SPN often reach large sizes before clinical detection, and the average case measures more than 10 cm in size.⁷ Similarly, our case also presented with pain and swelling in abdomen since 3 months and previously no other complaint was present.

In accordance with our case, most common localization of SPN is the tail of the pancreas, followed by the head and the body. Unusual presentations include multicentric tumors in the pancreas and extrapancreatic sites, such as the mesocolon, retroperitoneum, omentum, liver and duodenum, possibly representing synchronous tumor spread.^{11,12}

The differential diagnosis of SPN is wide and includes lesions such as acinar cell carcinoma, mixed acinar cell neoplasm, pancreatoblastoma, and pancreatic endocrine neoplasm. All share a relatively solid, cellular appearance, which is distinct from that of conventional ductal adenocarcinoma and most cystic neoplasms. Specific histologic features are helpful such as squamoid corpuscles in pancreatoblastoma. However, immunohistochemistry is widely considered the most helpful diagnostic technique in most instances.⁷

Malignant SPN, designated as a solid-pseudopapillary carcinoma, occurs in 15% of adult patients. Criteria which distinguish potentially malignant tumors and which are classified as 'SP carcinoma' are: 1) angioinvasion; 2) perineural invasion; and 3) deep invasion of the surrounding pancreatic parenchyma.

A recent study showed that some histological features, such as extensive necrosis, nuclear atypia, high mitotic rate, immunohistochemistry findings of expression of Ki-67 and sarcomatoid areas may be associated with aggressive behaviour.¹³ In the present case criteria for malignancy were identified and diagnosis of solid pseudopapillary carcinoma was done. Hence, as per our knowledge this is one of the rare presentation of this tumor considering the both age and gender of the patient.

Given their low malignant potential and the excellent overall prognosis, surgical resection has been the standard of care in the management of SPN.¹² Tumor enucleation and incomplete excision should be avoided due to the risk of tumor dissemination, development of a pancreatic fistula.¹⁴

CONCLUSION

Above case emphasise on certain points regarding the evaluation of pancreatic tumors:

- Early and careful diagnosis of pancreatic tumors is essential in children because essentially all pancreatic tumors have a better clinical outcome than most pancreatic tumors in adults.
- All the differential diagnosis of pancreatic masses in children should be carefully evaluated as it may promote correct preoperative diagnostic and prognostic work-up and appropriate treatment.

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REFERENCES

1. Tsukimoto I, Watanabe K, Lin JB, Nakajima T. Pancreatic carcinoma in children in Japan. *Cancer.* 1973;31(5):1203-7.
2. Chung EM, Travis MD, Conran RM. Pancreatic tumors in children: Radiologic-pathologic correlation. *Radiographics.* 2006;26(4):1211-38.
3. Jaffe R. The pancreas. In: Stocker J, Dehner L, eds. *Pediatric pathology.* 2nd ed. Philadelphia, Pa: Lippincott Williams & Wilkins. 2002:797-834.
4. Frantz VK. Tumors of the pancreas. In: *Atlas of Tumor Pathology.* Washington DC: Armed Forces institute of Pathology. 1959:32-3.
5. Chen X, Zhou GW, Zhou HJ, Peng CH, Li HW. Diagnosis and treatment of solid-pseudopapillary tumors of the pancreas. *Hepatobiliary Pancreat Dis Int.* 2005;4(3):456-9.
6. Pettinato G, Manivel JC, Ravetto C, Terracciano LM, Gould EW, di Tuoro A, et al. Papillary cystic tumor of the pancreas: a clinicopathologic study of 20 cases with cytologic, immunohistochemical, ultrastructural, and flow cytometric observations, and a review of literature. *Am J Clin Pathol.* 1992;98(5):478-88.
7. Odze RD, Goldblum JR. *Surgical Pathology of the GI tract, liver, biliary tract, and pancreas.* 2nd ed. Elsevier. Saunders., 2009:944-6.
8. Hamilton SR, Aaltonen LA. *World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of the Digestive System.* Lyon: IARC Press, 2000.
9. Kosmahl M, Seada LS, Jänig U, Harms D, Klöppel G. Solid-pseudopapillary tumor of the pancreas: its origin revisited. *Virchows Arch.* 2000;436:473-80.
10. Von Herbay A, Sieg B, Otto HF. Solid-cystic tumour of the pancreas. An endocrine neoplasm? *Virchows Arch A Pathol Anat Histopathol.* 1990;416:535-8.
11. Papavramidis T, Papavramidis S. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in the English literature. *J Am Coll Surg.* 2005;2:965-72.
12. Klöppel G, Maurer R, Hofmann E, Lüthold K, Oscarson J, Forsby N, et al. Solid-cystic (papillary-cystic) tumors within and outside the pancreas in

- men: report of two patients. *Virchows Arch.* 1991;418:179-83.
13. Tang LH, Aydin H, Brennan MF, Klimstra DS. Clinically aggressive solid pseudopapillary tumor of the pancreas. A report of two cases with components of undifferentiated carcinoma and a comparative clinicopathologic analysis of 34 conventional cases. *Am J Surg Pathol.* 2005;29:512-9.
 14. Reddy S, Cameron JL, Scudiere J, Hruban RH, Fishman EK, Ahuja N, et al. Surgical management of solid-pseudopapillary neoplasms of the pancreas: a large single-institutional series. *J Amer Coll Surg.* 2009;208:950-9.

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