

Case Report

Pseudo Papillary Tumor of Pancreas in a Child

Farhan Abbas Baloch

Abstract Solid pseudopapillary tumor of the pancreas is a rare pancreatic tumor. Mostly these tumors are found in women who are in their third decade of life. The tumors have a low malignant potential. A case of 9-year-old female was observed who presented with dull abdominal pain, abdominal heaviness, mass abdomen for 2 and a half years duration. The mass was excised. The histopathological examination revealed Pseudo Papillary Tumor of Pancreas.

Keywords: Solid pseudopapillary tumor of pancreas, childhood tumor of pancreas

Introduction

Solid pseudopapillary tumor of the pancreas was first described by Frantz in 1959.¹ Solid pseudopapillary tumors are rare tumors and account for 1-2% of all primary tumors of pancreas. There are many synonyms for solid pseudopapillary tumors which include papillary cystic neoplasm, solid-cystic papillary tumor or Gruber-Frantz tumor. Mostly these tumors are found in young women in the second or third decade.² In children these tumors are rarely seen.³ Mostly the patients present with a palpable abdominal mass and a dull abdominal pain. A low malignant potential is suspected in these tumors and their prognosis is extremely good.^{4,5} WHO in 1996, reclassified this tumor and now solid pseudopapillary tumor of pancreas is a universally recognized entity.

Case history

A 9 -year-old female patient presented with history of abdominal pain and abdominal heaviness for 2 and half years duration. The pain was dull and not associated with anorexia, nausea, vomiting, and weight loss. Her past medical or surgical history & family history was not significant. Her baseline investigations were normal and hepatitis profile was negative for both hepatitis B and C. Her serum VMA and amylase were within normal limits. The ultrasound abdomen showed solid mass in left hypochondrium. The CT scan showed a well-defined mass in the pancreatic tail. So

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Article Received : August 16,2010

Acceptance Date: September 07,2010



Figure 1: Solid Pseudopapillary Tumor of Pancreas. The tumor is solid and well circumscribed

the patient had undergone exploratory laprotomy, and a solid mass of 10 x 7 x 4 cm, present in the tail of pancreas at splenic hilum was found. Bistal pancreatic my was performed Lymph nodes were seen to be enlarged

Gross Examination: The tumor was well circumscribed predominantly solid mass. Cut surfaces were greyish brown, firm and solid with few areas of hemorrhage. Frank necrosis was not identified (Fig.1)

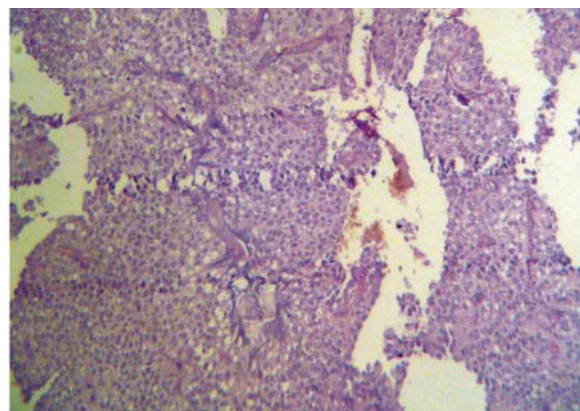


Figure 2: Pseudopapillary formations due to focal necrosis (H&E X 100)

Microscopic Examination: The tumor was very cellular sparing the adjacent pancreas. Tumor cells were arranged in solid sheets and pseudopapillae. Individual tumor cells were uniform, small to medium sized, polygonal with acidophilic cytoplasm and bland vesicular, ovoid nuclei. Few nuclei showed nuclear grooves and indistinct nucleoli. Mitotic figures were few. No vascular invasion was seen. The surrounding stroma was fibrocollagenous. (Fig. 2, 3, 4). The diagnosis of Solid pseudopapillary tumor of the pancreas was made.

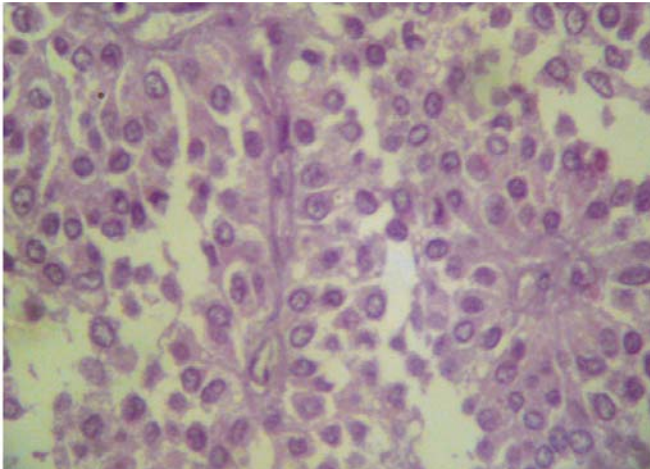


Figure 3: Pseudopapillae of Solid Pseudopapillary Tumor of Pancreas (H&E, 400).

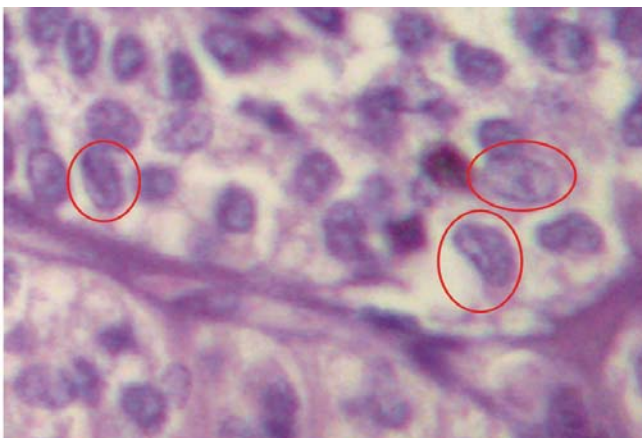


Figure 4: Nuclear grooves in Solid Pseudopapillary tumour of Pancreas (1000x) .

Discussion

Approximately 718 Solid pseudopapillary tumors of the pancreas cases have been reported in the literature.⁶ which were mostly seen in young women. These are slow-growing tumors with indolent

course. These tumors are commonly located in the tail of pancreas.

Most of the solid pseudopapillary tumors of the pancreas are large with solid and cystic areas. The present case however showed only solid pattern and no cystic spaces were identified. Initially tumors begin as solid masses in which there are many poorly supported tiny vessels and then the cells away from the small vessels undergo degenerative changes due to insufficient blood supply; however the cells adjacent to the vessels remain intact. This results in pseudopapillary and cystic spaces.⁷

Histologically, these tumors are composed of uniform epithelial cells arranged in solid sheets, nests and well-formed acinar structures. Squamoid corpuscles can also be present; however these structures were not seen in the present case. Controversy exists about the cell of origin. Some consider it to be of uncommitted cells origin, while others consider it to be arising from intercalated duct cells or centroacinar cells.⁸

Local recurrences and metastases are unusual. Liver being the most common site for metastases. Rare lymph node and peritoneal metastasis have been reported.⁹ Despite the presence of disseminated disease, clinically the tumor has a protracted course and overall 5-year survival rate is 97%.¹⁰ The tumor has a low malignant potential, however the features of its aggressive behavior, when present may include venous invasion, diffuse growth pattern, extensive tumor necrosis, significant nuclear atypia and high mitotic count.¹¹ Gross and microscopic appearances are characteristic to make the diagnosis of this tumor easy. In few difficult cases immunohistochemistry can be done to exclude other tumors of the pancreas.¹²

Immunohistochemically, the solid pseudopapillary tumors of the pancreas shows positive reactivity for keratin, desmoplakin, trypsin, chymotrypsin, amylase and vimentin. Focal positivity has been found for neuron-specific enolase and various islet cell hormones such as insulin and glucagons. In view of this immunohistochemical pattern it is suggested that solid pseudopapillary tumors of the pancreas arise from primitive pancreatic epithelial cells with predominance of exocrine features but having capacity for dual (endocrine and exocrine) differentiation. The presence of progesterone receptors and its well-known predilection for females suggest that it is a hormone-dependent tumor.¹³ Solid pseudopapillary tumors of the pancreas are surgically treated. The prognosis is good after resections because of its indolent growth.

Conclusion

Solid pseudopapillary tumors of the pancreas are rare pancreatic tumors mostly seen in young women. Accurate diagnose of this tumor is necessary, as surgical resection is curative. Our patient is on close follow up with no untoward complication reported so far.

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