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Solid Pseudopapillary Neoplasm: A Case Report of a Rare Pancreatic Tumor

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Abstract
Solid Pseudopapillary Neoplasm (SPN) of pancreas is a rare neoplasm with a reported incidence of 0.1-2.7% of all pancreatic tumors. SPN is mostly seen in young females. It has a low malignant potential and has an excellent prognosis. We report a case of 41 years old female who presented with an abdominal lump. A mass measuring9.4×6.2 cm was noted on CECT. The diagnosis of Solid Pseudopapillary Neoplasm of Pancreas was confirmed on histopathological examination. Being a rare tumor of pancreas, SPN is often missed on radiological examination. A solid/cystic mass on gross examination and microscopic presence of solid and papillary arrangement of cells is highly suggestive of SPN. Solid areas comprise of sheets and cords of round or ovoid, bland cells arranged on the perimeter of delicate fibrovascular septa. Malignant SPN, designated as a solid pseudopapillary carcinoma, is seen in about 15% of adult patients. Surgical resection of the tumor is the mainstay of therapy of SPN.Strict post-operative follow-up is necessary to keep a check on local and distant metastasis.
Keywords: *Solid Pseudopapillary Neoplasm, SPN, Pancreas.*

Introduction

Solid pseudopapillary neoplasm (SPN) of pancreas is a rare primary neoplasm of pancreas with a reported incidence of 0.17% to 2.7% of all pancreatic tumors.[1] It is a peculiar tumor of low malignant potential and has an inexplicable predilection for adolescent girls and young females. First described by Frantz in 1959, it was given numerous names, until World Health Organization named it "solid pseudopapillary neoplasm" of pancreas in 1996. The male to female ratio is 1:10 and the mean age at presentation is 22 years.[2] SPN has a relatively benign course, however in about 15% of cases it presents as malignancy with metastasis and invasion of adjacent organs.[2] Despite its locally aggressive behaviour, the tumor has a low-grade malignant potential and tends to have a favourable

prognosis, even in the presence of metastatic disease.

Case Report

A 41 years old female presented to the surgical department with complaints of abdominal pain, weight loss and a palpable abdominal mass. Ultrasound abdomen shows a complex cystic mass measuring (8.4×6.2) cm in peripancreatic region containing echogenic debris and thin septae. Contrast Enhanced Computed Tomography of abdomen revealed a well-defined round to oval thin-walled hypodense lesion measuring (9.4×7.3)cm in the neck and body of the pancreas. Wall thickness was 3mm and multiple foci of calcification were noted in the wall. Thin septae up to 1mm were seen. Impression of mucinous tumor of pancreas was given on radiology. The patient underwent distal pancreatectomy with splenectomy and the specimen was sent for histopathological examination.

Gross examination showed a pancreatic mass measuring (11×11×9) cm present at one end and a spleen measuring (14×7×3)cm at the other end, connected tofibro fatty soft tissue measuring (15×5×1.5)cm. Cut section of pancreatic mass was cystic and drained out 10 ml of straw coloured fluid. The cyst was unilocular and was filled with grey brown to yellow friable tissue measuring (8×6×5) cm. Cyst wall was variably thickened measuring 4mm to 8mm, was firm and gritty to cut. Spleen was grossly unremarkable. Microscopic examination revealed pseudo papillae, solid nests and trabecular arrangement of small to medium sized polygonal cells having occasional bland nuclei with indentation, perinuclear vacuoles and moderate amount of eosinophilic to clear cytoplasm. Marked hyalinization of stroma and focal eosinophilic globules were also seen. Areas of necrosis, hemorrhage, cholesterol clefts, fibrosis, scattered chronic inflammatory cells and dilated congested blood vessels along with compressed normal pancreatic parenchyma seen. Occasional mitosis was seen and the resection margins were free from tumor. Sections from spleen were unremarkable microscopically. Histomorphological diagnosis of Solid Pseudopapillary Tumor of Pancreas was thus assigned. Postoperatively the patient is on regular follow-up and is doing well.

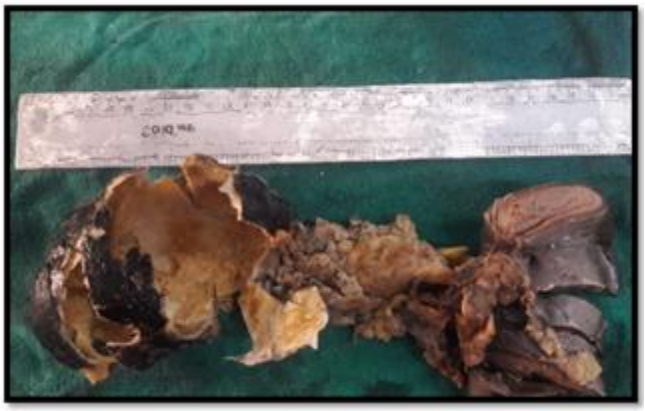

Fig.1: Cut section showing cystic pancreatic mass with attached spleen

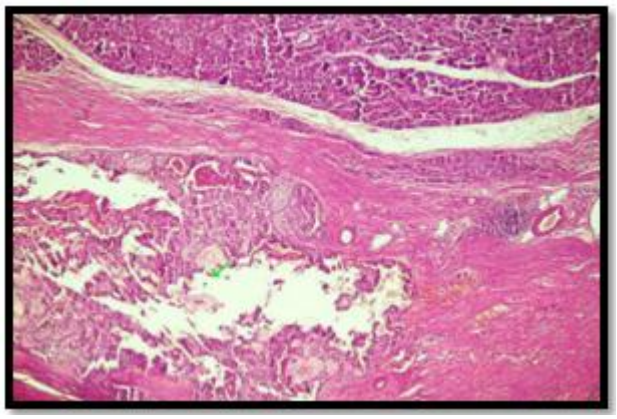

Fig.2.showing tumor (green arrow) and normal pancreatic tissue (top).

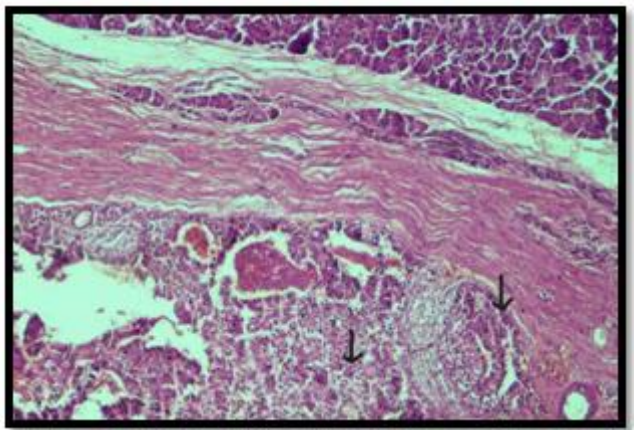

Fig 3. showing pseudopapillary pattern of tumor cells (arrows)

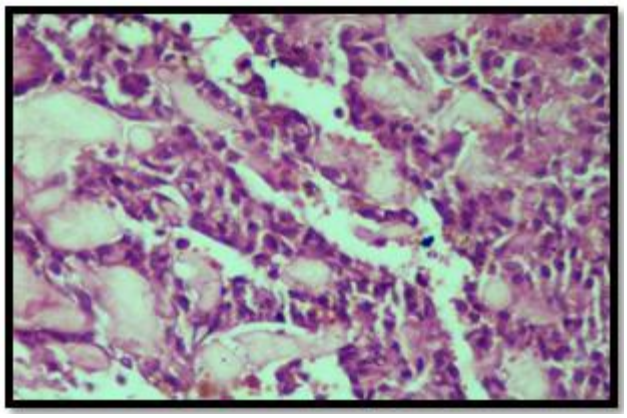

Fig.4 showing uniform polygonal tumor cells havingbland nuclei and moderate amount of eosinophilic to clear cytoplasm.

Discussion

SPN is a rare neoplasm with low malignant potential and has tendency to affect young females usually in the second and third decade of life due to involvement of sex hormones.[3] Although SPN has low malignant potential, up to 15% of SPN patients develop metastasis. The most commonly involved sites of metastasis are the liver, regional lymph nodes, mesentery, omentum, and peritoneum[1]. A study by Yu PF et al[3] showed that local invasion of the duodenum, stomach, spleen or major blood vessels may also occur. The clinical presentation of SPN is nonspecific. Often, the patients are asymptomatic for a long period of time, until the tumor reaches a substantial size and compresses the adjacent organs. Sometimes the patient may have mild abdominal pain, nausea, poor appetite, loss of weight, or palpable abdominalmass.[4,5] Predominance of SPN in females has been reported at a ratio of 10:1 and the mean age of presentation is about 22 years. SPN of the pancreas has peculiar pathological characteristics. On gross examination, the mass is usually large, well-defined and encapsulated. Areas of tissue necrosis, hemorrhage and cystic change are also seen.[6] In the present case, grossly the tumor was a unilocular cyst, containing straw coloured fluid. The cut surface was filled with grey brown to yellow friable tissue. Cyst wall was fibrotic and gritty to cut. Microscopy shows two types of cellular arrangements: solid and papillary.[6] Some areas comprise of sheets and cords of round or ovoid, bland cells arranged on the perimeter of delicate fibrovascular septa.[6] Eosinophilic globules composed of α1-antitrypsin might also be seen. Mitotic activity is usually low and true necrosis is uncommon; however, cystic degeneration is frequent. The pathological diagnosis is dependent on the presence of solid areas alternating with pseudopapillary pattern and pseudo-rosettes. In the present case, pseudopapillary pattern, trabecular pattern and solid nests of small to medium sized polygonal cells were seen. SPNs are typically positive for B-catenin, Vimentin, NSE, alpha-1-antitrypsin and alpha-1-antichymotrypsin, estrogen and progesterone.[6] In the present case, the tumor cells were reactive for B-catenin and Vimentin.

SPN is very difficult to diagnose preoperatively as the radiological appearance of SPN is unique. Computed tomography can show a heterogeneous mass, with a combination of solid and cystic components in various proportions. However, it is more difficult to diagnose SPN radiologically when the tumor is smaller than 3-cm in diameter. For SPNs smaller than 3-cm, the cystic component may not be obvious. In the present case initial radiological impression was mucinous tumor of pancreas. Histopathology is thus of great significance in diagnosing SPN.

The most important differential diagnosis is with neuroendocrine tumors of pancreas and mucinous cystadenocarcinomas both of which have relatively less favourable prognosis when compared to SPN. Surgical resection is the mainstay of therapy for SPN. Distal pancreatectomy with or without splenectomy, pylorus preserving pancreatoduodenectomy, Whipple's procedure are some of the modalities that can be pursued. Enucleation can be done for smaller SPN's. Lymph node excision is extremely rare. Metastasis to liver and peritoneum is observed in about 15% of cases. Overall, 5-year survival rate is about 95%.[7] Malignant SPN, designated as a solid pseudopapillary carcinoma is seen in about 15% of adult patients.[7]

Angioinvasion, perineural invasion and deep invasion of the surrounding pancreatic parenchyma are the various criteria that distinguish potentially malignant tumors, which are classified as Solid papillary carcinoma. Other histological features such as extensive necrosis, nuclear atypia, high mitotic rate, Ki-67 proliferative index, and sarcomatoid areas may be associated with aggressive behavior.[7]

Conclusion

SPN is a rare pancreatic tumor with low malignant potential and an unknown origin, incidence of which is increasing by the day. Hence a pathologist should always suspect SPN in a young female who presents with a solid cystic mass. Even in the presence of metastasis, prognosis is favourable. Surgical resection with clear margins is the mainstay of treatment although a strict follow-up is necessary to check on local or distant metastasis.

Sources of Support- Nil
Conflicts of Interest- Nil

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