

Case Reports

Solid Cystic Papillary Epithelial Neoplasm of Pancreas: a series of three cases with review of literature

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Solid-cystic- papillary epithelial neoplasms of the pancreas (also referred to as papillary serous epithelial neoplasm, solid- pseudopapillary tumor) are uncommon benign or low grade malignant tumors occurring pre-dominantly in young women.¹ While occurring often in adolescent girls, they are rare in childhood, in older women, and in men.² These tumors have an excellent prognosis and after complete surgical removal, more than 95% patients are cured. They are evenly distributed in the pancreas^{3,4}, present as large solitary masses (size range 3 to 18cms, average 8 to 10 cms) and are usually well demarcated from the remaining pancreas so that they can be easily removed surgically.¹ However; occasionally they invade the surrounding pancreatic parenchyma.^{3,5} These tumors can recur locally⁴ even as late as 3 to 10 years after tumor resection, so long term follow up is important.⁶ Only a few metastasize.^{3,7} Three patients with solid-cystic-papillary epithelial neoplasms of the pancreas are reported.

Case Report

Case 1

A 16 year old girl from Peshawar presented with a history of abdominal mass. No other clinical details were available. Laparotomy had been performed and biopsy diagnosis from Peshawar was Wilm's tumor. The case was sent to us for second opinion. We received five paraffin blocks and partly five Hand E stained slides. All the sections showed a neoplastic lesion with partly solid and partly pseudo papillary pattern, the tumor cells were monomorphic with round to ovoid nuclei and inconspicuous nucleoli. Mitoses were rare (Figure 1). In areas, the tumor cells were radially arranged around minute fibro vascular stalks. In one of the sections, benign pancreatic tissue was seen with no infiltration by the tumor. Immunohistochemically, the tumor cells showed positivity for vimentin and NSE (neuron specific enolase), while chromogranin A, synaptophysin, cytokeratin MNF and Cytokeratin CAM 5.2 were negative. Based on histopathological and immunohistochemical features, a diagnosis of solid-cystic papillary epithelial neoplasm was favored and clinical and radiological correlation was advised.

Case 2

A 27 year old woman from Multan presented with a history of lower abdominal pain and swelling. We received

Figure 1. Photomicrograph showing a neoplastic lesion with partly solid and partly pseudopapillary pattern (Mag: 10X).

Figure 2. Photomicrograph showing monomorphic tumor cells, polygonal with abundant pink cytoplasm and central ovoid nuclei (Mag: 40X).

the specimen coded as "midline retroperitoneal mass". Grossly, it was nodular measuring 6.5x5x4 cms. Cut surface was light brown with hemorrhagic areas. The entire specimen grossly appeared to be the lesion and no normal tissue was identified. Multiple sections were submitted. Sections showed a neoplastic lesion composed of sheets of tumor cells. In areas, pseudo papillary fronds were identified. The tumor cells were monomorphic, polygonal in shape with abundant pink cytoplasm and central ovoid nuclei (Figure 2). There was no nuclear atypia and no significant mitotic activity. Necrosis was absent. However, foci of calcification, cholesterol clefts and collections of foamy macrophages were seen. In one section, a thin rim of benign pancreatic parenchyma was identified. Immunohistochemically,

benign pancreatic parenchyma was identified. Immunohistochemically, the tumor cells showed positivity for vimentin and NSE and focal positivity for cytokeratins MNF and CAM 5.2. The tumor cells were negative for synaptophysin and chromogranin A. Based on histological and immunohistochemical features, a diagnosis of solid cystic papillary epithelial neoplasm of the pancreas was favored and clinical correlation was advised.

Case 3

A 14 year old girl from Peshawar presented with a history of upper abdominal pain and mass in upper abdomen. Surgery was performed in Peshawar and specimen was sent to us coded as "tumor Mass." On gross examination, the specimen consisted of a nodular piece of tissue, which was encapsulated and measured 12x8x6 cms. On one side, normal appearing pancreatic tissue was attached. Outer surface was smooth. On sectioning, cut surface was solid with focal cystic and hemorrhagic areas. Multiple sections were submitted. Microscopic examination showed a tumor composed of uniform polygonal cells arranged around delicate and hyalinised fibro vascular stalks. These cells had uniform nuclei and abundant pink cytoplasm. In areas, the tumor cells were forming solid sheets and had clear to pink cytoplasm, mitoses were rare. Focally, the lesion was seen to infiltrate the normal pancreatic tissue. Focal vascular and neural invasion was also seen (Figure 3). Immuno histochemistry showed positivity for Vimentin and NSE, and focal positivity for cytokeratin MNF. There was negativity for carcinoembryonic Antigen (CEA), synaptophysin and chromogranin A. Based on the above features, a diagnosis of solid-cystic papillary epithelial neoplasm with infiltration of adjacent pancreatic tissue, vascular and neural invasion was given and close follow up was advised.

Discussion

Grossly, solid-cystic papillary epithelial neoplasms of pancreas are encapsulated, have lobulated light brown solid cut surface with zones of hemorrhage, necrosis and cystic spaces filled with necrotic debris. Microscopically there is a solid monomorphous pattern with variable sclerosis and merges with a pseudo papillary pattern. Tumor cells are uniform, polygonal with indented nuclei, inconspicuous nucleoli, and a clear to eosinophilic cytoplasm, which is glycogen and mucin negative, mitoses are uncommon.^{1,2} Criteria for malignancy are not well established. Perineural or vascular invasion with or without deep invasion of surrounding tissue may indicate malignant behavior.¹ Nuclear atypia, mitotic rate and prominent necrobiotic cell nests (cells with pyknotic nuclei and eosinophilic cytoplasm) have also been described as helpful histologic parameters of malignancy. Immuno histochemically, the tumor cells show diffuse positivity for NSE Vimentin and focal positivity for alpha 1 antitrypsin and alpha 1 antichymotrypsin.^{8,9} Some tumors stain for cytokeratins and S 100 protein while most are negative for synaptophysin and chromogranin.^{3,10}

Cytologically, papillary structures with delicate vascular cores and covered by several layers of bland tumor cells are seen. These features are distinct from those of other cystic pancreatic tumors and preoperative cytologic diagnosis may lead to prompt appropriate surgical treatment.^{11,12} Characteristic ultrasound findings are well-encapsulated solid and cystic masses and in the appropriate clinical setting, a radiologic diagnosis can permit curative surgery.¹³

In the three cases, surgical specimen was available in two cases, and these two tumors measured 6.5 x .54 cms (case 2) and 12x 8x6 cms (case 3). In the third case (case 1), only paraffin blocks were received and so tumor size was not known. The age range of our three cases was 14 to 27 years with a mean age of 19 years. In the study by Zinner et al⁴ of 7 cases tumor size ranged from 7 to 20 cms while mean age was 22 years. Grossly the two cases in which specimens were available (cases 2 and 3) were nodular and encapsulated. Histologically, all three cases showed the usual histological features associated with this tumor and these were supplemented by the immunohistochemical findings. In two of the cases (cases 1 and 2), histologic criteria for malignancy^{1,3} were absent. However, in case 3, although there was little nuclear atypia and no increased mitotic activity, there was focal infiltration of adjacent normal pancreatic tissue as well as focal vascular and neural invasion. In Zinner's study⁴, four patients had local invasion and one patient had local invasion with liver metastases. However, in a study by Jung et al.¹⁴ none out of six children had either local invasion or distant metastases. Long term follow up is important in these tumors, even in the absence of any of the above features, since they can metastasize in the absence of all histologic criteria for malignancy. Therefore, even the

Figure 3. Photomicrograph showing vascular and neural invasion H&E magnification 10X.

follow up is important in these tumors, even in the absence of any of the above features, since they can metastasize in the absence of all histologic criteria for malignancy. Therefore, even the benign appearing tumors must also be classified as borderline tumors¹ and long term follow up is warranted. In our three cases, case 3 must be carefully followed up as it shows several of the histologic features suggestive of malignancy i.e. invasion into adjacent pancreatic parenchyma and focal perineural and vascular invasion.

References

1. Solcia E, Capella C, Kloppel G. Tumors of the pancreas. Atlas of Tumor Pathology, third series, Fascicle 20, Washington D.C. Armed Forces Institute of Pathology, 1995.
2. Oertel JE, Oertel YC, Heffess CS, Pancreas. In Stemberg SS ed. Diagnostic Surgical Pathology. Third ed. Philadelphia: Lippincott, Williams and Wilkins 1999, pp. 1495-96.
3. Nishihara K, Nagoshi M, Tsuneyoshi M, et al. Papillary cystic tumors of the pancreas: assessment of their malignant potential. *Cancer* 1993;71:82-92.
4. Zinner MJ, Shurbaji MS, Cameron JL. Solid and papillary epithelial neoplasms of the pancreas. *Surgery* 1990;108:475-80.
5. Matsunou H, Konishi F, Yamamichi N, et al. Solid, infiltrating variety of papillary cystic neoplasm of the pancreas. *Cancer* 1990;65:2747-57.
6. Sclafani LM, Reuter VE, Coit DG, et al. The malignant nature of papillary and cystic neoplasm of the pancreas. *Cancer* 1991;68:153-8.
7. Matsunou H, Konishi F. Papillary-cystic neoplasm of the pancreas: a clinicopathologic study concerning the tumor aging and malignancy of nine cases. *Cancer* 1990;65:283-91.
8. Lieber MR, Lack EE, Roberts JR Jr, et al. Solid and papillary epithelial neoplasm of the pancreas. An ultrastructural and immunocytochemical study of six cases. *Am J Surg Pathol* 1987;11:85-93.
9. Yamaguchi K, Miyagahara T, Tsuneyoshi M, et al. Papillary cystic tumor of the pancreas: an immunohistochemical and ultrastructural study of 14 patients. *Jpn J Clin Oncol* 1989;19:102-11.
10. Miettinen M, Partanen S, Fraki O, et al. Papillary cystic tumor of the pancreas. An analysis of cellular differentiation by electron microscopy and immunohistochemistry. *Am J Surg Pathol* 1987;11:855-65.
11. Naresh KN, Borges AM, Chinoy RF, et al. Solid and papillary epithelial neoplasm of the pancreas. Diagnosis by fine needle aspiration cytology in four cases. *Acta Cytol* 1995;39:489-93.
12. Trivedi N, Sharma U, Das PM, et al. NAC of papillary and solid epithelial neoplasm of pancreas--a case report. *Indian J Pathol Microbiol* 1999;42:369-72.
13. Lee YR, Kim Y, Koh BH, et al. Solid and papillary epithelial neoplasm of the pancreas with peritoneal metastasis and its recurrence: a case report. *Abdom Imaging* 2003;28:96-8.
14. Jung SE, Kim DY, Park KW, et al. Solid and papillary epithelial neoplasm of the pancreas in children. *World J Surg* 1999;23:233-6.