

Solid pseudopapillary epithelial neoplasm: a rare cause of intractable abdominal pain in young women

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Abstract

Solid pseudopapillary epithelial neoplasms of the pancreas are extremely rare and usually affect young women. It distinguishes itself from other pancreatic neoplasms with its low malignant potential. These tumors have a long asymptomatic period and are usually detected when they have grown to a very large size. We report a case of a 16 year old female patient who presented with intractable abdominal pain since past 1 month. A provisional diagnosis of Solid Pseudopapillary Epithelial Neoplasm was made on CT scan and confirmed by biopsy and histopathology. Early preoperative recognition of this tumour and confirmation by histopathology is emphasized because complete resection usually results in cure.

Keywords: Pseudopapillary, Epithelial, Pancreas, Solid pseudopapillary neoplasm

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Introduction

Solid and papillary epithelial neoplasms of the pancreas (SPEN) are extremely rare accounting for only 0.17 to 2.7% of all non-endocrine tumors of the pancreas [1]. It distinguishes itself from other pancreatic neoplasms with its low malignant potential and predilection for young female patients. These tumors have a long asymptomatic period and are usually detected when they have grown to a large size. The tumour is often diagnosed incidentally by abdominal examination, ultrasound, or CT scan of the abdomen. There are usually no abnormalities in the clinical laboratory

tests such as serum levels of amylase or in pancreatic cancer markers such as CA19-9, Carcinoembryonic antigen, and alpha-fetoprotein [2]. Early preoperative recognition of this tumour and confirmation by histopathology is important because complete resection usually results in cure. Metastasis is rarely seen, but when present usually involves the liver. It is therefore mandatory to establish an early diagnosis and attempt surgical excision even in large or metastasizing tumors, since complete excision offers an excellent prognosis [3].

Case Report

We present a case of a 16 year old girl who presented with a history of intractable right sided abdominal pain radiating to the back since past 1 month. On examination, a mass measuring 7 x 6 cms was felt in the right hypochondriac region. The clinical laboratory tests such as serum amylase level and tumour markers including AFP, CA19-9 and CEA were within normal limits. Computerized Tomography of the abdomen revealed well defined inhomogenously enhancing solid and cystic lesion involving the head and body of pancreas with compression of portal vein and splenic vein. A biopsy was done which confirmed the diagnosis of Solid Pseudopapillary Epithelial Neoplasm (SPEN) of pancreas. The tumour was resected and sent for histopathology. Grossly, the tumour was globular, grey brown in colour, solid – cystic and measured 8.5 x 7.5 x 3.5 cms. The tumour was surrounded by a fibrous pseudocapsule. Cut surface showed a well circumscribed, well

Manuscript received: 6th September 2017

Reviewed: 16th September 2017

Author Corrected: 24th September 2017

Accepted for Publication: 29th September 2017

encapsulated, soft, friable tumour with solid and cystic areas. The solid areas had a variegated appearance with areas of hemorrhage and necrosis. The cystic spaces were filled with bloody fluid and semisolid tissue debris. Microscopically, the tumour was well encapsulated with cells arranged in solid nests with abundant poorly supported tiny blood vessels. The tumour cells were small to medium sized, polygonal in shape with moderate amounts of clear to eosinophilic cytoplasm and relatively uniform ovoid nuclei, some showing characteristic longitudinal grooving.

The tumour cells distant to the blood vessels appeared to be degenerating, while the viable cells were seen forming a cuff around the blood vessels imparting a pseudopapillary architecture. Large areas of stromal hyalinization with intracellular and extracellular eosinophilic globules were seen in numerous foci. Groups of foamy macrophages with clusters of cholesterol clefts, surrounded by foreign body giant cells were seen focally. Tumour cell nests were seen within the fibrous capsule but not penetrating the full thickness of the capsule. Blood vessels close to the capsule showed angioinvasion.

No adjuvant chemotherapy or radiotherapy was given. The patient was doing well on three months of follow up.

Discussion

Solid pseudopapillary epithelial neoplasm is rare and far less common than the other pancreatic tumours, including ductal adenocarcinoma, cystic and neuroendocrine tumors [4]. This uncommon typically benign tumour is found mainly in young non-Caucasian women between 2nd and 3rd decades of life, although rare cases have been reported in children and men. It seems to have a predilection for Asian and African – American women. Although most of these tumors exhibit benign behavior, malignant degeneration can occur[5].

The cell of origin of SPEN remains controversial, but most researchers agree on a theory of a primitive epithelial cell as the cell from which tumour differentiates [6].

Patients with SPEN are often clinically asymptomatic. They may present with a gradually enlarging abdominal mass or complain of vague abdominal pain or discomfort. The abdomen is usually non tender on palpation, but obstructive symptoms may occur if the tumour grows large enough to compress adjacent viscera. There are usually no abnormalities in clinical laboratory tests or in pancreatic cancer markers[5].

SPEN has distinctive pathologic features. They can occur in every part of the pancreas but they are slightly more common in the tail. The size of the tumors ranges from as small as 1.5 cms to as large as 30 cms in diameter. Grossly, it appears as a large and encapsulated mass, generally well-demarcated from the remaining pancreas. Cut sections show the alternation of solid and yellowish areas with cystic, frequently necrotic and hemorrhagic zones. In smaller SPEN, there are often variable amounts of fibrosis, and cystic changes can be less prominent.

Histologically, they are generally characterized by solid areas which alternate with a pseudopapillary pattern, and cystic spaces which are the results of gradual degenerative changes occurring in the solid neoplasm. Solid areas are formed by cords of small to medium sized, polygonal, monomorphous cells, separated by small vessels which exhibit a variable degree of perivascular collagen deposition. Tumour cells present eosinophilic and vacuolar cytoplasm, around an often grooved ovoid nucleus, containing a nucleolus and dispersed chromatin.

Occasionally cells contain aggregates of hyaline, diastase resistant, PAS-positive cytoplasmic globules of varying size. Near the cystic spaces, accompanying the degenerative changes, it is possible to see aggregates of foamy histiocytes, cholesterol clefts, foreign body giant cells and hemorrhage.

The tumour tissue is usually well demarcated from the normal pancreas by a fibrous capsule [7]. Perineural invasion, angioinvasion or deep infiltration of the surrounding acinar tissue do not indicate an accelerated malignant behavior, because solid-pseudopapillary neoplasms in which the above mentioned histological criteria of aggressive behavior are not detected may also metastasize. Consequently all solid pseudopapillary neoplasms are currently classified as low-grade malignant neoplasms [8].



Figure-1: (a) Large solid cystic mass noted in the head and body of pancreas intraoperatively. (b) Excised mass measuring 8.5x7.5x3.5cms.



Figure 2: (a) Gross examination revealed a globular, grey brown, solid-cystic tumour. (b) Cut surface showed a well circumscribed, well encapsulated, soft, friable tumour with solid and cystic areas along with areas of hemorrhage and necrosis.

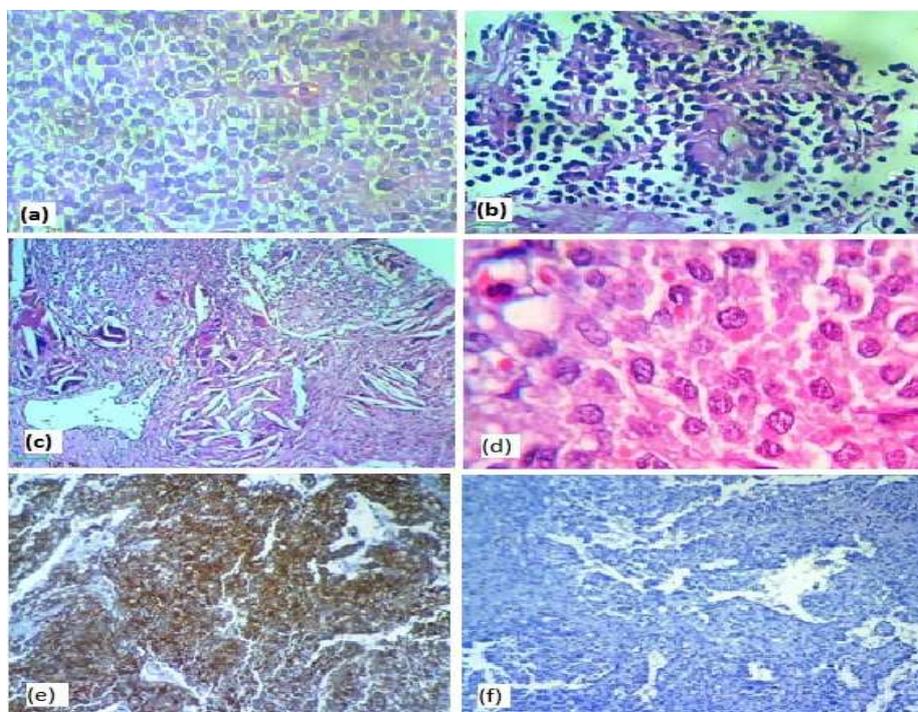


Figure 3: (a) The solid areas of the tumour was composed of small to medium sized, polygonal tumour cells with eosinophilic cytoplasm and uniform ovoid nuclei, some showing characteristic longitudinal grooving. (b) The tumour cells distant to the blood vessels appeared to be degenerating, while the viable cells were seen

forming a cuff around the blood vessels imparting a characteristic pseudopapillary architecture. (c) Clusters of foamy macrophages and cholesterol clefts were seen focally. (d) PAS positive eosinophilic globules were seen in numerous foci. (e) Tumour cells were positive for vimentin. (f) Chromogranin stain was negative.

SPEN can be confused with other cystic neoplasms. Microcystic adenomas are found predominantly in middle aged and elderly women. They are composed of several small cysts (less than 2 cms) and have a honeycomb appearance on cross section. Mucinous cystic neoplasms including cystadenoma and cystadenocarcinoma are multilocular cystic tumors that may have a solid component. The multilocularity seen on CT and MRI usually enables differentiation. It is important to distinguish between these types of cystic neoplasms because the microcystic adenoma has little malignant potential whereas the mucinous cystadenoma frequently undergoes malignant degeneration [3].

Conclusion

SPEN is a rare neoplasm that primarily affects young women[9]. While clinical signs and symptoms are relatively nonspecific, characteristic findings on imaging and confirmation by histopathology separate these from the more malignant pancreatic tumors [10]. Although surgical resection is generally curative, a close follow up is advised in order to diagnose a local recurrence or distant metastasis [9].

Funding: Nil, Conflict of interest: None

Permission of IRB: Yes

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How to cite this article?

Hegde S, Samartha V, Philipose T. R. Solid pseudopapillary epithelial neoplasm: a rare cause of intractable abdominal pain in young women. *Int J Med Res Rev* 2017;5(09):877-880. doi:10.17511/ijmrr. 2017.i09.08.