Case Report

Solid - Pseudopapillary tumor of pancreas in an elderly female: Case report of a challenging, rare entity

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ABSTRACT

Solid-Pseudopapillary tumor is a rare pancreatic neoplasm with uncertain to low malignant potential. This rare neoplasm with many pseudonyms occurs predominantly in young woman under the age of thirty years. We present a rare case of Solid-Pseudopapillary tumor of pancreas in an elderly female. The tumor was located in the tail of pancreas; clinically and radiologically diagnosed as Mucinous cystadenocarcinoma. Preoperative CT guided FNA revealed numerous papillary tissue fragments with slender branching fibrovascular cores lined by several layers of uniform tumor cells with round to oval eccentrically placed bland nucleus and abundant light basophilic, finely vacuolated cytoplasm. Occasional tumor cells showed longitudinal nuclear grooves and intracytoplasmic hyaline inclusions. Some of the papillary fragments showed mucinous change in their stalks. Cytological features were suggestive of Solid-Pseudopapillary tumor of pancreas. Histopathological examination of resected specimen confirmed the diagnosis.

Key words: Frantz's tumor, pancreatic tumors, solid cystic papillary neoplasms

INTRODUCTION

Solid-Pseudopapillary tumor of pancreas (SPTP) is the most recent descriptive term given to this characteristic and enigmatic pancreatic tumor of low malignant potential, seen predominantly in adolescent girls and young women. The cytological and histopathological features of this tumor are highly characteristic and help to differentiate this tumor from other cystic tumors of pancreas. In this report, we describe the cytological and histological findings in a rare case of Solid-Pseudopapillary tumor of the pancreas in an elderly female.

CASE REPORT

A 47 year old female presented with pain in the left hypochondrial and left lumbar region since 2 months. The pain was insidious in onset, gradually increased in intensity and radiated to the back. Per abdomen examination did not reveal any organomegaly or lump. Computed Tomography (CT) revealed a large well defined heterogeneous solid cystic mass measuring 8x7cms, in relation to the tail of pancreas. The mass was radiologically diagnosed as Mucinous cystadenocarcinoma of the pancreas. CT guided Fine Needle Aspiration (FNA) of the mass was performed and smears were referred for examination.

FNA smears were highly cellular with numerous papillary tissue fragments with slender branching fibrovascular cores lined by several layers of tumor cells. In addition there were dispersed populations of tumor cells. These tumor cells were uniform and revealed round to oval eccentrically placed nucleus with bland nuclear chromatin, inconspicuous nucleoli and abundant light basophilic, finely vacuolated cytoplasm. Binucleation and multinucleation were seen. Occasional tumor cells showed longitudinal nuclear grooves and intracytoplasmic hyaline inclusions. Some of the papillary fragments showed mucinous change in their stalks [Figure 1]. Foamy histiocytes and multinucleated histiocytic giant cells were seen in the background. Cytologic diagnosis of Solid-Pseudopapillary tumor of the pancreas was rendered.

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The patient underwent distal pancreatectomy with splenectomy. On gross examination there was a well-circumscribed and encapsulated mass in the tail of the pancreas measuring 9x6.5cm. It was partly cystic with solid friable tumor tissue and areas of haemorrhage and necrosis. Normal pancreatic tissue was identified at the periphery. The spleen weighed 170 gm and was gray brown/tan in appearance.

Microscopically, the tumor had a thick fibrous capsule. Solid areas of the tumor were composed of cords of small uniform cells, which were forming pseudopapillae [Figure 2]. Tumor cells had round to oval nucleus with finely dispersed nuclear chromatin, moderate eosinophilic granular cytoplasm and infrequent mitoses. Tumor cells showed frequent nuclear grooves and folding. Cystic areas showed hemorrhage and necrosis. The tumor was showing capsular invasion and minimal infiltration into the surrounding normal pancreatic tissue. The tumor was also infiltrating in to attached omentum. Proximal surgical resection limit was however free of tumor.

DISCUSSION

Solid-Pseudopapillary tumor of pancreas is an extremely rare neoplasm of the pancreas that normally occurs in young females.[1] It was first described in 1959 by Frantz.[2] Various synonyms for this unique exocrine epithelial neoplasm of pancreas include papillary cystic neoplasm, papillary epithelial neoplasm, papillary and cystic tumor, papillary and cystic epithelial carcinoma, papillary and solid neoplasm, solid and cystic acinar cell tumor and Gruber-Frantz's tumor. The latest consensus designation for this tumor is Solid-Pseudopapillary tumor of pancreas (SPTP). It is very difficult to establish the real incidence of SPTP of the pancreas, because of the variety of synonyms ascribed to it, as well as misdiagnosed cases as acinar cell carcinoma, nonfunctioning islet cell tumor, pancreatoblastoma, cystadenoma and even adenocarcinoma.[3]

SPTP accounts for only 1 to 2% of exocrine pancreatic tumors, and occurs predominantly in young females.[4] Very few cases have been documented in older patients.[5] In only 7% of cases, men are affected, but generally they are 10 years older than women.[6]

Clinically, this tumor causes few symptoms. SPTP of the pancreas are frequently diagnosed during investigation of gastrointestinal complaints such as abdominal pain, anorexia, weight loss, nausea and vomiting or abdominal masses, in case of large tumors, or they are incidentally found in smaller tumors (50% of the cases).[3] Obstructive jaundice is seen in case of tumor location in the head of the pancreas.[7]

Tumor is localized in head, body and tail of the pancreas in the ratio of 4:2:4. Pre-operatively fine needle aspiration can be used for obtaining a cytologic diagnosis. Cytological smears in SPTP are highly cellular and show monotonous population of small cells arranged in aggregates and papillae with fibrovascular cores. Cells have bland nuclear chromatin and may show grooving.[8]

Grossly it is usually a well circumscribed tumor ranging in size from 2-25 cm diameter. It has a variegated appearance with solid, cystic and papillary areas with foci of necrosis and hemorrhages. These degenerative changes are probably related to vascular ischemia. The clinical differential diagnosis includes all the cystic and solid lesions of the pancreas, like inflammatory pseudocyst, mucinous cystic tumors, microcystic adenoma and mucinous cystadenocarcinoma.

To the pathologist, SPTP represents a challenge. The characteristic light-microscopic feature of the tumor shows

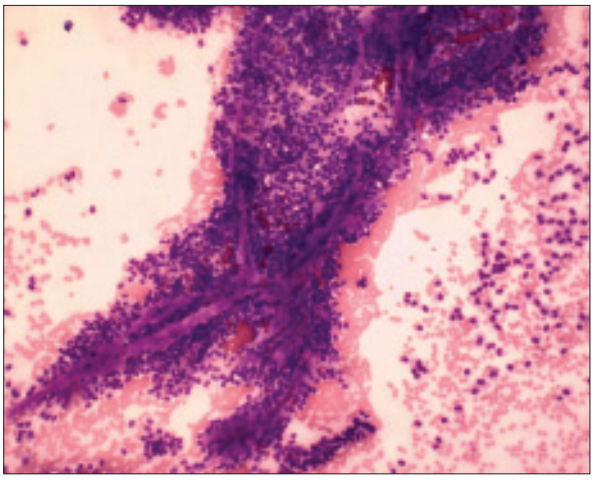

Figure 1: Papillary structures are lined by cytologically bland cells and marked mucinous change is seen in their central cores (Giemsa,10x)

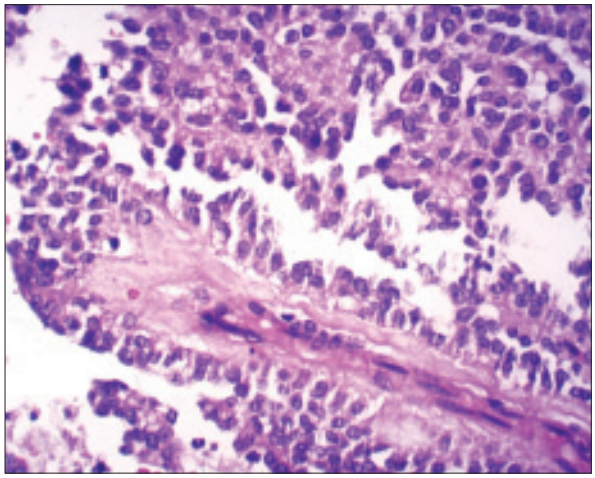

Figure 2: Small uniform tumor cells, forming pseudopapillae (H and E, 20x)

a mixture of papillary and solid patterns. The papillary structures are the fibrovascular stalks surrounded by tumor cells. The tumor cells are small, uniform cells with eosinophilic granular cytoplasm. The cystic areas contain abundant necrotic material, blood, cholesterol crystals and foam cells. Although on gross examination degenerative cystic changes may lead to confusion with cystic neoplasms of the pancreas, the characteristic microscopic features aid in the diagnosis of SPTP. These tumors are typically positive for vimentin, neuron-specific enolase (NSE), alfa1-antitrypsin, and alfa1-antichymotrypsin. Another major morphologic differential diagnosis is the islet cell tumors or pancreatic endocrine tumors (PET). Again immunohistologic analysis is the most reliable differential diagnostic tool for the separation of SPTPs and PETs. Keratin reactivity differs in the 2 tumor types; it is strong and diffuse in the PETs and weak and focal in the SPTPs. In fact, Klimstra *et al*[9] suggested that intense keratin positivity essentially excludes a diagnosis of SPTP. In contrast, SPTP consistently demonstrates uniform reactivity for vimentin. Moreover, labeling for chromogranin and synaptophysin shows focal positivity in all SPTPs. In contrast, one or both of those markers are seen diffusely in more than 90% of PETs. No immune labelling for any enteropancreatic neuropeptides is expected in SPTPs. Notohara *et al*[10] recently demonstrated that CD10 and CD56 typically were coexpressed by SPTPs, whereas PETs infrequently and only weakly expressed both of those determinants.

Histological parameters predicting the tumor's aggressive behavior include: Capsule thickness of more than 2 mm, high nuclear grade, prominent necrobiotic nests, capsular invasion into the surrounding normal pancreatic tissue and other tissues, vascular invasion and metastasis.[11]

In conclusion, SPTP is a rare low-grade malignant tumor usually seen in young women with few clinical symptoms. Radical resection is the treatment of choice and offers an excellent prognosis even in case of metastases or local invasion.

The localization and presence of local invasion affect the surgical management.[3] The various surgical procedures are Whipple's operation, pylorus preserving pancreaticoduodenectomy, distal pancreatectomy with or without splenectomy, enucleation and excision. Liver metastasis should be treated with resection.[1] The role of neoadjuvant chemotherapy is described in only a few case reports. It was done in case of advanced disease with invasion of the superior mesenteric vein with good response.[12,13] Surgical resection with adjuvant chemotherapy is also reserved for tumors with aggressive histological features.

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