Solid pseudopapillary tumor of pancreas: A lesser known entity-diagnosis and pitfalls: A case report

ABSTRACT
Solid pseudopapillary tumor (SPT) is a rare pancreatic neoplasm with a reported incidence of 0.1% to 2.7% of all pancreatic tumors. Because radiological presentation of pancreatic tumors is quite overlapping, distinctive features in fine needle aspiration cytology (FNAC) helps in its diagnosis preoperatively. Being a low-grade malignancy presenting predominantly in young females, correct preoperative diagnosis minimizes the need of extensive surgery. SPT carries good prognosis without any adjuvant chemotherapy/radiotherapy in most cases, even in the presence of metastatic disease. On the other hand, aggressive surgical resection is required for ductal adenocarcinoma which is more common pancreatic tumor (90%). We report here a case of a 49-year-old female diagnosed as SPT. The importance of the need for the radiologist, pathologist, and surgeon to be familiar with SPT is highlighted so that it is more often diagnosed as there are significant therapeutic and prognostic implications.

Key words: Cytology; females; pancreas; pancreatic tumor; solid pseudopapillary tumor (SPT)

Introduction
Solid pseudopapillary tumor (SPT) was first described by Frantz in 1959 and defined by the World Health Organization in 1996. SPT is a rare tumor because it still remains underdiagnosed despite easily identifiable cytological and histopathological features. SPT shows dual differentiation — epithelial and neuroendocrine. Differentiation along exocrine cell lines has been postulated for SPT on the basis of trypsin and chymotrypsin positivity. However, neuron-specific enolase (NSE), and synaptophysin positivity favors an endocrine origin. An undiagnosed SPT may unduly subject the patients for aggressive surgery. They may even develop diabetes mellitus postoperatively. Variable clinicopathologic and radiologic presentations of this less understood tumor will be discussed along with diagnostic pitfalls so it can be identified more frequently.

Case Report
A 49-year-old female came to the surgical oncology outpatient department (OPD) with complains of vague abdominal discomfort/pain and early satiety. There were no complains of associated fever, nausea/vomiting, diarrhea, and weight loss. Systemic examination was unremarkable. No local tenderness or lump was noted. Hematological and serological investigations were within normal limits. She had previous reports of abdominal ultrasound showing a mass lesion in the head-body region of the pancreas and fine needle
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aspiration cytology (FNAC) of pancreatic mass reported as adenocarcinoma.

Now, at our institute, contrast-enhanced CT (CECT) was done that revealed a well-defined heterogeneously enhancing exophytic solid mass lesion arising from head and body of the pancreas measuring 8.2 cm × 14 cm × 11 cm. Large necrotic component, cystic areas, internal septation, and calcification were seen with few subcentimeter lymph nodes. No pancreatic/bile duct dilatation were noted. Radiological impression was malignant pancreatic tumor with differential diagnosis of serous cystadenoma (SCN), branch duct intraductal papillary mucinous neoplasm (IPMN), and acinar carcinoma. CT-guided FNAC smears were stained with hematoxylin and eosin (H&E) stain. Hypercellular smears showed a monomorphic picture. Many branching papillary fragments with central metachromatic fibrovascular core were seen. Individual cells were small with round-to-ovoid nuclei. Nuclei were hyperchromatic with occasional grooves and fine granular chromatin [Figure 1]. Cytological diagnosis of SPT was made. Adequate surgical resection of pancreatic tumor was done. Histopathological examination of excised specimen showed tumor cells arranged in solid sheets with intervening areas of degeneration characterized by formation of pseudopapillary structures. Individual tumor cells were uniform, small/medium sized, polygonal with moderate-to-abundant eosinophilic cytoplasm, and bland vesicular nuclei [Figure 2]. Few nuclei showed indentation and indistinct nucleoli. Aggregates of foamy macrophages were additionally noted along with few cholesterol crystals and foreign body giant cell reaction. Extensive areas of coagulative necrosis and hemorrhage were noted. No lymph node involvement was seen. With these classical histological features, final diagnosis of SPT of the pancreas was made. Immunohistochemistry for Beta-catenin (β-catenin) showed nuclear positivity.

Discussion

This rare neoplasm of the exocrine pancreas is almost exclusively found in women in all age groups and races. Mean age for SPT is 25-35 years. It is rare in males, children, and older females. Recently, SPT cases in females of fifth-sixth decade have been reported. The growing incidence of SPT is due to its increased detection rate. Patients usually present
with vague gastrointestinal symptoms. The tumor is detected on routine abdominal examination and/or on imaging exploration. Being a retroperitoneal organ, pancreatic tumors may not always be palpable. SPT is characterized by long asymptomatic course and nonspecific symptoms. Therefore, it is not unusual that they are detected when grown to a remarkable size (8-10 cm).\[14,6] CT is the initial imaging modality of choice for detection, characterization of SPT, and depicting relationships with adjacent structures. Magnetic resonance imaging (MRI) is more accurate in characterizing and differentiating SPT from other tumors. Imaging study shows a well-circumscribed, lobulated exophytic lesion with a definite peritumoral capsule and minimal vascularization. Large tumors show both solid and cystic areas, whereas small lesions have only solid areas that make it difficult to differentiate from ductal adenocarcinoma. Degenerative changes without dilatation of the bile duct system are noted. SPT should always be part of differential diagnosis in any pancreatic tumor, but being a rare tumor, it often escapes detection or is misdiagnosed. Accuracy of CT alone in cystic pancreatic tumor diagnosis is 60%. With FNAC, 75-80% lesions can be definitely diagnosed as SPT or have SPT/low-grade epithelial neoplasm in differential diagnosis. Though MRI is better, CECT with synchronous FNAC in all pancreatic tumors is more cost-effective in developing countries, where correct diagnosis is required at a minimal cost. Cytological findings are quite distinctive and include hypercellular smears with many highly characteristic Chinese letter-like large branching papillary fragments having a central fibrovascular core. The cells are small and monomorphic with round-to-ovoid nuclei with little pleomorphism. Nuclei show finely granular cytoplasm, small nucleoli, and occasional grooves. The cytological findings in the present case showed numerous cellular papillary fragments composed of monomorphic, bland, round-to-oval cells as previously described in literature. Fine vacuolation in the cells as described by Pettinato et al. and nuclear grooving described by Cappelari et al. are typical findings of this tumor that were noted. Pseudorosettes and many loosely cohesive cells/bare nuclei were seen in background.\[7,8] However, intracytoplasmic inclusions were not present. SPT is misdiagnosed not only at radiology but also cytologically. It may be difficult to obtain a satisfactory aspirate at times because of extensive necrosis and hemorrhage and its retroperitoneal location. It is misdiagnosed as endocrine tumor when rosette formation is more common than papillary structures. The absence of speckled pattern of chromatin and cytoplasmic granularity rules out the possibility of endocrine neoplasm such as islet cell tumor. Pancreatoblastoma is a tumor of childhood. It can be misdiagnosed as adenocarcinoma as well when rosettes are mistaken for acinar formation. Well differentiated adenocarcinoma showing minimal atypia may be difficult to differentiate from SPT. Nuclear membrane irregularities, hyperchromatic nuclei with coarse chromatin, and necrosis favor adenocarcinoma over SPT.

In FNAC, β-catenin, E-cadherin, and cluster of differentiation (CD) 10 are good immunohistochemical markers for SPT diagnosis.\[9] Other markers are vimentin, nonspecific enolase, Alpha-1 Antitrypsin (α1-antitrypsin), Alpha 1-antichymotrypsin (α1-antichymotrypsin), and progesterone receptors. Studies are still needed to determine sensitivity and specificity of these markers on cytology smears to establish their diagnostic utility.\[2,10]

Surgery is mainstay of treatment. Some authors advocate aggressive treatment whereas others consider simple tumor resection to be adequate. Many studies favor the latter. Depending on tumor location, simple enucleation, distal pancreatectomy, pancreaticoduodenectomy or Whipple operation is done. Overall a 5-year survival is >90%, including patients with metastatic disease due to amenability of metastases to resection. Surgery is generally curative. No chemotherapy/radiotherapy is required.\[11,6,10]

Conclusion

We should maintain a high index of suspicion for the diagnosis of SPT in evaluation of pancreatic tumor of any age/sex or any type-solid/cystic, symptomatic/asymptomatic so that it is recognized and diagnosed on a more consistent basis. Clinico-radio-cytological correlation is strongly emphasized. Recognition of SPT becomes more important because of less aggressive treatment for this tumor with much better prognosis and quality of life.

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Conflicts of interest
There are no conflicts of interest.

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Kimura's disease is a rare, chronic inflammatory disorder of unknown cause. It is endemic in Asia, affecting more number of males than females, with a ratio of 3:1. The typical clinical manifestations include a triad of painless unilateral cervical lymphadenopathy or subcutaneous masses predominantly in the head and neck region, blood and tissue eosinophilia, and an elevated serum immunoglobulin E (IgE) levels.

Variable conditions both benign and malignant may mimic Kimura’s disease both clinically and on fine needle aspirates. The confirmatory diagnosis is established only by histopathological examination. We report a case of Kimura’s disease in a patient who underwent multiple investigations in view of her past history and family history of pulmonary Koch’s. Fine needle aspiration cytology (FNAC) was performed thrice with consistently similar result of reactive lymphadenitis with numerous histiocytes and eosinophilia. The final diagnosis of Kimura’s disease could finally be established only on histopathological examination.

Key words: Eosinophilia; fine needle aspiration cytology (FNAC); Kimura’s disease; lymphadenopathy; postauricular; reactive lymphadenitis

Introduction

Kimura’s disease is a benign, chronic inflammatory soft tissue disorder of unknown origin. It occurs predominantly in young adults with the age range of 27-40 years and the