

Case Report

Solid Pseudopapillary Tumor of the Pancreas: A Case Report

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Abstract

Introduction: Solid pseudopapillary tumor (SPT) of the pancreas is rare, accounting for 0.13-2.7% of all pancreatic tumors. It has specific clinical, pathological and radiological characters that make it quite different from other pancreatic tumors.

Presentation of case: A 15-year-old girl with SPT of the pancreas diagnosed after surgical resection with histopathology and immunohistochemistry confirmation. As no clear role for adjuvant treatment, she was elected for follow up.

Conclusion: Although SPT is a rare tumor without notable symptoms. Complete surgical excision is the treatment of choice. The place of chemotherapy or radiotherapy needed to be elucidated.

Keywords: Solid pseudo-papillary pancreatic tumors; pancreatic tumors; cystic tumors

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Introduction

Solid pseudopapillary tumor of the pancreas (SPT) is an uncommon pancreatic neoplasm. This study reports a case of SPT of the pancreas, and our experience in diagnosis and management over three years.

Case Report

A 15-year-old girl with no prior medical or surgical history referred to our oncology center as a case of recurrent hemorrhagic pancreatic cyst for further management. The condition started 5 months back when she started to complain from epigastric pain, nausea and vomiting with decreased appetite. On physical examination, her abdomen was not distended but tender on palpation in epigastric region. There were no palpable masses, and bowel sounds were audible. On laboratory testing, she had a serum chemistry panel and complete blood count within normal limits except CA 19.9 was 93.68 U/ml. A computed tomography scan revealed well – defined cystic lesion of 5 cm diameter with thick soft tissue wall of pancreatic body (Fig 1).



Figure 1 CT abdomen before surgical interference

Ultrasound-guided fine-needle aspiration (FNA) biopsy yielded pictures of simple hemorrhagic cyst. The patient underwent a distal pancreatectomy with splenectomy where an 8 x 4 x 1.5 cm well-circumscribed mass was removed from pancreatic body and tail. Histological examination revealed loosely cohesive round cells with uniform nuclei, clear cytoplasm with rare mitosis (Fig 2);

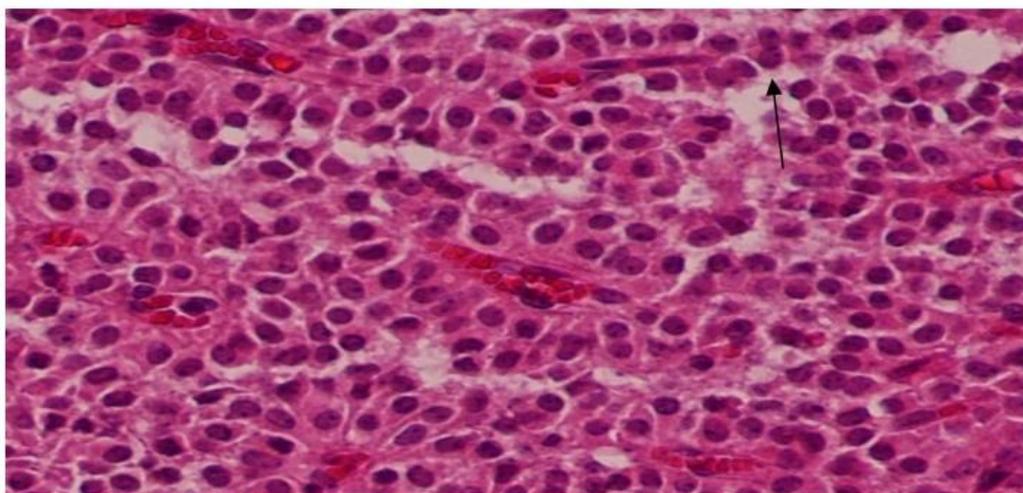


Figure 2 pseudo papillary tumor (63XH&E)

tumor size: 3 x 2 cm, presences of lymphovascular and perineural invasion, margins were uninvolved by tumor with distant from closet margin was 0.1 cm and one lymph node was negative for metastasis (0/1). Immunohistochemical staining showed neoplastic cells to be positive for vimentin, CD10, CD 56, and Bet-Catenin (Fig 3a &b),

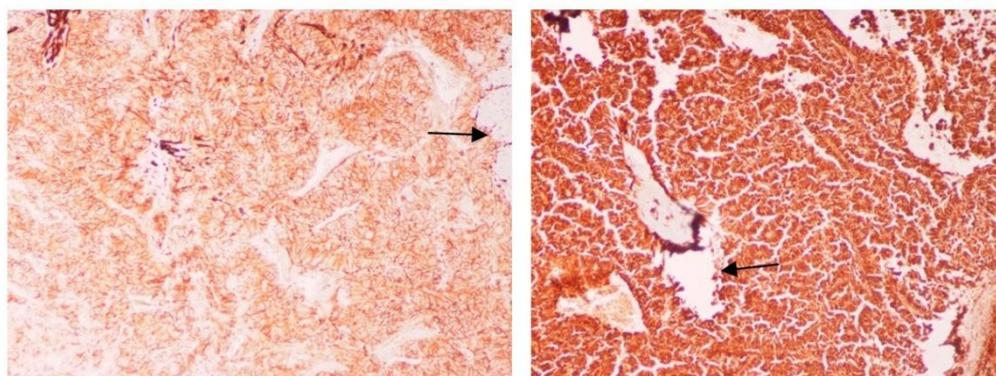


Figure 3a (IHC 40x) positive for CD 56 (left) and positive for vimentin (right)

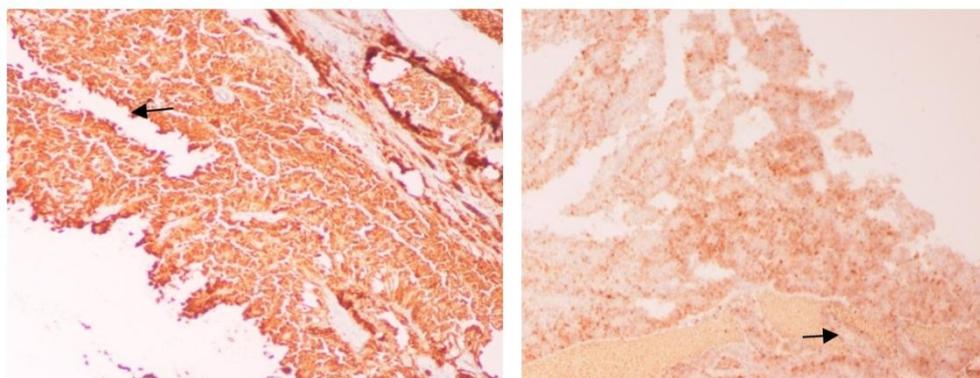


Figure 3b (IHC 40x) positive for beta-catenin (left) and positive for CD10 (right) while negative for chromogranin. Focal weak positivity is noted for neuron specific enulase (NSE)

The patient had a smooth recovery and was discharged 10 days after operation. Re-evaluation post operation by CT scan revealed no evidence of tumor residual. Also there was decrease in serum CA19.9 U/ mL with serial measurements, with last one near the normal ranges. From that time, she is under follow up without evidence of local or systemic recurrence.

Discussion

During the period from June 2011 to November 2013; 36 patients were diagnosed as primary pancreatic tumors in King Abdullah Medical City Oncology Center; our case is the first and only one diagnosed as solid pseudo papillary which represents 2.7% of all pancreatic tumors; this is corresponding to international percentages [1]. SPT is firstly described by Frantz in 1959[2].

Different names of this tumor were reported and according to its macroscopic and microscopic pathological characteristics over the years. It has been given multiple descriptive names such as papillary-cystic tumor, solid cystic tumor, papillary epithelial neoplasm, solid, and papillary neoplasm, papillary tumor of the pancreas, or Frantz's tumor until it was defined by the World Health Organization (WHO) in 1996 as a "solid pseudo papillary tumor" of the pancreas [3].

In a large retrospective review included 718 patients with SPT, more than 90 % of the patients were young women with 10:1 predominance over men and usually in the second or third decade of life [4]. Clinical features are usually vague and nonspecific including abdominal pain, dyspepsia, early satiety, and vomiting due to bulky tumor compressing the upper abdomen. Up to 20% of patients discovered accidentally either on imaging or at operation for unrelated pathology [5, 6].

The pathogenesis still remains unknown, although its tendency to affect young women has suggested that sex hormones may contribute in the origin of SPT. In the immunohistochemical staining for sex hormone-receptor proteins and clinicopathological characteristics, no difference has been attributable to gender alone [7]. Sun et al reported that 62.5% of SPT patients are infected with hepatitis B virus (HBV), which can induce over-expression of β -catenin in tumor cells, indicating that HBV infection may be involved in the pathogenesis of SPT, which, however, has not been confirmed by other researchers [8]. Radiological findings include the presence of an encapsulated mass with two components solid and cystic on either CT scan or MRI, with MRI notably better for identification of special tumor characteristics such as the presence of a capsule, hemorrhage, necrosis or cystic degeneration [9]. Pathologically, SPTs are identified as well demarcated from adjacent pancreatic tissues by fibrous capsule. Microscopic examination yields mixed solid and cystic components with regions of necrosis and hemorrhage. Characteristic findings include the presence of solid areas alternating with pseudopapillary formations, foamy histiocytes, nuclear grooves and cytoplasmic globules [10].

As the origin of this tumor remains unclear, the immunohistochemistry is needed for accurate diagnosis. SPT is typically positive for vimentin (Vim), a-1-antitrypsin (AAT), a-1- antichymotrypsin (AACT), and neuron specific enolase (NSE) [11]. However, the unique immunohistochemical features with expression of CD56 and CD10 have not reached an agreement in a recent study, also SPT cells may reveal focal immunoreactivity for cytokeratin (CK) and synaptophysin (Syn), abnormal nuclear location of β -catenin, and presence of progesterone receptors (PR), and may express galectin-3, all of which are useful in differentiating SPT from endocrine pancreatic tumor [12].

Furthermore, SPT rarely develops outside the pancreas and up to 95% are localized to it at the time of diagnosis [13]. Extra pancreatic SPT cases have been reported in retroperitoneum, liver, omentum, and mesocolon. Some of them were considered to arise from an ectopic pancreas [14].

Although they have low malignant potential, they can invade locally and when distant metastasis present, usually considered as synchronous disease to the liver and/or peritoneum. Any patient with a solid and partly cystic mass of the pancreas especially in females under 35 years of age SPT should be added to the differential diagnosis which should include microcystic adenoma, mucinous cystic neoplasm, nonfunctioning islet cell tumor, pancreatic adenocarcinoma, pancreaticoblastoma, cystic degeneration of solid neoplasm and calcified hemorrhagic pseudocyst [15].

The prognosis of SPTs is good, even with local recurrence, as well as metastases or invasions. More than 95% of patients with SPTs limited to the pancreas are cured by complete surgical excision. The overall 5-year survival was estimated to be 95% in a review of 718 patients reported in the English literature. The role of adjuvant therapy in treatment of SPT is unclear, with few studies demonstrated a role for gemcitabine and radiotherapy to downsize large tumors or treat the rare unrespectable cases [16]. Local recurrence is reported to be less than 10%, and usually within 4 years of surgery. Recurrence, local invasion, and limited metastases are not contraindications for resection, and long-term survival has also been observed in patients with SPTs [17].

Owing to the favorable prognosis and excellent long term survival, even in the presence of local recurrence or stable metastases, predictive factors of survival are difficult to identify. However in some review articles considered the presence of vascular invasion, tumor size greater than five cm and low nuclear grade as poor prognostic factors [18]. On other study done by Washington et al showed that the clinical and pathologic features of SPT, including diffuse growth, venous invasion, nuclear pleomorphism, mitotic rate, necrosis and dedifferentiation, are related to its aggressive behavior or metastatic potential [19].

Kemi and Grant evaluated the chromosomal changes in SPT and considered DNA aneuploidy, double loss of X chromosomes, trisomy for chromosome 3, unbalanced translocation between chromosomes 13 and 17 are associated with its aggressive behavior and may be indicators of its possible metastatic potential [20].

In our case, the diagnosis was based on radiological, pathological and immunohistochemical finding. Although that the patient had many apparently bad prognostic features as the presence of lymphovascular and perineural invasion with close surgical margin, but in lack of sufficient evidence from literature and good prognosis even in the presence of local recurrence or metastasis, the case was elected for only follow up and now continued for 13 months without evidence of local or systemic recurrence.

Conclusion

SPT is a rare pancreatic tumor with non specific presentation. The accuracy of diagnosis may be improved by immunohistochemical staining as such tumors are typically negative for cytokeratin and pancreatic enzyme markers, but positive for vimentin, CD 10, CD 56. Whenever possible, surgical resection is the treatment of choice with excellent long-term survival. Further studies needed for establishing guidelines for SPT treatment and clarify the role of chemotherapy or radiotherapy.

Ethical consideration

Ethical approval to conduct the study was sought from the IRB review committee before the commencement of the study.

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