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Case Report

Interesting Case of Solid Pseudopapillary Epithelial Neoplasm of Pancreas (SPEN) In A Pregnant Woman.

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Abstract

Solid pseudopapillary tumours (SPT) are rare, cystic neoplasm of the pancreas with low malignant potential. It usually occurs in females in second to third decade of life and have favourable diagnosis. We present a case of SPT which was diagnosed incidentally in a 19 year old pregnant female at 12 weeks who later had an abortion at 20 weeks of gestation. Pancreatic pseudopapillary tumors are rare neoplasms with low malignant potential and sound be kept as differential diagnosis while evaluating pancreatic tumors.

Keywords: Papillary Tumors, Pancreas.

Introduction

Solid pseudopapillary tumour (SPT) is a rare neoplasm of the pancreas, usually characterized by a well encapsulated mass lesion with a low malignant potential. It predominantly occurs in young females, in their second to third decade of life and has a favourable prognosis, with over 95% of patients reported as being disease-free after surgical resection and with less than 2% mortality.[1] Histogenesis remains unclear: acinar, endocrine, ductal and progenitor cells have all been postulated as possible origins for this tumor. It is usually asymptomatic or minimally symptomatic[1]. These tumors can be visualized and differentiated from other pancreatic lesions with a multimodality approach involving ultrasonography (US), computed tomography (CT) and magnetic resonance imaging (MRI). Complete resection is curative in most cases. In this article, we present an asymptomatic presentation of SPT in a young pregnant lady.

Case Report

A 19-year-old woman with 3 months of amenorrhea and no significant prior medical or surgical history came for routine antenatal care. On physical examination, her abdomen was soft, non-tender and non-distended. Review of systems and vitals was otherwise unremarkable.

Ultrasound was performed Philips Epiq 5G ultrasonography machine, contrast enhanced CT was done 16 slice Multidetector CT (SIEMENS® SOMATOM EMOTION 16) and MRI was done in 1.5 Tesla MRI scanner (SIEMENS® MAGNETOM AVANTO) (Fig 1) and were performed. A single live intrauterine gestation corresponding to 12 weeks 4 days was noted with NT 1.2 mm. An incidental note of a well-defined, heterogenous, solid-cystic lesion with papillary projections was noted in the head of the pancreas measuring ~ 6.8 x 5.6 x 5.8 cm with increased internal vascularity. The lesion was causing mass effect on the adjacent liver parenchyma and gall bladder superolaterally but with no obvious infiltration. Inferolaterally, it was abutting the left kidney with no extension. The lesion was abutting main portal vein and IVC, but the intraluminal flow was maintained and there no obvious infiltration into the walls of the vessels. Rest of the pancreas was unremarkable and no other focal lesions were noted elsewhere. The MPD was noted to be prominent and CBD was normal.

On laboratory examination, her liver function tests, serum amylase, lipase and total Leucocyte Counts were within normal limits. She was advised for follow up ultrasonography with nil surgical intervention during the antenatal period.

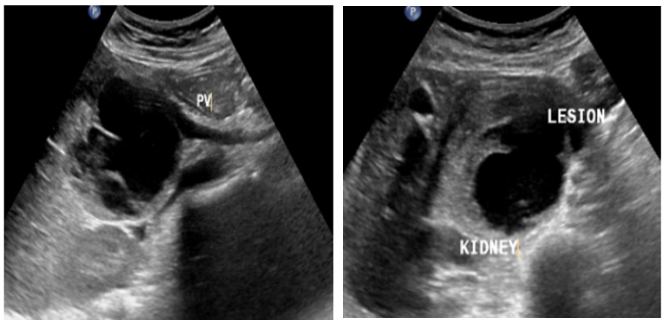

Fig. 1 Well-defined lesion in the head of the pancreas in RUQ ultrasonography.

After 2 months, she presented with spontaneous abortion, underwent dilatation and curettage after which she was referred for contrast enhanced CT abdomen and pelvis (Fig 2) for further characterisation of lesion.

On CECT, A well-defined, heterogeneously enhancing solid-cystic mass with few enhancing thin intrinsic septations was

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noted in the head of pancreas. The lesion was abutting gall bladder, liver, right kidney, portal vein and IVC as seen on ultrasonography with no discernible infiltration into the surrounding structures. No filling defect was noted in portal vein or IVC. No regional lymphadenopathy/ ascites was noted.

On MRI (Fig 2)T1 heterogeneously hypointense and T2 heterogeneously hyperintense solid-cystic lesion was noted in the head of the pancreas with restricted diffusion of solid components on DWI. Rest of the findings were consistent with CT.

Fine needle aspiration (FNA) showed tumor cells with papillary architecture, which were consistent with solid pseudopapillary neoplasm.

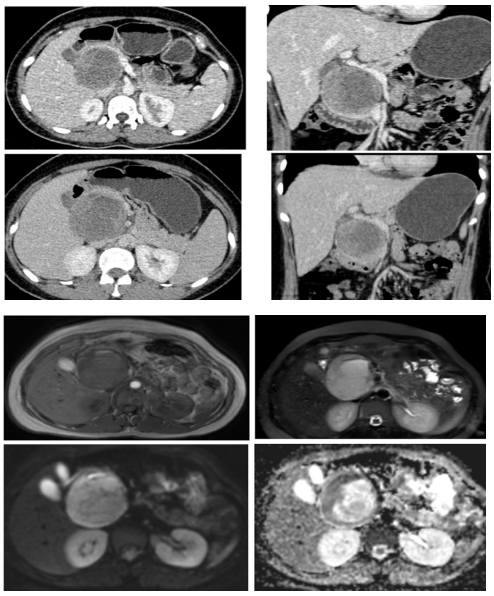

Fig.2 CECT shows heterogeneously enhancing solid-cystic lesion. MRI characteristics of the lesion – T1 hypo and T2 hyperintense heterogenous lesion showing restricted diffusion.

Discussion:

Solid pseudopapillary tumor (SPT) of the pancreas, first described by Frantz in 1959 is a rare exocrine pancreatic tumor, representing only about 1% of all tumors of the pancreas. Different names for this tumor were used until it was defined by the World Health Organization (WHO) in 1996 as a "solid pseudopapillary tumor" of the pancreas. [2] The origin of solid pseudopapillary tumors is unclear. Many investigators vouch for the theory that SPTs originate from multipotent primordial cells, whereas others suggest an extrapancreatic origin, from genital ridge angle-related cells. [3] This rare tumor seems to have a predilection for young Asian and African-American women. The male to female ratio is 1:10 and the mean age at presentation is around 22 years. It is often clinically asymptomatic or present with a gradually enlarging abdominal mass. Jaundice is a rare presentation. [4] Although most SPTs exhibit benign behavior, malignancy can occur in about 15% of cases, manifesting as metastases or invasion of adjacent structures. [3] Various studies indicated that metastasis most commonly occurred to liver and the omentum. [5, 6] The majority of such tumors are located in the pancreatic body and tail. The morphological appearance of SPT varies from solid to cystic components with cellular degenerative changes. [7] They are characteristically positive for α*1*-antitrypsin, CD56, CD10, and vimentin. [3] The majority of tumors are diagnosed through ultrasound or CT scan of the abdomen, but MRI also defines the hypervascular, well-encapsulated, round tumors with mixed cystic and solid components. Echo-endosonography may provide FNA biopsy with the possibility of pre-operative pathologic diagnosis. [3] Despite the locally aggressive features, the tumor has a low-grade malignant potential and tends to have a favourable prognosis, even in the presence of metastatic disease. Overall 5-year survival is as high as 97% in patients undergoing surgical resection[8]. Neither vascular, or perineural invasion has been a factor for predicting tumor recurrence or overall survival of patients. [4] Surgery is the treatment of choice, even in the case of distant hepatic metastasis or local recurrence. [9]

Conclusion: In conclusion, pancreatic pseudopapillary tumors are rare neoplasms with low malignant potential. Imaging features in ultrasonography, computed tomography and MRI are all helpful in accurate diagnosis.

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