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Solid Pseudopapillary Tumour of Pancreas - A rare case report

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Abstract : A 15 year old female presented to our surgical OPD with complaints of abdominal pain one week duration localized to right lower quadrant with no other significant positive history. General and systemic examination was normal. Per abdomen was soft with mild tenderness noted in right iliac fossa with no palpable mass or organomegaly. Basic blood investigations, fasting and postprandial blood sugar and coagulation profile were within normal limits. USG Abdomen, CECT Abdomen and MRI Abdomen were suggestive of Solid Pseudopapillary Tumour of Pancreas. Tumour markers like Ca 19-9, CEA, AFP were within normal limits. CT guided biopsy of the tumour were consistent with Solid Pseudopapillary Tumour of Pancreas. Subtotal pancreatectomy and appendicectomy was done. Post operative period was uneventful. Histopathology came out as Solid Pseudopapillary Tumour of Pancreas.

Keyword : Solid Pseudopapillary Tumour of Pancreas, Solid and papillary tumor, Frantz tumor.

INTRODUCTION:

Solid pseudopapillary tumour of pancreas is a rare entity usually remains asymptomatic for years and comes into light only when investigated for other reasons. Here we present a rare case of SPT identified while investigating for subacute appendicitis.

CASE HISTORY:

A 15 year old female presented to our surgical OPD with c/o abdominal pain 1 week duration, acute in onset, localized to right lower quadrant, not radiating, no aggravating/ relieving factors. No h/o fever, vomiting, bladder / bowel disturbances and no significant past history, menstrual & family history. General examination and other systemic examination was normal. Vitals were within normal limits. Per abdomen was soft with mild tenderness noted in right iliac fossa with no palpable mass/ organomegaly.

INVESTIGATIONS:

Basic blood investigations, fasting and postprandial blood sugar and coagulation profile were within normal limits. USG Abdomen showed a well defined hypoechoic mass with posterior enhancement measuring 5.2*4.5*4.8cm with a volume of 59 ml with suboptimal visualization of appendix and Probe tenderness in right iliac fossa. CECT Abdomen **(Fig:1)** showed a well encapsulated hypodense lesion to isodense noted involving distal body & tail of pancreas with well defined smooth margins showing homogenous contrast enhancement with no surrounding fat stranding of size 5.0*4.0*4.7cm suggestive of Solid Pseudopapillary Tumour of Pancreas. MRI Abdomen **(Fig:2)** showed a well defined mildly heterogenous, mildly enhancing non calcified, lobulated, solid, spherical mass of 5.0*4.5*4.5cm is seen in the tail with hypointense signal on T1 & isointense signal on T2 weighted images. Diffusion restriction is seen indicating cellularity. The tumour smoothly indents stomach with no invasion of splenic vessels and no obvious communication with the pancreatic duct. Suggestive of Solid Pseudopapillary Tumour of Pancreas. Tumour markers like Ca 19-9, CEA, AFP were within normal limits. CT guided biopsy of the tumour were consistent with Solid Pseudopapillary Tumour of Pancreas.

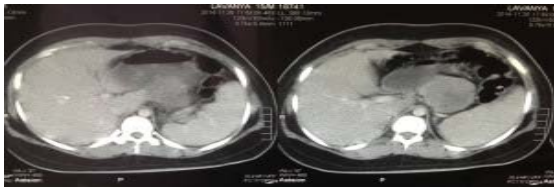

Fig:1 CECT Abdomen

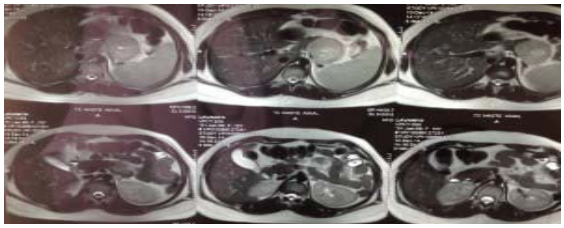

Fig:2 MRI Abdomen

OPERATIVE STEPS:

Abdomen was opened by means of a roof top incision. The gastrocolic omentum was opened and a solid well encapsulated tumour of size 5.0*4.8*4.7cm occupying the tail and distal body of pancreas encasing the splenic vessels was noted. There was no infiltration into and surrounding structures/ ascites/ peritoneal metastasis and the liver surface was normal. Appendix was 9cm in length, paracaecal and inflamed with minor adhesions to lateral peritoneum. Spleen was released from its attachments and brought forward. Posterior dissection of pancreas was done upto the level of portal vein. Splenic artery and vein were ligated separately and then subtotal pancreatectomy **(Fig:3)** was done using 75mm linear cutting staplers. The pancreatic stump was reinforced by doublebreasting with 3-0 polypropylene sutures. Appendicectomy was done and abdomen closed with 2 drainage tubes.

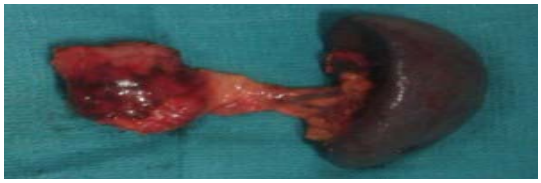

Fig:3 Subtotal Pancreatectomy With Splenectomy Specimen

Post operative period was uneventful. Patient was started on liquid diet from 3rd day and solid diet from 4th day. Drainage tubes were removed on 6th day. Serum and drainage fluid amylase and lipase, fasting and postprandial blood sugar were within normal limits. Histopathology came out as Solid Pseudopapillary Tumour of Pancreas.

DISCUSSION:

Solid pseudopapillary tumor (SPT) was first described by Frantz in 1959 as a “papillary tumor, benign or malignant” in a report of three cases; in 1981 was proposed as a distinct tumor entity, and in 1996, was defined by World Health Organization (WHO) as “solid pseudopapillary tumors” for the international histological classification of tumors of the exocrine pancreas. The molecular studies of solid pseudo-papillary tumor reveal that the tumor is similar to tumors originating in acinar cells and distinct from ductal cell adenocarcinoma. It is also known as solid and papillary tumor, solid-cystic tumor, papillary-cystic tumor, papillary epithelial neoplasm, papillary and solid neoplasm, solid and pseudopapillary epithelial neoplasm, solid and cystic acinar cell neoplasm, and Gruber-Frantz tumor. It comprises 1-2% of non-endocrine pancreatic neoplasms. The mean age 30-35 years, 90% are women and is not associated with any clinical syndrome. It seems to have a black racial predilection. The tumour is unique to pancreas mainly derived from centroacinar cells. Grossly the tumour is large (mean 9 cm), usually encapsulated, hemorrhagic, necrotic, rarely multifocal. Microscopically it is a cellular tumor, resembles pancreatic endocrine neoplasm or CNS ependymoma. It has pseudopapillae which are due to solid nests minus cells degenerating away from the small vessels; resemble rosettes in cross section. It also has round/ oval nuclei, finely stippled chromatin, nuclear grooves, indistinct nucleoli, few mitoses and also foam cells. It may have pseudocystic areas. Tumor cells infiltrate without any surrounding stromal reaction. The tumour stains positive for Vimentin, CD10, CD56, Intense membranous claudin 5 and cytoplasmic claudin 2, Estrogen and progesterone receptors, focal neuroendocrine markers, Chymotrypsin and trypsin, Nuclear and cytoplasmic beta-catenin, cyclin D1, nuclear E-cadherin, paranuclear dot like CD99. It stains negative for Chromogranin, CEA, acinar and ductal markers. It is almost always associated with mutations in exon 3 of the beta-catenin gene, causes abnormal immunostaining patterns for beta-catenin (nuclear and cytoplasmic, compared to membranous staining in normal pancreas) and overexpression of cyclin D1. The poor prognostic factors are venous invasion, high nuclear grade and “necrobiotic nests”. Metastases occurs in 10-15% to liver or peritoneum. Patients usually survive even with metastases. Surgical wide excision is the treatment of choice and it has excellent prognosis. Differential diagnosis includes Acinar cell carcinoma, Adrenal cortical tumors, Pancreatic endocrine tumor and Pancreatic pseudocyst.

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