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SOLID PSEUDOPAPILLARY NEOPLASM OF PANCREAS- CASE SERIES

Surgery

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ABSTRACT

The prevalence of solid pseudopapillary neoplasms (SPN) of the pancreas is highest in the second and third decade and primarily affects young women. SPN was classified as a rare "low grade malignant pancreatic tumour" in the 2019 WHO classification of malignancies of the digestive system. SPT has a good prognosis and is generally passive. Most common location in adults is body and tail, whereas it is head of the pancreas in children. Usually occurs as a single large mass. The gold standard investigation of choice is contrast enhanced computed tomography (CECT). Primary surgical resection is considered as the treatment of choice for SPNs. The procedure will depend on the location of the tumor. Cytohistologic examination with immunohistochemistry is required to confirm the diagnosis. Patients usually do well with a 5-year survival rate of about 97%. In this article we discuss the presentation, diagnosis, and treatment modalities of three cases of solitary pseudopapillary neoplasm of pancreas.

KEYWORDS

Pancreatic neoplasm, Solid pseudopapillary neoplasm, Pancreaticoduodenectomy

INTRODUCTION:

Pancreatic is one of the most lethal malignant neoplasms, and is usually diagnosed at an advanced stages in about 80-90% of the patients leading to very poor 1-year survival rate of 24% and a 5-year survival rate of meagre 9%.[1] Solid pseudopapillary neoplasms (SPN) of the pancreas predominantly affects young women with highest incidence in second and third decade. WHO classification of tumours of the Digestive System 2019 described SPN as a rare “low grade malignant pancreatic tumour”.[2] SPT is a relatively indolent entity with excellent prognosis. In this article we discuss the presentation, diagnosis and treatment modalities of three cases of solitary pseudopapillary neoplasm of pancreas.

Case Report:

A timid 16-year-old girl was brought by her parents with complaints vague pain in the left upper abdomen for past 5 days. There was no history of vomiting, loss of weight or appetite. There was no history of comorbid illness. On examination, her vitals were stable and a mass of size 4x4 cm was palpable in the left hypochondriac region, which was firm, non-tender and did not move with respiration. There was no organomegaly. Her baseline blood investigations were within normal limits. She underwent ultrasonogram of abdomen which revealed a cystic mass of size 7x5 cm with internal septations and solid areas. She was further evaluated with Magnetic Resonance Imaging of abdomen, which showed a heterogenous T2, STIR hyperintense and T1 hypointense large lobulated mass of size 8.7x7.1x5.6cm with central necrotic and cystic areas arising from the tail of pancreas abutting the splenic hilum and compressing the left renal vein.

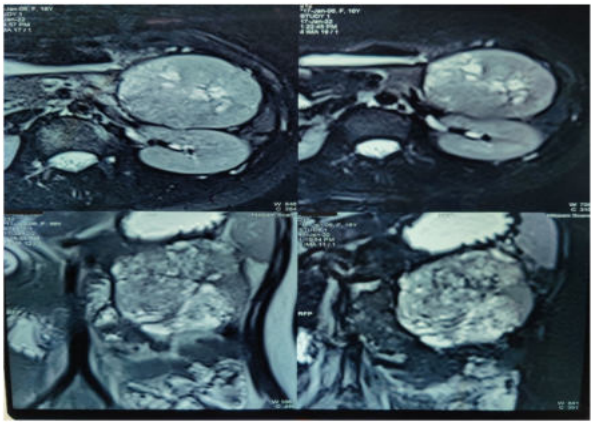

Figure 1 MR Image showing the distal pancreas mass compressing the left renal vein

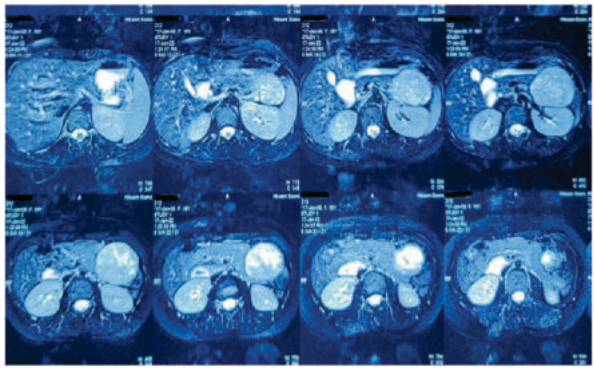

Figure 2 MR image showing mass in close proximity to the spleen.

She underwent laparotomy wherein a solid mass was seen arising from the tail of pancreas.

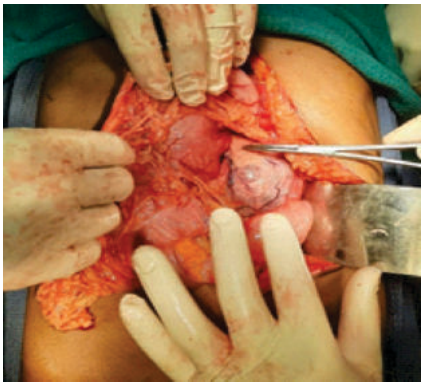

Figure 3 Intraoperative picture showing the mass arising from the tail of pancreas.

Distal pancreatectomy and splenectomy were done. Post operative period was uneventful. Histopathological examination (HPE) showed Solid Pseudopapillary Tumour of Pancreas.

Case 2:

A 20-year-old lady planning her second pregnancy underwent a routine ultrasonogram of abdomen which showed an incidental well defined solid hyperechoic vascular and partly heterogenous avascular mass of size 9.0x6.5x8.5 cm arising from the tail of pancreas. On

examination a mass of size 3x2 cm was palpable in left hypochondrium, which was tender and did not move with respiration. On further evaluation with CECT abdomen confirmed a well-defined mass arising from tail of pancreas with peripheral solid and cystic areas suggestive of solid pseudopapillary tumour of pancreas.

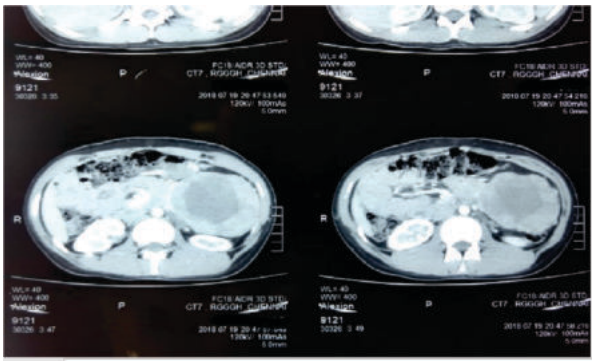

Figure 4 CECT image showing the mass arising from the tail of pancreas.

She was taken for exploratory laparotomy.

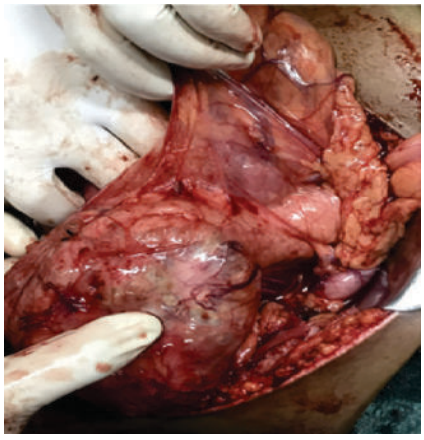

Figure 5 Intraoperative picture showing the mass arising from the tail of pancreas.

Intraoperatively, a mass of size 12x8 cm with solid and cystic components seen arising from the tail of the pancreas, adherent to the splenic vessels. Patient underwent distal pancreaticosplenectomy. She recovered well post operatively. HPE was consistent with solid pseudopapillary tumour of pancreas.

Case 3:

A young girl was complaining occasional abdominal pain and abdominal distension for the past one month. There was history of anorexia. There was no history of vomiting or comorbid illness. On examination, her vitals were stable and a mass of size 4x5 cm was palpable in the epigastric region, which was firm, non-tender and did not move with respiration. There was no organomegaly. Her baseline blood investigations were within normal limits. On evaluation with CECT abdomen confirmed a well-defined mass of size 8 arising from heads and uncinate of pancreas with peripheral solid and cystic areas suggestive of solid pseudopapillary tumour of pancreas. She was planned for Whipple's procedure after explaining the morbidity and mortality of the same. She tolerated the procedure and pancreaticoduodenectomy was done. Her postoperative period was uneventful and recovered well.

DISCUSSION

Virginia Kneeland Frantz observed a peculiar tumor of pancreas in a 19-year-old girl in 1927. She observed four similar cases and published them under the title “Tumors of Pancreas” in 1959 and described these non-functional island cell tumors as Papillary Pancreas Neoplasms.[3] WHO reclassified these tumors and named them as Solid Pseudopapillary Neoplasm (SPN) of pancreas. WHO classification of tumours of the Digestive System 2019 described SPN as a rare “low grade malignant pancreatic tumour”, accounts for less than 10% of the cystic neoplasms of pancreas and 1-2% of all pancreatic tumors.[2,4]

SPN has a female preponderance with female to male ratio up to 10:1 and most commonly occurs in second and third decade.[5] They are diagnosed at old age in males and they appear to have a poorer prognosis than their counterpart. The most common clinical presentation in adults is a nonspecific upper abdominal pain, upper abdominal mass and bloating sensation. Pediatric population most frequently present with a palpable abdominal mass.[5] Acute rupture of the tumor presenting as an acute abdomen spontaneously or following trauma is a rare presentation most common in children.[6] However, around a third of the patients had an imaging-based incidental diagnosis.[6] Basic blood workup and specific tumor markers are rendered negative. Most common location in adults is body and tail, whereas it is head of the pancreas in children.[7] Usually occurs as a single large mass with a mean diameter of 5 cm, and can be multicentric. Gross characteristics include encapsulated solid mass with cystic components indicative of hemorrhagic degeneration.[6]

SPN is a unique neoplasm due its riveting origin from the genital-ridge-related cells that were adhered to the pancreatic tissue during development.[8] The aberrant Wnt signalling appears to be the defining genetic abnormality of these neoplasms. The majority of SPNs are caused by acquired activating mutations of the CTNNB1 oncogene, which lead to nuclear -catenin accumulation and downregulation of E-cadherin, and almost all of them are connected to hyperactivation of the Wnt signalling pathway. Other genes involved include TFE3 and LEF1.[9] Expression of progesterone receptors (PR) is one of the immunohistochemical of the neoplasm, and could play a role in replication of tumoral cells considering its prevalence young women.[10]

The gold standard investigation of choice is contrast enhanced computed tomography (CECT). It provides clear definition of the tumor capsule and distinguished characterisation of the solid and cystic components, including hemorrhage, necrosis and calcifications. Contrast material provides high portal-venous phase enhancement in the solid component. Magnetic resonance imaging provides fine characterization of cystic component and hemorrhagic degeneration is visualised as hyperintense and also heterogenous signal intensity on T1- and T2- weighted images. Contrast enhanced Endoscopic Ultrasonogram can be used for diagnosis but it considered less useful than CECT but it is extremely useful for guided aspiration and biopsy.

Cytohistologic examination with immunohistochemistry is mandatory for the diagnosis of SPN.[11] Histologic examination displays two randomly mixed components combined with variable amounts of hemorrhage and pseudocystic changes: (1) a solid component, composed of poorly cohesive cells admixed with numerous myxoid fibrovascular cords and (2) a pseudopapillary component, corresponding to discohesive small and monomorphic neoplastic cells that are detached from these fibrovascular stalks.[11] Multiple studies propose the following immunohistochemical profile, β-catenin, CD99, chromogranin, trypsin, BCL10, and E-cadherin.

Primary surgical resection is considered as the the treatment of choice for SPNs. The procedure will depend on the location of the tumor. Tumors involving the body and tail of pancreas can be managed by distal pancreatectomy with splenic preservation if feasible. Tumors involving the head of pancreas shall be managed by pylorus preserving pancreaticoduodenectomy considering that tumors involving the head of pancreas are common in pediatric population and it significantly reduces postoperative morbidity.[12,13] Enucleation can be an alternative approach when the tumor is smaller than 2 cm, its distance to the main duct is more than 2–3 mm and especially when it is localized in the head of the pancreas.[14]

CONCLUSION:

Clinicians should be aware of this enitity when approaching a child or young female presenting with an abdominal mass. Radiological imaging modalities are useful in diagnosing and more often than not diagnosed as an incidental finding. Cytohistologic examination with immunohistochemistry is required to confirm the diagnosis. Primary surgical resection is the mainstay of treatment of these neoplasms. Further studies are required with regard to need for lymphadenectomy and approach to unresectable and metastatic tumors. Patients usually do well with a 5-year survival rate of about 97%.

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