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Solid Pseudopapillary Neoplasm: A Rare Presentation

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I. Background

Solid pseudopapillary neoplasm (SPN) of pancreas is a rare epithelial tumour which accounts for about 1- 3% of exocrine tumour and represent 0.2-2.7% of pancreatic carcinoma. It usually occurs in younger age group with a clear female preponderance among patients. The clinical presentation of SPN is non-specific, patient normally can present with abdominal discomfort, abdominal pain or palpable abdominal mass. In view of there is no symptoms unique to SPN and no much features to differentiate in between SPN and pancreatic pseudocyst, hence clinicians can make the wrong diagnosis and thus provide wrong management.

II. Case Presentation

We are presenting a case of 18 years old who presented with epigastric mass for 1 year associated with constitutional symptoms and abdominal pain. Per-abdomen revealed 10cm x 12 cm mass which was tender on palpation at left upper quadrant. CT abdomen reported as large pancreatic cyst causing portal vein compression and patient underwent percutaneous drainage in which the content was haemorrhagic fluid. Ca 19-9 was 5.5; CEA level less than 2; fluid for amylase showed 806 and the cytology result is negative. CT abdomen reassessment showed that the size of cystic mass remains static, subsequently decision made to proceed with surgical intervention. She underwent open distal pancreato-splenectomy and intraoperatively noted huge tumour measuring 15cm x 15 cm at distal pancreas. Histopathology revealed features of solid pseudopapillary neoplasm with negative for nodal metastasis (pT3N0M0). Patient responded well to the treatment and did not required chemotherapy.

Photo 1: Intraoperative demonstrated tumour measuring 15cm x 15cm

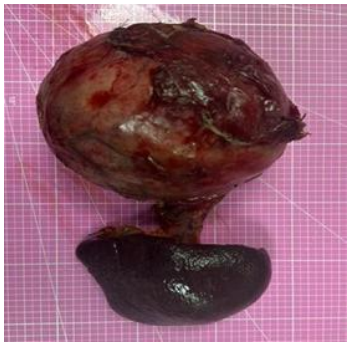

Photo 2: operative specimen

III. Discussion

SPN of pancreas is slow, growing, non-functioning tumour usually arising from the body and tail regions of pancreas, in which accounts for 1%-2% of exocrine pancreatic neoplasm. The mean age at presentation is 28.5 years, with a female-male ratio of 10:1. The clinical symptoms are not specific with the commonest presentation is abdominal pain or discomfort. Patient also can come with abdominal mass, anorexia, weight loss and fever. The standard radiological investigation used to diagnose SPN is computed tomography (CT) which will demonstrate well demarcated large heterogenous solid-cystic mass due to hemorrhagic degeneration, and calcification can be seen at the periphery of mass. MRI shows a well-defined mass with heterogenous density in the T1 and T2 images. The definite management of SPN is surgical resection which compromise from distal pancreatectomy and splenectomy to Whipple's procedure. Complete surgical resection warrants good prognosis in which can improve cure rate >95% and increase the 5 years survival rate. Our case was initially treated as pancreatic pseudocyst as patient presented with non-specific symptoms and the investigations done pointed towards the diagnosis of pancreatic pseudocyst. However, patient was counselled for surgical intervention in view of failed radiological drainage, and surprisingly the histopathological examination (HPE) report turned out to be SPN.

IV. Conclusion

It is suggested for the clinical practitioner to be more aware when dealing with the young woman who presented with pancreatic mass. A precise diagnosis can be made with the aid of radiological imaging such as CT abdomen and ultrasound guided biopsy, which can lead to better outcome and less morbidity.

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