ISSN 2515-8260 Volume 09, Issue 07, 2022

Solid Pseudopapillary epithelial neoplasm (SPEN) of Pancreas :a rare tumour of pancreas

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Solid pseudopapillary epithelial neoplasm (SPEN) of the pancreas are exceptional cystic exocrine tumour of the pancreas with low incidence rate. It is most prevalent in young women between 30 and 40 years of age [1]. It is also known as a Frantz or Hamoudi tumour. SPEN has very low-grade malignant potential, with only 5% of patients developing metastases but it is more aggressive and locally invasive in men. Early detection of this tumour of this tumour is important as , it usually has a good prognosis following surgical resection but poor prognosis factors are male sex, vascular or local invasion, size >5 cm, and necrosis or cellular atypia[2] . SPEN can present similar to other pancreatic tumours. This case report aims to describe the clinical and pathological findings in a patient diagnosed with SPEN and present the treatment and its outcome.

CASE REPORT

A 17-year-old girl presented to the general surgery outpatient department at our institute with intermittent abdominal pain in upper abdomen from 1 month, radiating to back associated with a history of vomiting, loss of appetite. On abdominal examination an intraabdominal, retroperitoneal lump was palpable in epigastric region extending to left hypochondrium region. Contrast enhanced computed tomography(CECT) showed a well-defined rounded hypodense, non – enhancing parenchymal solid mass lesion measuring 8.3 × 7.5 x 7.5 cm arising from the anterior part of the tail of the pancreas causing mass effect on greater curvature of stomach and jejunum. Patient planned for surgery. Intraoperatively, a 10 x 8 x 8 cm mass involving the distal body and tail of pancreas abutting splenic artery and vein producing gross dilatation of splenic vein. Distal pancreatectomy with splenectomy done. Specimen resected enbloc and was sent for histopathology. **Histopathology** of the resected tumour reported a solid pseudopapillary neoplasm of the pancreas. The diagnosis of solid pseudopapillary epithelial neoplasm (SPEN) of the pancreas was confirmed on histopathology of the resected tumour . The patient had uneventful recovery post-operatively.

DISCUSSION

Solid Pseudopapillary Epithelial Neoplasm of the pancreas, an exceptional cystic exocrine pancreatic tumour. This is more prevalent in young women between 30 to 40 years of age [1]. Although most of these tumours exhibit benign behaviour but malignant degeneration can occur[2]. Our patient had an uneventful recovery following the surgery. Our patient was 17 years of age, which is in the lower range of presentation age. SPEN can occur in any part of pancreas but they are more commonly located in the pancreas tail. Size can vary from 1.5

cm to as large as 30 cm in diameter. The extent of surgical resection depends upon location, metastasis and invasiveness of tumour[4-6]. The most commonly reported procedure for this tumour in the literature is a distal pancreatectomy with splenectomy. Following a good surgical resection, patients generally have an excellent prognosis, and several studies have reported a disease-free survival rate of >95 % [1,8]. The risk factors of poor prognosis are; male patients, vascular or local invasion, tumour size> 5 cm, metastasis, histology with necrosis or cellular atypia, and unresectable tumours [2,7,8,9,11]. Kim et al. reported high-grade malignancy features and recurrence as significant factors for poor prognosis [10].

Conclusion

SPEN is rare neoplasm affecting young females , it is crucial to detect SPEN at early stage. SPEN is to be consider as a differential diagnosis in young women presenting with abdominal pain and lump in upper abdomen. Early surgical resection of the tumour generally curative and has good prognosis. A close follow up is required to observe local recurrence or distant metastasis. Further studies are required to understand the pathogenesis, identify biomarkers and risk factors for recurrence.

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