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Solid Pseudo-Papillary Tumour of Pancreas: A Rare Surgical Entity

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Abstract

Solid pseudo-papillary tumour (SPT) of pancreas is a rare epithelial tumour of low malignant potential predominantly affecting young women. It is a relatively indolent entity with favourable prognosis. The atypical presentation causes a delayed or misdiagnosis of these pathology. The common location in the pancreatic head and tail often necessitates surgical intervention. This paper presents a 44-year-old female with intra-abdominal mass diagnosed with solid pseudo papillary tumour that underwent successful distal pancreatectomy and splenectomy.

Keywords: Clinical trials; Pseudo-papillary tumour of pancreas; Pancreatic surgery; Distal pancreatectomy; Pancreatic tumour

Introduction

Solid pseudo-papillary tumour (SPT) of pancreas is a rare epithelial tumour of low malignant potential accounting for 0.17 to 2.7% of primary tumours of pancreas affecting predominantly adolescent girls and young women [1]. First described by FRANTZ in 1959, referred to by variety of terms including FRANTZ tumour, solid cystic tumour, papillary cystic tumour, solid and papillary epithelial neoplasm and solid pseudo-papillary tumour of pancreas. The SPT characterised by unique clinical and pathological features. Most common in females with a ratio of 10:1, mean age 35 years, less than 10% incidence over 40 years of age. More frequent in non-caucasians with highest incidence in Japan. These are slow growing relatively benign tumours, though metastasis has been reported. It is difficult to diagnose as the presentation varies from non-specific abdominal pain to incidental findings in asymptomatic patients. Diagnosis are mainly based on radiological evidence and occasionally tissue biopsy [2]. We report a case of SPT which was successfully managed by surgical intervention.

Case History

A 47 years old female with no previous medical comorbid condition presented with acute Pain in epigastrium and left hypochondrium region associated with multiple episodes of vomiting and inability to pass flatus and stools for two days. On physical examination, there was abdominal tenderness with the presence of guarded abdomen. Computed tomography (CT) of abdomen showed a large well encapsulated heterogeneously enhancing mass of size 12.8 × 10.1 cm on left side of abdomen, possibly arising from pancreatic tail appearing as space-occupying lesion or retroperitoneal tumour (Figure 1). There was no associated Lymphadenopathy. CT guided biopsy was done but it was inconclusive. On the basis of clinical and radiological findings patient was planned for surgery. Intra-operative findings showed a large cystic lesion arising from distal pancreas present adherent to transverse colon and splenic vessels (Figure 2). En-bloc removal of mass along with distal pancreatectomy and splenectomy was performed. Post-operatively, the patient was started on liquid diet on the second post-operative day and semisolid diet on next day. The patient's recovery was uneventful and was discharged

on the fifth post-operative day. The patient has been asymptomatic for 1 year and engaged in her routine activities. Histopathology revealed SPT of pancreas with immune histochemistry showing tumour cells positive for Beta-catenin (nuclear), CD10, vimentin and negative for chromogranin. All the 15 lymph nodes excised were for malignancy.

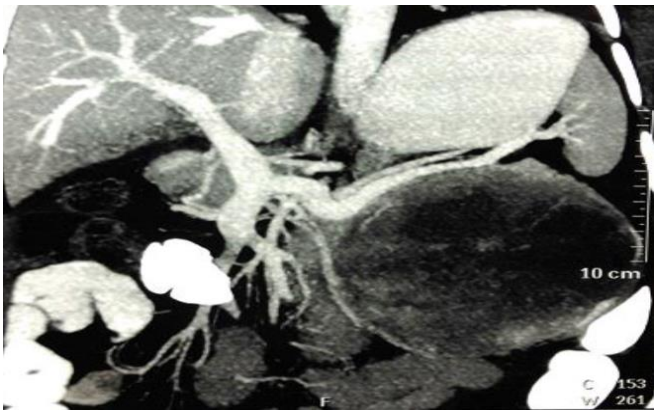

Figure 1: Computed tomography (CT) of abdomen showed a large well encapsulated heterogeneously enhancing mass of size 12.8x10.1 cm on left side of abdomen, possibly arising from pancreatic tail.

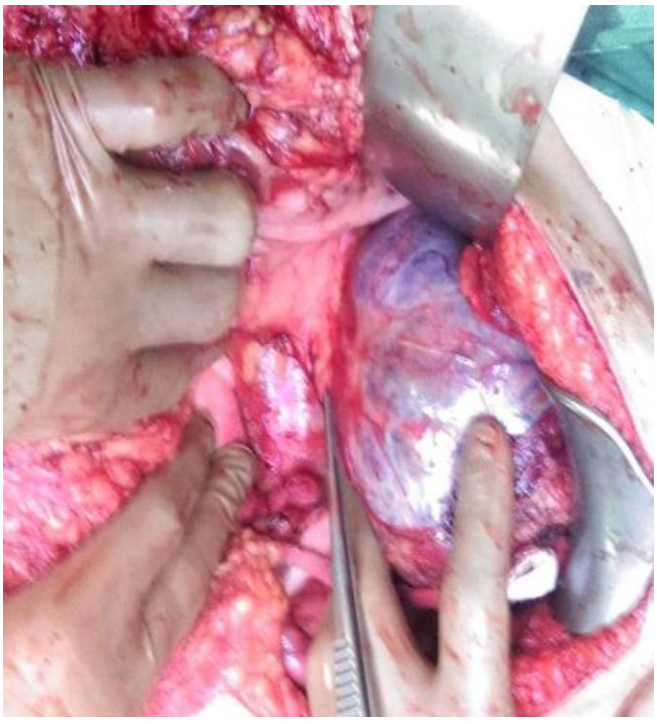

Figure 2: Large cystic lesion arising from distal pancreas.

Discussion

SPT of pancreas is rare tumour with good prognosis after complete resection. Early diagnosis due to radiological advancement and increased awareness of these neoplasm has led to their increased prevalence in recent years. Female predominance has been attributed to the proximity of primordial pancreatic cell to the ovarian ridge during development. CT being investigation of choice show well encapsulated, hypodense mass with various solid and cystic components although MRI is preferable to CT for demonstrating the presence of a capsule, haemorrhage or cystic degeneration. The tumor may involve any portion of pancreas but the head and tail are most common [2,3]. Tumour markers of SPT are APT, beta catenin, cyclin D1, D3, vimentin, antitrypsin, NSE and Progesterone. Malignant transformation seen in 15% adults and 13% children with greater risk in elderly males [4]. Metastasis are more common to liver, lymph node and peritoneum is seen in 10- 15% patients. The tumour may occur anywhere in the pancreas and presents macroscopically as a round, deceptively well demarcated lesion. Surgery is the only definitive treatment for SPT with cure rate of greater than 95% with complete resection, even in the presence of local invasion, recurrence or limited metastasis [5]. En bloc resection of tumour without intra-operative rupture with formal lymphadenectomy can be undertaken in addition to resection of synchronous/metachronous metastasis. Surgical de bulking can be effective for metastatic disease involving liver [5,6]. In contrast to other pancreatic tumours invasion of portal vein or superior mesenteric artery does not indicate tumour unresectability. The role of adjuvant therapy in treatment of SPT with few studies demonstrating a role of gemcitabine and radiotherapy to downsize the large tumour or treat un resectable disease is still unclear. Thus, early surgical intervention in a suspected case is the best treatment option as was done in our patient. Open as well as laparoscopic approach is safe and feasible option for such patients [7].

Conclusion

SPT of the pancreas are rare and frequently present in young female relatively indolent entity with favourable prognosis thus a treatable pancreatic tumor. While clinical signs and symptoms are relatively nonspecific, characteristic findings on imaging and histology separate these tumors from the more malignant pancreatic tumors. Resection remains the mainstay in treating this disease in view of its premalignant potential. Complete surgical excision is the treatment

of choice and can be achieved through an open or minimal access technique as per the surgeon expertise.

Author's Contribution

The authors contributed in study concept and design; data acquisition, drafting of the manuscript; critical revision of the manuscript for important intellectual content, material support and study supervision.

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