Volume : 2 | Issue : 12 | Dec 2013 • ISSN No 2277 - 8160

Research Paper | Medical Science

A Non-functional Pancreatic Neuroendocrine Tumour Presenting as a Pseudopancreatic Cyst-a Case Report.

Dr Sapana Bothra	3rd year Surgery PG resident
Dr Vishal S Shah	2nd year PG resident
Dr Praful M Brajprasad	1st year PG resident
Dr Parth Dalal	Assistant Prof
Dr Nina M Shah	HOU

ABSTRACT

Pancreatic neuroendocrine tumors (PNET) have an incidence of one per 100,000 individuals per year representing 1-2% of all pancreatic tumors. PNETs are a heterogeneous group with various clinical presentations and lineage. About 60% of the PNETs are non-secretory & non-functional although. Non-functional PNETS are incidentally diagnosed in most of the cases. They continue to be tumors with a low incidence and few studies directed toward early detection and management have been carried out. Currently, CT scans are the studies most used for detection. The characteristics of NF-PNETs make their detection difficult and new strategies are needed for early detection and management.

In the following case report we have discussed a patient with a non-functional PNET which was treated as a pseudopancreatic cyst based on the radiological studies which later turned out to be a PNET on biopsy studies and immunohistochemistry.The treatment is multidisciplinary, however surgical resection at present is the only curative therapy available.Surgical treatment is preferred in patients without evidence of unresectability with longer survival. Although asymptomatic non-functional tumours can be managed conservatively.(reference required).

KEYWORDS : Pancreatic neuroendocrine tumour , Pseudopancreatic cyst, cystogastrotomy

INTRODUCTION:

Pancreatic neuroendocrine tumour(PNET) belonging to the category of Gastroenteropancreatic neuroendocrine tumours(GEP-NETs) can originate within the pancreas or from similar neuroendocrine cells outside the pancreas.(Figure 2) It is unclear if whether the pancreatic tumour originates from the usual cells of Islets of Langerhans, or from diffuse neuroendocrine pluripotent cells. About 95% of the pancreatic tumours are adenocarcinomas and only 1-2% of clinically significant pancreatic neoplasms are GEP-NETs. PNETs can be functional viz gastrinomas, insulinomas, glucagonomas or VIPomas,or non-functional. About 60% of the PNETs are non-secretory & non-functional[2] which either do not secrete, or the quantity or type of product do not cause a clinical syndrome such as chromogranin A, neurotension or pancreaticpolypeptide(PP), although blood levels may rise. In the following case report ,we have discussed a patient with a non-functional PNET which was treated as a pseudopancreatic cyst based on radiological studies which turned out to be a PNET[3] on biopsy studies(Figure 1) and immunohistochemistry.

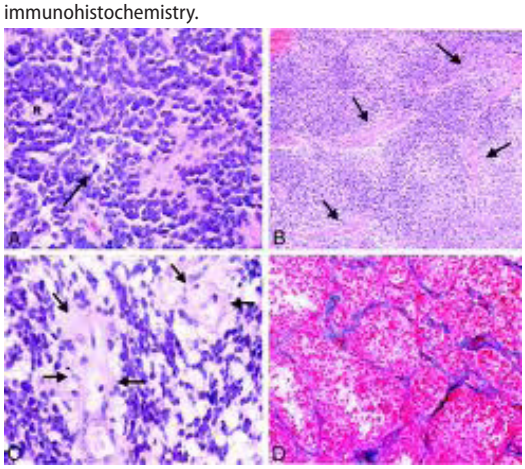


Figure 1

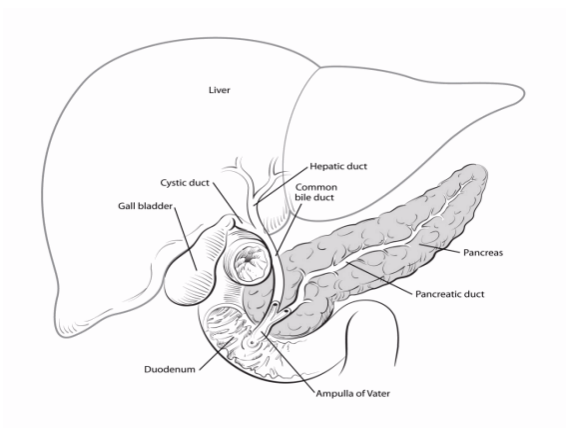


Figure 2

Case report:

A 67 year old male patient presented with complaints of upper abdominal fullness and pain and anorexia of one month duration. There was no history of vomiting, jaundice or weight loss or similar complaints in the past. On examination the patient was moderately built and nourished with no significant abnormal sign on general examination with mild tenderness on per abdominal examination.

INVESTIGATIONS:

Routine blood investigations were within normal limits except the blood sugars which were in the diabetic range. X-Ray chest and abdomen were normal. USG abdomen revealed a multiloculated well defined hypoechoic lesion of 81mm*85mm*117mm in relation to head and body of pancreas. MDCT abdomen showed a well defined thick walled cystic lesion showing multiple thin septations along anterior aspect of head & proximal body of pancreas, about 116mm*103mm*96mm with wall thickness of 10mm and the lesion compressed and

displaced stomach anteriorly, and splenic vein & artery, hepatic artery posteriorly. Rest of the pancreas, liver, gall bladder, CBD were normal. There was a perilesional lymph node of 12*11mm adjacent to the head of pancreas. (Figure 4)

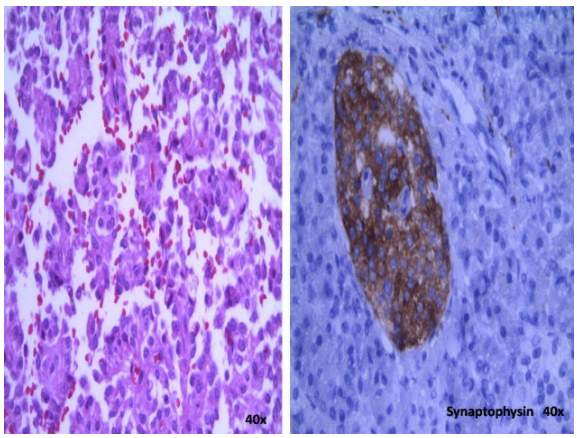


Figure 3

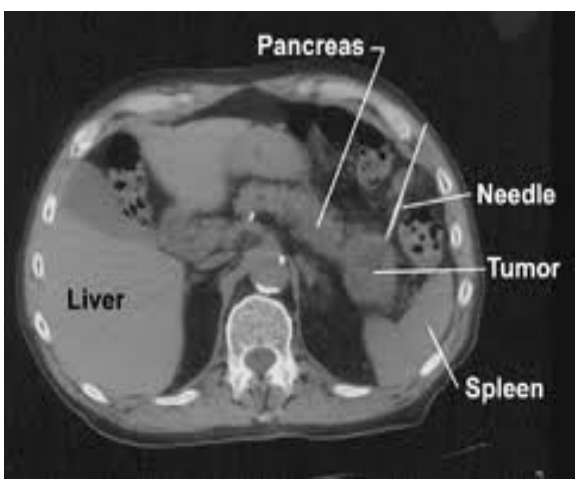


Figure 4

These features were suggestive of pancreatic pseudocyst with lymphadenopathy. MRCP revealed a cystic lesion of 12*10cm in peripancreatic region along head and body with internal septations and thickened wall. The lesion was compressing on the lesser curvature of stomach & left lobe of liver & was not separately seen from the pancreatic parenchyma and was not communicating with the pancreatic duct. Serum amylase(129) & serum lipase(340) were mildly elevated. Keeping cystic neoplasm of pancreas in mind as a differential diagnosis, tumour markers like CA 19-9(9.36) & CEA(3.83) were analysed which were within normal limits.

MANAGEMENT:

Considering the size of the cyst and its obstructing nature cystogastrotomy was planned. On exploration after doing a gastrotomy, on aspirating the cyst fresh blood was aspirated which was unlikely of a pseudocyst fluid. On opening the cyst torrential bleedin was encountered which was eventually managed. While doin the anastomosis, the wall of the cyst was found to be fragile and was tearing on passing sutures. Thin strips of cyst wall were sent for pathological examination. PATHOLOGY: Grossly irregular gray-tan soft tissue bits, sections of which on microscopy showed pink round polygonal cells in nests and chords with fibrovascular septae, mild nuclear pleomorphism and dark chromatin indicative of neuroendocrine tomour of pancreas of low grade.(Figure 3) Immunohistochemistry studies showed low grade neuroendocrine tumour of pancreas which was positive for synaptophysin and chromogranin. RESULTS: Post operative course of the patient was uneventful. Serum chromogranin was elevated(578.78). Patient was discharged on the 7th post-op day. On follow up after one month patient was asymptomatic & USG revealed a 74mm*46mm*62mm sized multiloculated cystic lesion in head of pancreas with rest of the USG being normal. Considering the age of the patient and the symptom free nature of patient, no further medications were advised. On follow up after 6 months patient was asymptomatic & USG revealed 98mm*49mm*55mm sized cystic lesion in head of pancreas.At present patient is doing well with no medications.

DISCUSSION:

NETs arise from various neuroendocrine cells whose function is to serve at various neuroendocrine interface. Pancreatic neuroendocrine tumours[4] can be functional or non-functional. Functional tumours are classified according to the hormone they secrete, for eg; gastrinoma secretes gastrin, insulinoma secretes insulin .Most functioning tumours are benign, while more than 50% of non-functioning tumours are malignant.Non functional tumours are usually slow growing and they produce symptoms mainly because of the tumor bulk or metastasis. PNETs can present with symptoms like abdominal fullness ,pain abdomen, vomiting, jaundice. All these symptoms[4] are mainly because of the tumour bulk compressing up on the neighbouring structures. Because of their non-secretory and asymptomatic nature these tumours usually present late and diagnosis is usually delayed. PNETs are fairly rare and are reported in 2-4 people per million annually.The incidence of pancreatic neuroendocrine tumors appears to be rising, due in part to heightened awareness of the disease, improved diagnostic techniques and an increased rate of incidental diagnoses during evaluations for other conditions. Contrast enhanced computed tomography[5] being the most sensitive investigation. EUS being the other useful investigation. For patients with pancreatic neuroendocrine tumors that havemetastasized, prognosis is poor. The overall prognosis & survival of the patients with PNETs is far better than those with exocrine pancreatic cancer.PNETs have been classified[6] by the WHO as well differentiated ,moderately differentiated, &poorly differentiated. The first two are of the low grade variety with good prognosis. The treatment is multidisciplinary, however surgical resection at present is the only curative therapy available. Although asymptomatic non-functional tumours can be managed conservatively.

REFERENCES

1. Franko J, Feng W, Yip L, Genovese E, Moser AJ | 2. Jensen, R. T.; Berna, M. J.; Bingham, D. B.; Norton, J. A. (2008). "Inherited pancreatic endocrine tumor syndromes: Advances in molecular pathogenesis, diagnosis, management, and controversies" | 3. Cystic pancreatic neuroendocrine tumors: a clinicopathologic study. | Singhi AD, Chu LC, Tatsas AD, Shi C, Ellison TA, Fishman EK, Kawamoto S, Schulick RD, Wolfgang CL, Hruban RH, Edil BH. | 4. Burgess A. An overview of pancreatic neuroendocrine tumoursNissen NN, Kim AS, Yu R, et al. Pancreatic neuroendocrine tumors: Presentation, management, and outcomes. | 5 Tan, E. H.; Tan, C. (2011). "Imaging of gastroenteropancreatic neuroendocrine tumors". World Journal of Clinical Oncology | 6 Klimstra, D. S.; Modlin, I. R.; Coppola, D.; Lloyd, R. V.; Suster, S. (2010). "The Pathologic Classification of Neuroendocrine Tumors". Pancreas 39 (6): 707–712. |