www.jmscr.igmpublication.org Impact Factor 3.79
ISSN (e)-2347-176x

IGM Publication Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

A Rare Case of Non-Functioning Pancreatic Neuroendocrine Tumour and Its Surgical Management

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Abstract

Pancreatic neuroendocrine tumors (PNET) consist of a rare group of neoplasms that arise from the pancreatic endocrine cells which are scattered among exocrine tissue of pancreas. They account for 2-4% of pancreatic neoplasms. The incidence is 1/100000. People frequently harbour a symptomatic PNET, and are found only on autopsy studies. They are usual sporadic but more than 10% are associated with genetic syndromes hese tumours can either be functional or non-functional attributing to the secretion of specific hormones. In this case report we are discussing about Pancreatic neuroendocrine tumour which was managed surgicaly.

INTRODUCTION

Pancreatic neuroendocrine tumors (PNET) consist of a rare group of neoplasms that arise from the pancreatic endocrine cells (1) which are scattered among exocrine tissue of pancreas (2). They account for 2-4% of pancreatic neoplasms. The incidence is 1/100000. People frequently harbour a symptomatic PNET, and are found only on autopsy studies. They are usual sporadic but more than 10% are associated with genetic syndromes eg: MEN1, Von hippel landau disease, Von recklinghauesen disease and tuberous sclerosis complex. These tumours can either be functional or non-functional attributing to the secretion of specific hormones.

CASE REPORT

This is a case report discussing a case of non-functioning Pancreatic neuroendocrine tumour and our management of the same. A fifty years old male came to the surgery department with complaints of vague abdominal discomfort on and off for the past four years. He denied any relevant medical or surgical antecedents. Physical examination of the patient revealed no organomegaly. Laboratory data was found to be

within normal range. USG abdomen was done which revealed "A peripancreatic mass seen to be causing mild displacement of the portal vein and fatty liver." Hence CECT whole abdomen (Fig 1) was done and revealed " A well defined round homogenous minimal enhancing lesion arising from the head of pancreas. No pancreatic duct dilatation. No cystic component or calcification seen - suggestive of Pancreatic neuroendocrine tumour." Chest Xray was performed and metastases was ruled out. Thereafter patient underwent Exploratory Laprotomy. Intraoperatively a globular mass of size 7x5cm (Fig 2) was seen arising from the head of the pancreas. As the tumour was found to be well away from the pancreatic duct, Enucleation of the tumour was performed which was sent for Frozen section which confirmed it to be benign in origin. The patient had an uneventful postoperative course.

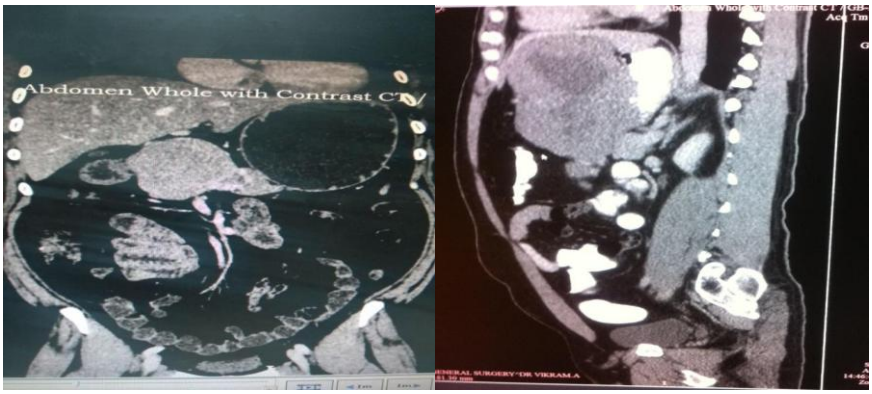

Fig 1: CECT abdomen showing PNET in the head of pancreas

Fig 2: Intra operatice picture- a well-encapsulated globular mass of size 7x5cm was found over the head of pancreas extending to the retro pancreatic space. Postoperative specimen appearance

Microscopic appearance (Fig 3)- well-encapsulated neoplasms composed of cells arranged in nests, cords, trabeculaea and islands. Individual cells showing round nuclei with peppery chromatin and moderate amounts of esinophillic cytoplasm. Intervening areas shows thin walled blood and fibrous strands.

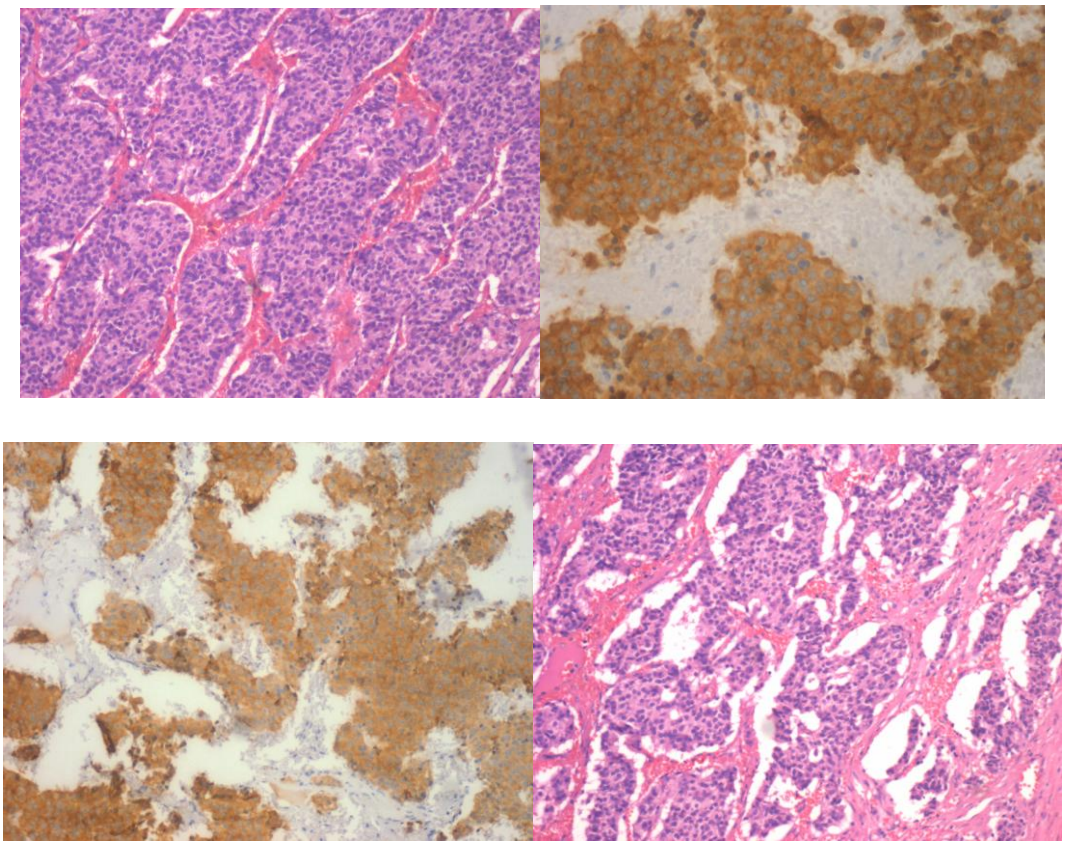

Fig 3: Histological appearance

Immunohisto chemistry was done which was positive for synaptophysin and chromogranin.ki 67<5%. Hence suggestive of Neuroendocrine tumor - type I

Patient was reviewed at regular intervals. An ultrasound abdomen was performed at 3months and a CECT whole abdomen was performed at an interval of 6months and recurrence was ruled out.

DISCUSSION

NF-PNETs are rare entities. The proportion is widely variable from 48% (by Ito El at) all the way upto 71% (Kazanjian et all). They can either be functional or non functional with a range of 15-30% being NF-PNET. (3)

CLASSIFICATION

WHO classification of tumors depends on the Metastases, Ki 67 index, Angionivasion, Tumor size, Histological differentiation and Hormonal syndrome. The prognosis is the worst for Poorly differentiated Carcinoma.

WHO Classification on NETs

1. well-differentiated tumour

1A) Benign - <2cm contained in the pancreas,no angioinvasion perineural invasion, <2mitosis/10HPF, <2%Ki-67

1B) Uncertain-2and >2 cm contained in the pancreas, angioinvasion perineural invasion, >2mitosis/10HPF, >2%Ki-

2. well- differentiated carcinoma- low grade malignant, gross local invasion and or metastases

3. poorly differentiated carcinoma – High-grade malignant >10/10 HPF,>16%Ki-67

Grading:

Gx-grade cannot be assessed

G1- well differentiated

G2-Moderately differentiated

G3- poorly differentiated

G4- Undifferentiated

In this case, the tumour was a *well-differentiated benign non-functional tumor*. Despite the size being >2cm, the low in Ki67 index confirmed its benign nature – *Grade I.*

DIAGNOSIS

Clinical features and biochemical diagnosis:

PNETs have a varied presentation and depend on the status of the tumour. PNETs if symptomatic depend on the mechanical bulk , growth pattern, fibrosis and spread of the tumor. Functional tumors can produce syndromes due to predominant hormone secreted which form the basis for their diagnosis.(4) Diagnosis of a pancreatic non functional tumour should be suspected in patient who present with a more or less asymptomatic course.

Chromogranin A - a glycoprotein is one of the most diagnostic markers for diagnosis and follows up of NET. Elevated levels are found in 60-80% of NET both functioning and non functioning(5),

DIAGNOSTIC IMAGING

Ultrasound: PNETs present as a hypoechoic aspect that can't be differentiated from other tumours and could be easily missed because of inexperience, deep situations, located in the tail, flatulence and obesity.

EUS is another modality used. It is a basic outpatient examination, which allows detailed imaging and analysis of the pancreas, helpful for preoperative assessment of NET.

CT and MRI are very useful in the diagnosis of Net. CT with contrast is the most frequently used initial imaging method. NET express SS receptors. SRS utilises octreotide (synthesised from SS) to detect the tumor. This aids in the better staging, visualisation of occult tumor and SSA treatment. SRS combined with single photon emission CT (SPECT) is more sensitive than conventional imaging for diagnosing primary and metastatic tumors. PET has not proved advantageous for NET because of low metabolic activity in most of tumors(6)

SASI test when first described for gastrinomas but has proved significant for localising other symptomatic PNET(7) The role of chemotherapy is limited.

TREATMENT

In cases of localised PNETs, the mainstay of treatment is that it should be resected. Curative resection surgery for sporadic PNETs has been standardised using the SSI test. Haynes et al. in a case series of more than 139 patients concluded that those with incidental NF-TNPs should undergo tumor resection and careful postoperative surveillance even if pathological findings are suggestive of benign disease.(8) Surgery for patients with genetic syndromes is rarely curative (9,10,11). Medical treatment with drugs such as

somatostatin analogs are used in patients who do not benefit with surgery alone.(12,13) The local treatment for liver metastasis is well established and may vary from liver resection to RFA to chemo embolisation.

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