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A Mystification Intra-abdominal Mass - A Case Report

Dr.T.Gayathri, Dr.P.Anbalagan MS, Dr.A.Rajkumar MS MCH, Dr.D.Achuthan MS, Dr.K.Anbarasan MS, Dr.R.Saravanan MS

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ABSTRACT
A neuroendocrine tumor (NET) is a type of neoplasm which arises from specialized cells which have the traits of hormone producing nerve cells and endocrine cells and thus known as neuroendocrine cells. They are found throughout the body and have the capacity to produce hormones more than the normal level which results in symptoms[7]. These hormones have a specific effect on the activity of other organs or cells in the body. In India epidemiological report in the year 2017 was stated and common NETs were pancreas(42.9%), small intestine (22.1%), colorectum (9%), appendix (2.7%)[6].
KEY WORDS- Neuroendocrine tumor, GIT NET ,GI malignancy

I. INTRODUCTION

Neuroendocrine tumors constitute about 0.5% of all malignant conditions and 2% of all malignant tumors of gastrointestinal tract[1]. The clinical profile varies between an Indian population and western countries, as previous indian studies showed stomach as common primary site but recent studies showed pancreas . Other sites of NETs are GI tract, lungs , adrenal gland , thymus etc.

Carcinoid tumors comes under a type of neuroendocrine tumor but the term is no longer used currently[1].

II. CASE PRESENTATION

70yr old male came to outpatient department with complaints of loss of weight and appetite for the past one and half months .No h/o vomiting /abdominal pain/fever /abdominal distension/bowel or bladder symptoms /jaundice/haematemesis/malena/bony pain/breathlessness. Patient is a known case of diabetes mellitus and Parkinson's disease for 12years and patient is on treatment . No history of any previous surgeries.

On examination patient with ECOG score 3 , vitals stable and per abdomen examination no significant inspectory findings . On palpation mild tenderness over right lumbar with palpable mass , margins not well defined ,liver dullness not obliterated . Carnett test -negative .No significant percussion findings, bowel sounds were present.

Initial routine blood investigation was within normal range except anemia and blood sugar under control with oral medications. Peripheral smear -Microcytic hypochromic Anemia .Stool occult blood- negative.

Ultrasound abdomen was done which showed hypoechoic mass noted in right hypochondrium measuring 9.8*6.8cm in size with increased vascularity suggested CECT abdomen.

UGI scopy showed ulceroproliferative growth involving half of d2 ,able to negotiate beyond D2 which suggestive of periampullary growth and biopsy was taken which showed features of poorly differentiated carcinoma. CECT abdomen showed 98*68mm heterodense lesion with contrast enhancement noted arising from head of pancreas .CBD ,MPD not dilated . Fat plane between the lesion and theadjacent organ normal and no evidence ofinvasion ,suggestive of ?Gastrointestinal stromal tumor .

Inspite of repeated blood transfusion(5 units preoperatively)their was fall in hemoglobin which may be due to tumor mass.

As definitive diagnosis wasn't arrived we proceeded with MRI/MRCPabdomen(FIG:1) (FIG:2) which showed right sided retroperitoneal mass of size 8.02*6.49cm ,heterointensity(FIG:3) with moderate enhancement and necrotic areas. Liver /GB /spleen/kidneys are normal .No evidence of IHBR /MPD/CBD dilatation .Bladder and pelvic structures normal.So Differential diagnosis was arrived as Right sided retroperitoneal sarcoma or Malignant GIST (duodenal involvement).

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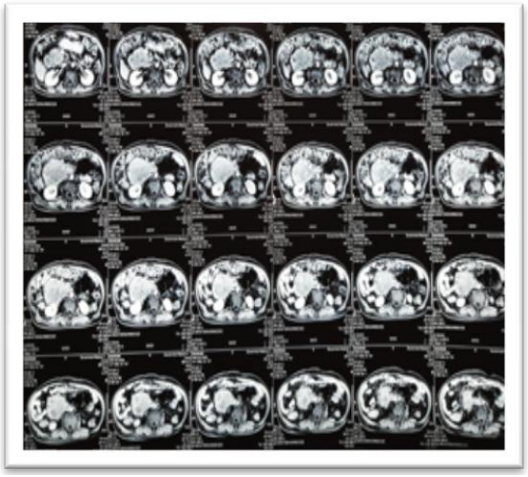

FIG 1 :MRI abdomen

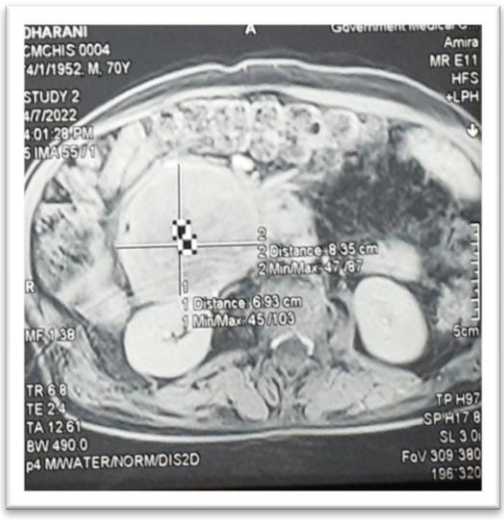

FIG 2 : T1 fat suppression contrast hyperenhancing lesion (axial)

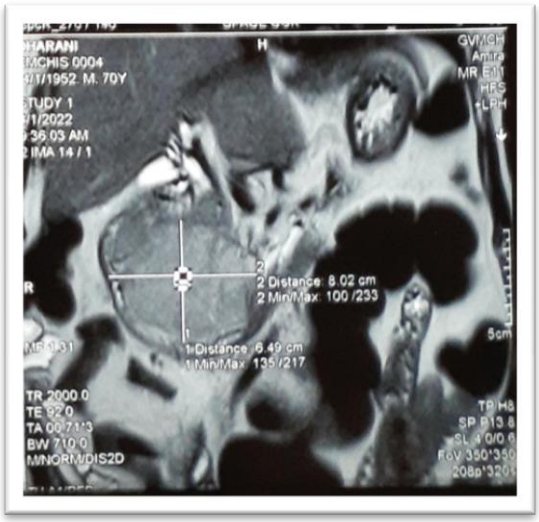

FIG 3 :MRCP – T2w (coronal) Well defined heterogenoushyperintense lesion

With the above findings we planned for EXPLORATORY LAPAROTOMY(FIG:4) electively. We proceeded with Whipple's Procedure (Resection of duodenal mass /cholecystectomy /Distal gastrectomy / pancreaticojejunostomy /end to side hepaticojejunostomy /Gastrojejunostomy/feeding jejunostomy) .Intraoperative findingsshowed cirrhotic liver /No liver /peritoneal Mets .Mass of size 8*7cm arising from d2/d3 , mobile mass with no vascular infiltration .Minimal free fluid in pelvis .GB and pancreas normal with no MPD dilatation.

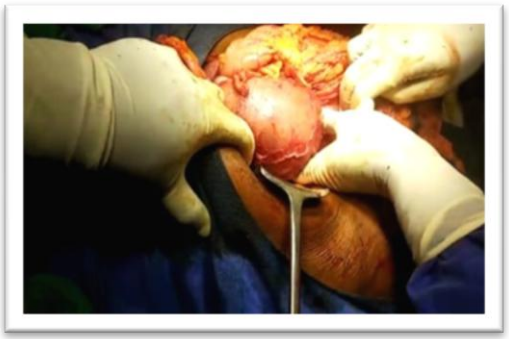

FIG 4: INTRAOPERATIVE FINDING OF TUMOR OF SIZE 8*6CM

On followup of HPE (FIG:5) report, MALIGNANT NEUROENDOCRINE TUMOR - DUODENUM 2ND PART (PERIAMPULLARY REGION) .Tumor invades the serosa extending into adjacent pancreas with resected margin free of tumor .Suggested IHC for confirmation and to rule out possibility of HEPATOID VARIANT OF ADENOCARCINOMA. Nodes are free from tumor infiltration. Immunohistological studies on poorly differentiated tumor component shows strongly positive for neuroendocrine markers-synaptophysin and chromogranin.Stains for insulin, gastrin, glucagon, somatostatin were negative. Ki67 proliferative index was <2%.

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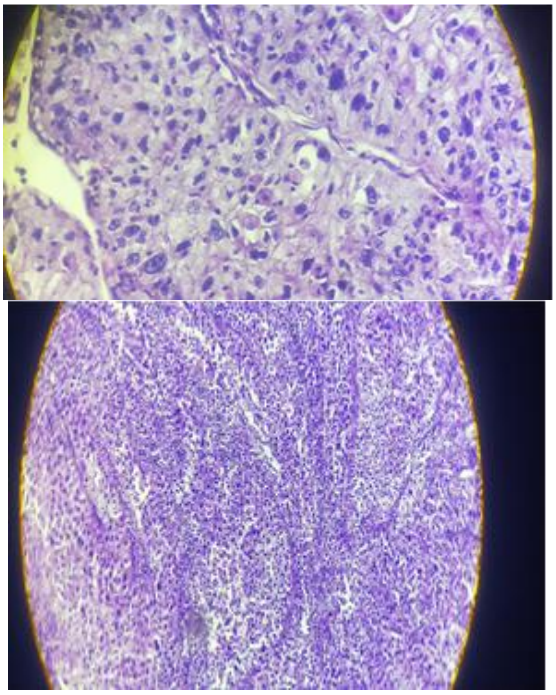

FIG : 5 SALT AND PEPPER APPEARANCE , suggestive of NETs.

III. DISCUSSION

Small bowel(midgut) NETs are more common than foregut or hindgut origin .Duodenum is relatively rare site accounting for approximately 3-4%[5]. The average age of presentation is 60-65 years with slight male predominance .Embryologic site of origin , functional status (defined as hormone secretion) and grade defines the key features behind the pathology of neuroendocrine tumors.The 2010 WHO pathology classification relies mainly on proliferation rates (ki67).

Management of NETs falls under two main therapies (I) tumor control and(II)control of symptoms due to excess hormone.Gastrinomas are the most common functional duodenal-NET (d-NET) and represent 60% of d-NET.Commonly arises from proximal duodenum involving 1st and 2nd part of duodenumand commonly spreads to local lymphnodes and hence lymphnode dissection is essential.

Periampullary duodenal NETs do not usually produce a hormonal syndrome but due to its location it may cause obstructive jaundice or bleeding. Most are small and are either ulcerative/polypoidal/nodular in nature. 50% have either lymphnode or liver metastasis[2].Depending on size of tumor local excision of tumor or whipple's is proceeded[2]. Duodenal type of NET is difficult to diagnose based on imaging due to its complex anatomy of duodeno-pancreatic region ,these tumors are challenging to diagnose and often misdiagnosed as pancreatic head tumor . EGD and direct tissue biopsy is most common and essential method of diagnosing D-NETs.

At present, the pathogenesis of duodenal-NETs is not completely clear, and further research is needed. The management protocol of D-NETs based on ENETS guidelines[1](FIG:6).

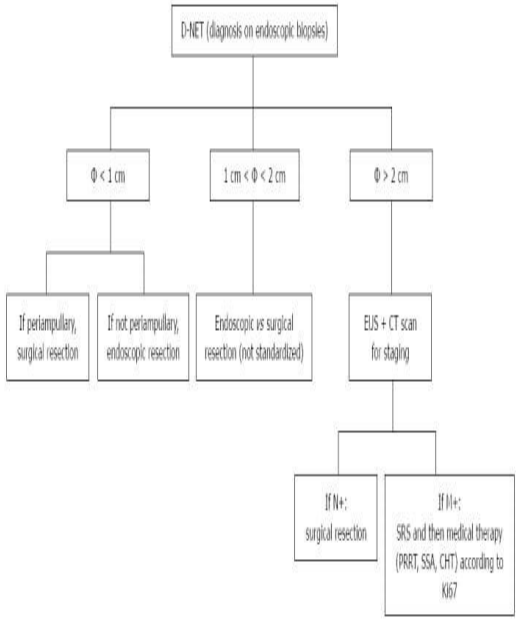

FIG :6 -European Neuroendocrine Tumor Society (ENETS) Guidelines
(EUS-endoscopic ultrasound; N+ nodes; M+ metastasis)

Immunohistochemistry is used to characterize the aggressivity of NETs,by assessing the proliferative index Ki-67, as well as neuroendocrine differentiation by assessing chromograninA (CgA) and CD56.

In my case 70yrs male presented with non specific symptoms , periampullary d-NET were definite diagnosis wasn't arrived based on imaging and hence proceeded for diagnostic laparotomy , on table we found that tumor was involving duodenum 2nd and 3rd part and hence proceeded for whipple's .

IV. CONCLUSION

I reported this case as a rare presentation due to large size of tumor as d-NETs are generally solitary and small, with 75% of tumors smaller than 2 cm in size . Thorough preoperative examination, detailed surgical evaluation, and early surgical treatment can avoid missed diagnoses and misdiagnoses of this disease.

Regular physical examination, including gastroscopy, can improvethe detection of d-NETs, diagnose the tumor stage early and predict

prognosis. Ultimately, the appropriate treatment can improve the patient's 5-year survival rate.
d-NETS at the ampulla of vater or periampullary area presents in more advanced stage of disease and have poor overall survival than d-NETs located elsewhere in duodenum.

Surgical resection of d-NETs should be followed by multisliceCT,somatostatin receptor scintigraphy(SRS) and CgA levels performed at 6-12months after surgery and then annually for minimum of 3 years[1].

Human ethics : Consent was obtained from the patient

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