ORIGINAL RESEARCH PAPER
Volume - 9 | Issue - 11 | November - 2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A SYNCHRONOUS INCIDENCE OF ASYMPTOMATIC JEJUNAL GIST IN PATIENT WITH NON- FUNCTIONAL PANCREATIC NEUROENDOCRINE TUMOR (PNET)

General Surgery

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ABSTRACT

Objective : Pancreatic NET and GIST represent rare neoplasms. This is a case report on a patient operated for PNET with incidentally found jejunal GIST inour unit.

Results : Patient underwent enucleation for pancreatic NET and jejuno-jejunal resection and anastomosis for GIST. Postoperatively patient was prescribed imatinib. Post operative period and followup were uneventful.

Conclusions : Sporadic non-functioning PNET was an incidental diagnosis, while jejuna GIST was intraoperative incidental diagnosis. Prognosis predominantly depends upon type, size and stage of presentation of PNET, as GIST was asymptomatic.

KEYWORDS

GIST, PNET, Jejunal, Non-functioning

INTRODUCTION

Neuroendocrine tumors are neoplasms that exhibit neuroendocrine phenotypes such as production of neuropeptides, large dense core secretory vesicles and the lack of neural structures.[1][2][3] Pancreatic NETs, traditionally called islet cell tumors representing 3% of all pancreatic tumors.[4]The islet cells arise from either neural crest cells or embryonic foregut endoderm.The incidence of NET has increased over the last three decades, likely from improvement in imaging modalities. PNETs have a wide spectrum of biologic behavior. Some are low grade and indolent, while others behave aggressively and have a propensity to metastasize. Overall survival with resected tumors is 55% but is only 15% with metastatic disease.

GISTs are the most common mesenchymal tumor of gastrointestinal tract that arise of the interstitial cells of Cajal. Stomach, being the most common site of origin, Jejunal GISTs are the rarest subtype. Jejunal GISTs are typically asymptomatic while small andmay be diagnosed incidentally during CT, endoscopy, during surgery or from symptomatic liver metastasis.[5] Exophytic growth with minimal or no luminal protrusion which is common, makes endoscopic diagnosis difficult. IHC and pathological tests are diagnostic.[6]

CASE REPORT

A 73yr old Hindu male patient presented with c/o. B/L flank pain to surgical OPD. Patient didn't have any other complaint like epigastric pain, epigastric fullness, weight loss, anorexia etc. patient was a known case of hypertension.

On per abdominal examination a lump was palpable in epigastrium, which was approax 2*1 cm in size and firm consistency with ill defined margins and no pulsations or peristalsis were present.

RADIOLOGICAL FINDINGS:*(Figure 1)*

On CECT, a large heterogenous soft tissue density lesion in pre-caval, pre-aortic region intending and displacing 3rd part of duodenum with possibility of nodal mass with mild necrosis likely. Biopsy correlation suggested.

On EUA, a large iso to hypoechoic mass with ill defined contour, areas of necrosis within was noted in precaval area (3x3cm). No calcification or hypervascularity. Abdominal lymphadenopathy with nodal mass likely tubercular. Biopsy from mass taken. Adv. cytoHPE, GeneXpert.
On HPE, features s/o. well diffferntiated NET WHO grade I.
And IHC- Synaptophysin and Chromogranin- Positive

INTRAOPERATIVE FINDINGS: *(Figure 2-3)*

On opening peritoneum, no metastases found in liver, omentum or peritoneal wall.

About 4*3 cm mass found in lower part of head of pancreas extending from curvature of duodenum to uncinate process. Enucleation done and sent for HPE.

A single discrete and firm precaval LN excised and sent for HPE.
Around 5-6cm from DJ junction, 2*2 cm jejunal submucosal exophytic mass was found which was resected and sent for HPE. Jejunal side-to-side anastomosis done.

RESULT

On Biopsy Report,

1. Cut surface brownish soft.
   Pancreatic neuroendocrine tumor- well differentiated WHO grade I Periphery and central areas reveal lymphoid tissue with lymphoid follicles *(Figure 4)*
2. Spindle cell neoplasm favouring
   GIST 3.2x1.5x1.2 GIST- spindle cell type, low grade low risk assessment *(Figure 5)*

DISCUSSION

The majority of PNETs are non-functional (60%-90%); however some may secrete gastrointestinal hormones that produce clinical syndromes. Most tumors are sporadic; however, some are associated with syndromes such as MEN syndrome, VHL syndrome, tuberous sclerosis, neurofibromatosis, von Recklinghausen syndrome. Tumors that produce pancreatic polypeptide, neurotensin, calcitonin are categorized as non-functional, as they do not produce a definable hormonal syndrome. They may present with distant metastasis but may not have symptoms until late in course of the disease.[7][8] Functioning tumors are likely to be benign, such as insulinomas, while glucagonomas are almost always malignant. Gastrinomas being most commonly associated NET with MEN I. multiple imaging modalities are useful including, CT, MRI, EUS, SRS, angiography. 68Ga Dotatate PET-CT seems to be an excellent modality for PNETs and may be imaging modality of choice.

Enucleation and pancreaticoduodenectomy are surgery options depending upon the size, type and location of tumor. The goal of surgical resection is to obtain negative margin even if adjacent organ has be resected.

Octreotide/lanreotide, sunitinb or everolimus are systemic treatments used to prolong progression free survival and to stabilize tumor growth in addition to relieving symptoms associated with functional tumors.[9][10]

Gastrointestinal stromal tumors are uncommon mesenchymal tumor of GIT. They can occur throughout the GIT, and small intestine is the second most frequent location after the stomach(2/3rd). Jejunal GISTs are extremely rare. Incidence of GIST is very low i.e. 2 in 1,00,000, while jejunal GIST is extremely rare accounting for 0.1-3% of all GI tumors.[11][12] The average size of symptomatic GIST tumors was 6cm compared to asymptomatic tumors measuring 2cm. generally GISTs does not spread to lymphatic system with the exception of Succinate dehydrogenase deficient GIST has lymphovascular invasion in pediatrics.[13]

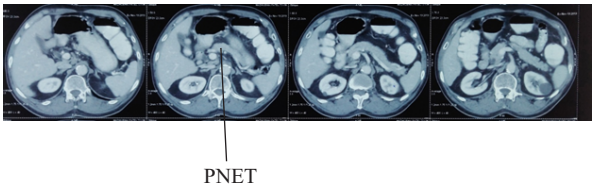
Patients presented with GI bleeding or obstruction can be evaluated by endoscopy with or without EUS and biopsy and exon mutation analysis.

The primary treatment for resectable GIST without evidence of metastasis is primary R_0 resection with consideration of adjuvant targeted therapy with imatinib mesylate after analysis of tumor size, mitotic rate and location of tumor.[14]

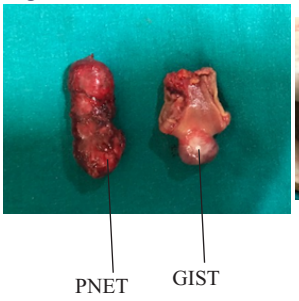
CONCLUSIONS

Management of PNETs requires a multidisciplinary approach and a thorough understanding of the biological behavior to determine optimal treatment and surgical therapy. Surgical resection continues to be the only curative therapy.

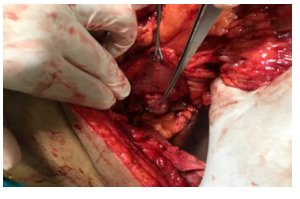
The principal and only potentially curative treatment for GIST is surgery. Survival in setting of recurrence and metastases is poor. TKI has dramatically transformed natural history of disease and the role of mutational analysis in the type and dose of TKI administered actively evolving. The role of imatinib has been expanded in neoadjuvant settings and adjuvantly following complete macroscopic and microscopic resection.[15]



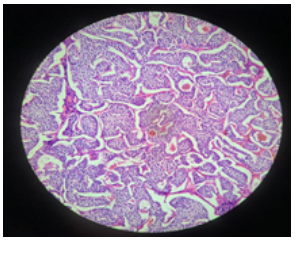
[Figure 1]



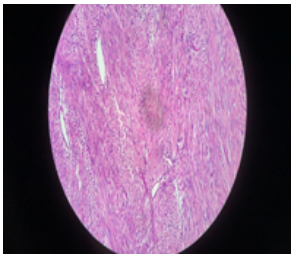
[Figure 2]



[Figure 3]



[Figure 4] PNET



[Figure 5] GIST

REFERENCES

1. *Highlights of the biology of endocrine tumours of the gut and pancreas.* Rindi G, Bordi C. 2003;10, Endocrine related cancer, pp. 427-436.
2. *Neuroendocrine tumours of the gastro-entero-pancreatic system.* Massironi S, Sciola v, Peracchi M, et al. et al. 2008;14, World J Gastroenterol, pp. 5377-5384.
3. *Neuroendocrine tumours of the pancreas.* Ehehalt F, Saeger HD, Schmidt CM, et al. et al. 2009;14, Oncologist, pp. 456-467.
4. Davies K, Conlon KC. 2009;11, Curr Gastroenteral Rep, pp. 119-127.
5. *Clinical entity andd treatment strategies for adult intussusceptions: 20 years' experience.* Wang L, Wu C, YU J, Hsiao C, Hsu C, Jao S. 2007;50, Dis Colon Rectum, pp. 1941-9.
6. *Adjuvant imatinib treatment improves recurrence free survival in patients with high-risk gastrointestinal stromal tumours (GIST).* Nilsson B, Sjolund K, Kindblom L, Meis-KIndblom J, Bumming P, Nilsson O et al. 2007;96, Br J Cancer, pp. 1658-8.
7. *Pancreatic neuroendocrine tumours (PNETs): incidence, prognosis and recent trend towards improved survival.* Halfdanarson TR, Rabe KG, Rubin J, et al. 2008;19, Ann Oncol, pp. 1727-1733.
8. *Gastrointestinal neuroendocrine tumours: pancreatic endocrine tumors.* Metz DC, Jensen RT,. 2008;135, Gastroenterology, pp. 1468-1492.
9. *Lanreotide in metastatic enteropancreatic neuroendocrine tumors.* Caplin ME, Pavel M, Cwikla JB, et al. 2014;371, N Engl J Med, pp. 224-233.
10. *Placebo-controlled, double-blind, prospective, randomised study on effect of octreotide LAR in the control of tumor growth in patients with metastatic neuroendocrine midgut tumors : a report from PROMID Study Group.* Rinke A, Muller HH, Schade Brittinger C, et al. 2009;27, J Clin Oncol, pp. 4656-4663.
11. *Bleeding Jejunal GIST : an uncommon cause of gastrointestinal bleeding.* Mohanaraj T, Hanif H, Zainal A.A. 2015;70, Med. J. Malaysia, pp. 31-32.
12. *Intestinal gastrointestinal stromal tumors presenting with gastrointestinal bleeding: different diagnostic modalities in three patients.* Koseoglu H, Solakoglu T., Akin F.E., Kilicarslan A., Akbaba S., Algin O. 2018;15, Electron. J. Gen. Med., p. em55.
13. *Pediatric gastrointestinal stromal tumors.* Pappo AS, Janeway KA. 2009;23(1), Hematol Oncol Clin North Am, pp. 15-34.
14. *NCCN Task Force report: update on management of patients with gastrointestinal tumors.* Demetri GD, von Mehren M, Antonescu CR, et al. 2010;8, J NAtl Compr Canc Netw, pp. S1-41; quiz S42-4.
15. *Surgical resection of gastrointestinal stromal tumors after treatment with imatinib.* Andtbacka R, Ng C, Scaife C, et al. 2007 Jan; 14(1), Ann Surg Oncol, pp. 14-24.