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A RARE CASE REPORT OF PRIMARY LYMPH NODE GASTRINOMA.

Gastroenterology

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ABSTRACT

Background: Gastrinoma, is a functioning Neuroendocrine tumor usually arising from gastrinoma triangle .(90%) with high metastatic potential to liver and lymph node.lymph node primary is a rare clinical presentation.hereby presenting a rare case of primary lymph node gastrinoma. **Methodology:** 26 year male diagnosed as a case of sporadic gastrinoma (2018) on basis of clinical,biochemical and imaging methods ,now presented with metabolic alkalosis, seizures, severe epigastric pain, vomiting and seizure of 2month duration .Clinical examination was unremarkable. localization studies revealed (68Ga DOTANOC scan) lesion of size 5*5cm in the gastrohepatic region.no lesion in gastrinoma triangle. After optimization patient was taken for surgery. 6*5cm cystic lesion in hepatogastric region was resected . whipples procedure done considering the occult nature of duodenal lesion even after duodenotomy and hyperechoic lesion in IOS in pancreas. Preoperatively patient condition improved markedly and gastrin level reduced to 27pg/ml from 10000pg/ml. Histopathology reported as grade 1 neuroendocrine tumour for the nodal tissue with no lesions in the pancreas or duodenum. **Results:** Our case is a primary lymph node gastrinoma as there is no lesion in the gastrinoma triangle and rest of pancreas.prognosis of primary lymph node gastrinoma is not defined so far as like metastatic lymph node gastrinoma.patient needs regular follow up to look for recurrence. **Conclusions:** Though lymph node metastasis is common with gastrinoma primary lymph node gastrinoma can be diagnosed after excluding the primary source.

KEYWORDS

Gastrinoma, Lymphnode

INTRODUCTION

Gastrinoma (Zollinger Ellison syndrome- ZES) is a rare neuroendocrine tumor (NET), with an incidence of 0.1،15 cases per million individuals worldwide[1].Gastrinoma is often misdiagnosed due to its relative rarity and lack of specific symptoms.

Typical symptoms include abdominal pain, secretory diarrhea and esophagitis. Gastrinomas are now known to most commonly arise within the wall of the duodenum (40–60%), followed by the pancreas (~20%)[2]. Primary lymph node gastrinomas have also been reported in the literature. Our's is a case of ZES where preoperative localisation and post operative Histopathology revealed a gastrinoma in a hepatogastric region, presumed to be a primary lymph node gastrinoma.

Case Summary

26 year old male patient presented with epigastric pain and vomiting of 2month duration. Patient was evaluated for same complaints in 2018 .OGD showed diffuse gastritis. CECT ABDOMEN showed heterogeneously enhancing wall thickening with prominent rugal folds in the body and fundus of stomach.

Two well defined heterogeneously enhancing mass lesion in hepatogastric region along lesser curvature and coeliac artery - nodal mass suspicious of gastrinoma. **S.gastrin** > 10000pg/ml. 68Ga DOTANOC scan reported as DOTANOC avid lesion of size 5*5cm in the gastrohepatic region and diffuse hypertrophied gastric rugal fold with minimal DOTANOC uptake.

After that patient lost follow up and now in April 2022 patient again presented with similar complaints of abdominal pain and vomiting of 2month duration. No h/o any diarrhea but h/o multiple episodes of seizures present.

On clinical examination vitals were stable and unremarkable clinical findings. OGD showed thickened gastric rugal folds and pangastritis.68Ga DOTANOC scan was repeated and showed 6.3*5.9cm DOTANOC avid lesion in the gastrohepatic region, no uptake anywhere else.

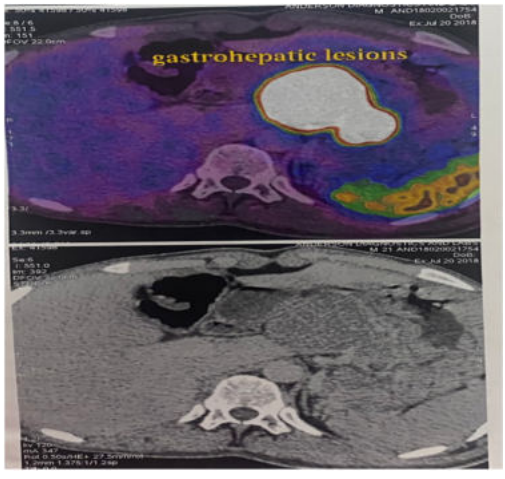

Figure 1) DOTANOC uptake in lymphnode in hepatogastric region

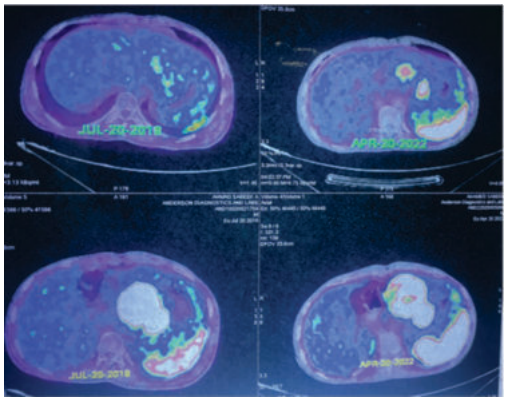

Figure 2:- repeat DOTANOC scan showing increase in size of lesion

CECT ABDOMEN showed 6*5cm lesion in the hepatogastric region likely arising from the lesser curvature of stomach and abutting the left lobe of liver . Replaced left hepatic artery arises from the left gastric artery.

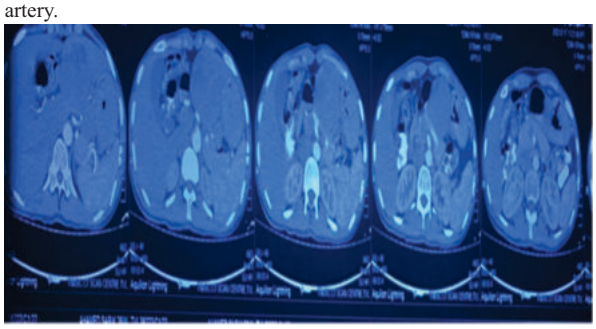

Figure 3) - CECT abdomen showing the lesion

S.chromogranin -13750ng/ml .
In the presence of 68Ga DOTANOC avid lesion and elevated s.chromogranin and s.gastrin level and thickened gastric mucosal folds it was diagnosed as a case of gastrinoma. **urine Homovanilic acid (HVA)** -3.97 (normal).**urine 5 hydroxyindole acetic acid (5HIAA)**-7.23 .To rule out MEN association patient was evaluated with S.PTH ,S. Calcium,USG neck and MRI brain and didn't show any abnormalities .So it was diagnosed as a case of sporadic gastrinoma. Patient developed metabolic alkalosis and seizures. Which on evaluation found to be because of excess acid secretion related alkalosis and the alkalosis in turn caused seizure episodes. This was managed with octreotide and PPI and adequate hydration .Patient was improved and after stabilising the patient proceeded with surgery.

Intraoperatively 6*5cm cystic lesion was seen in the gastrohepatic ligament which was getting feeding vessels from the left gastric artery.

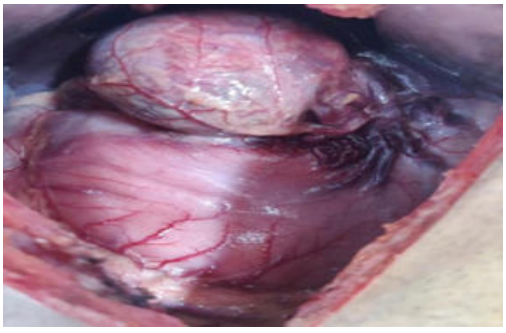

Figure 4) - intraop picture of the lesion

Lesion was completely separate from the stomach wall and pancreas. After dissecting out the lesion duodenotomy was done along the anterolateral wall of 2nd part of duodenum and entire duodenum was palpated .No lesion could be found out intraduodenally. Intraoperative USG was done and found to have a suspicious well circumscribed hyper vascular lesion in the head of pancreas near D1.

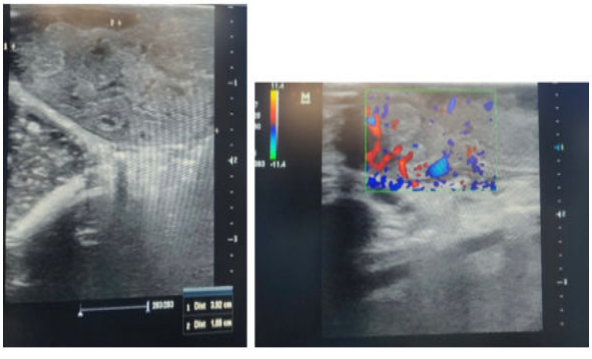

Figure 5) intraop ultrasound showing suspicious hyper vascular lesion in head of pancreas .

Considering the suspicious lesion in the head of pancreas and possibilities of missed small primary in duodenum ,we proceeded with whipples procedure.

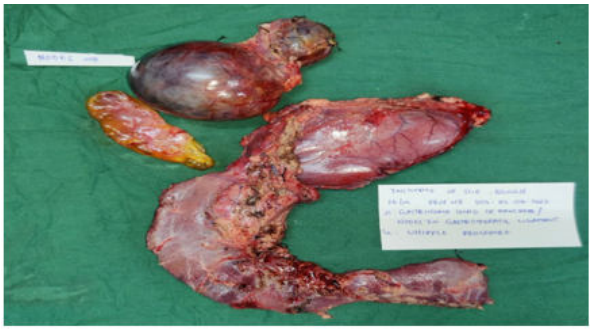

Figure 6) specimen showing hepatogastric lesion and whipples specimen

Post operatively patient improved symptomaticaly and showed improvement in electrolyte imbalance also.Post operative S.gastrin level on post op day 1 was 27pg/ml .Histopathology examination reported as grade 1 neuroendocrine tumour for the nodal tissue in the gastrohepatic ligament.no other lesions could be identified in the pancreas or duodenum.

DISCUSSION

Gastrinoma is a second most common functioning neuroendocrine tumour with incidence of 0.1 -15cases per million individuals worldwide[1].60-90 % of gastrinoma occurs within the gastrinoma triangle (passaro's triangle) ,which is an area bounded by the junction of the cystic/common bile duct posteriorly, the junction of the second/third parts of the duodenum inferiorly, and the junction of the pancreatic neck and body medially[3].90 % of it occurs in the duodenum and most common site is first part of duodenum.60-90% of gastrinomas were associated with metastases (primarily lymph-node/liver) .[4]

The existence of primary lymph node gastrinomas is a much debated and controversial topic in the literature. There have been many reports of patients with Zollinger - Ellison syndrome (ZES) who had only lymph nodes resected and were cured as assessed by biochemical testing and imaging studies in both the short and long term. Norton et al. showed in a series of 176 patients with ZES (138 sporadic, 38 MEN1) that 26 of 45 patients were disease free soon after lymph node only resection[5] .Norton et al.emphasized that a missed duodenal primary tumor could easily be mistaken for a primary lymph node gastrinoma. Even when imaging is negative for sporadic ZES, routine surgical exploration is warranted to minimize the chance of metastasis to the liver. Of those gastrinomas not localized to the duodenum or pancreas, the most common site of gastrinoma occurrence is within a lymph node found in the gastrinoma triangle. It has been theorized that pancreatic stem cells from the ventral pancreatic bud become dispersed and are incorporated into lymph tissue and the duodenal wall during embryonic development, which is supported by pathological studies that report findings of neuroendocrine cells in the region almost exclusively defined by the gastrinoma triangle[2].Intraoperative techniques used to localize a gastrinoma include sonogram, transillumination, and duodenotomy, which have been shown to detect significantly more gastrinomas (94% vs. 64%)[2], resulting in a higher cure rate. Our case is a primary lymph node gastrinoma as there is no lesion in the gastrinoma triangle or in rest of pancreas other than the lymphnode mass seen in the gastrohepatic region.Prognosis of primary lymph node gastrinoma is not defined so far as like metastatic lymph node gastrinoma.patient needs regular follow up to look for recurrence.

CONCLUSION

Once a diagnosis of sporadic gastrinoma has been made, a complete intraoperative search for a primary, including duodenotomy with total tumor excision should be the standard of care to maximize the chance for cure and minimizing further morbidity and mortality associated with either a repeat operation or metastasis.A missed duodenal primary tumor could easily be mistaken for a primary lymph node gastrinoma.

REFERENCES

[1] Zhang W-D, Liu D-R, Wang P, Zhao J-G, Wang Z-F, Chen L. Clinical treatment of gastrinoma: A case report and review of the literature. Oncology Letters 2016;11:3433–7. https://doi.org/10.3892/ol.2016.4397.
[2] Teng A, Haas C, Lee DY, Wang J, Lung E, Attiyeh F. Primary lymph node gastrinoma. A diagnosis of exclusion. A case for duodenotomy in the setting of a negative imaging for primary tumor: A case report and review of the literature. Int J Surg Case Rep

2014;5:849–52. https://doi.org/10.1016/j.ijscr.2014.08.019.
[3] Stabile BE, Morrow DJ, Passaro E. The gastrinoma triangle: operative implications. Am J Surg 1984;147:25–31. https://doi.org/10.1016/0002-9610(84)90029-1.
[4] Krampitz GW, Norton JA. Current management of the Zollinger-Ellison syndrome. Adv Surg 2013;47:59–79. https://doi.org/10.1016/j.yasu.2013.02.004.
[5] Norton JA, Alexander HR, Fraker DL, Venzon DJ, Gibril F, Jensen RT. Possible primary lymph node gastrinoma: occurrence, natural history, and predictive factors: a prospective study. Ann Surg 2003;237:650–7; discussion 657-659. https://doi.org/10.1097/01.SLA.0000064375.51939.48.