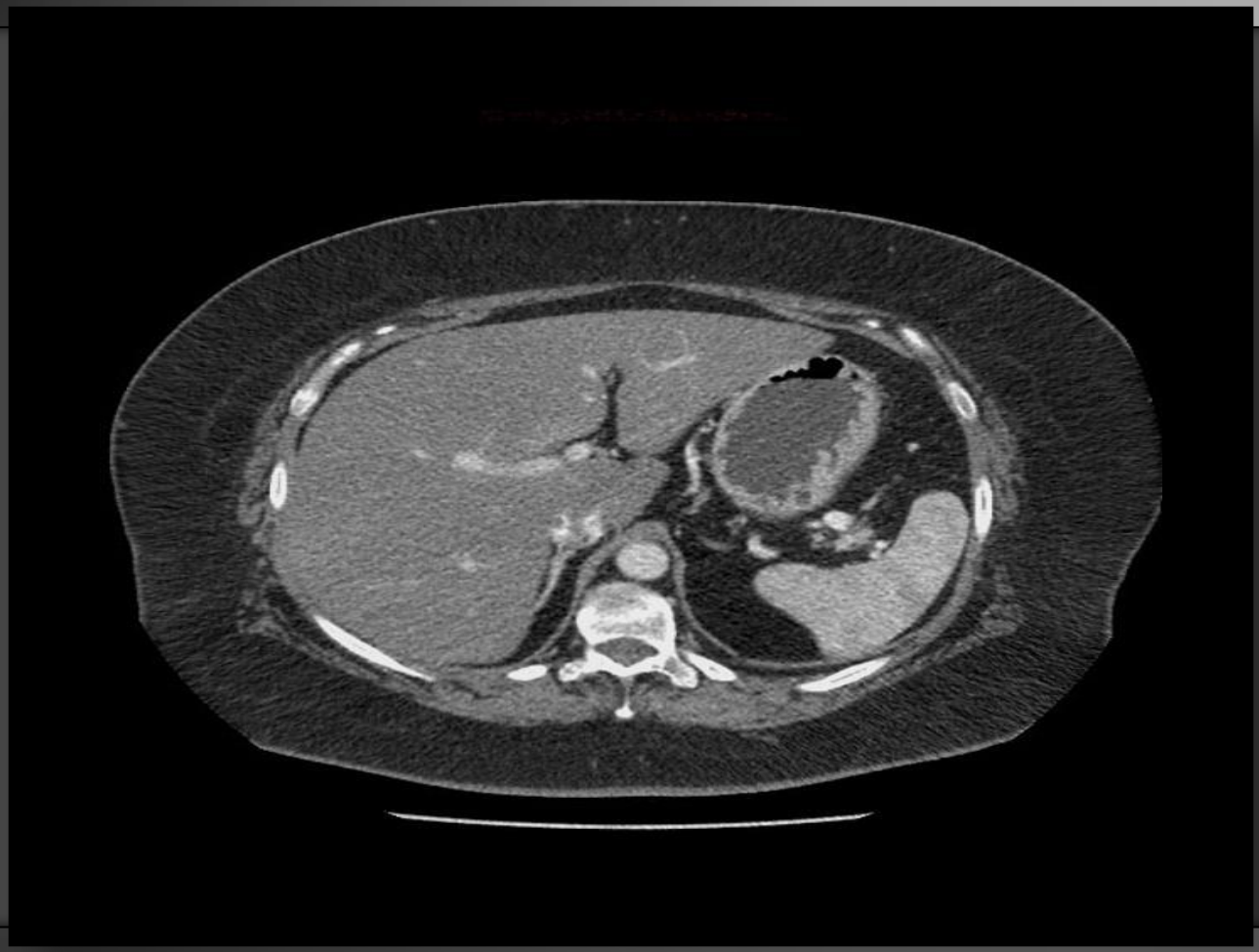
Enteropancreatic tumors

- Second most common manifestation of MEN1,
- 30% are malignant
- They tend to occur in parallel with hyperparathyroidism.
- tumors secrete peptide hormones that cause specific clinical syndromes.



MEN I with pancreatic neuroendocrine tumour

Modality CT

1. Gastrinomas

- most common enteropancreatic tumors in MEN1 patients
- result in the Zollinger-Ellison syndrome (ZES).
- ZES is caused by excessive gastrin production
- Occurs in > ½ of MEN1 pts with small carcinoid-like tumors in the duodenal wall or, less often, by pancreatic islet cell tumors.
- There may be > one gastrin-producing tumor, making localization difficult.

manifestation

- The excess acid production may cause;

1. esophagitis,
2. duodenal ulcers throughout the duodenum,
3. ulcers involving the proximal jejunum, and
4. diarrhea.

The ulcer is commonly refractory to conservative therapy such as antacids.

diagnosis

- Increased gastric acid secretion,
- elevated basal gastrin levels in the serum [generally >115 pmol/L
- an exaggerated response of serum gastrin to either secretin or calcium.

NB:Approximately one-fourth of all ZES occurs in the context of MEN1

Other causes of elevated serum gastrin levels should be excluded , such as :

1. achlorhydria,
2. treatment with H_2 receptor antagonists or proton pump inhibitors,
3. retained gastric antrum,
4. small-bowel resection,
5. gastric outlet obstruction, and
6. hypercalcemia,

2 . Insulinomas

- second most common enteropancreatic tumors in MEN1.
- Unlike gastrinomas, most insulinomas originate in the pancreas bed,
- becoming the most common pancreatic tumor in MEN1.
- The tumors may be benign or malignant (25%).

diagnosis

- hypoglycemia during a short fasting time
- inappropriate elevation of serum insulin and C-peptide levels.
- Large insulinomas may be identified by CT or MRI scanning.
- Intraoperative ultrasonography.

3 . Glucagonoma

- seen occasionally in MEN1,
- **manifestations** :

causes a syndrome of,

1. hyperglycemia,
2. skin rash (necrolytic migratory erythema),
3. anorexia,
4. glossitis,
5. anemia,
6. depression,
7. diarrhea, and
8. venous thrombosis.

- glucagon level is high.
- +/- ⬆ plasma ghrelin levels.
- The glucagonoma syndrome may represent a complex interaction between glucagon/ghrelin overproduction and the nutritional status of the patient.

4. VIPomas (1%) secrete VIP and cause profuse watery diarrhea, hypokalemia, and achlorhydria (WDHA, Verner-Morrison syndrome).

5. Somatostatinomas (0.7%) can cause diabetes mellitus, steatorrhea, and cholelithiasis.

- The pancreatic neoplasms differ from the other components of MEN1 in that approximately one-third of the tumors display malignant features, including hepatic metastases

Tx of Pancreatic Islet Cell Tumors

- Two features complicate the management.

1. the pancreatic islet cell tumors are multicentric, malignant about a third of the time, and cause death in 10–20% of patients.

2. performance of a total pancreatectomy to prevent malignancy causes diabetes mellitus, a disease with significant long-term complications that include neuropathy, retinopathy, and nephropathy.

general concepts in tx;

- (1) Islet cell tumors should be resected because medical therapy for the hormonal effects are generally ineffective.

- (2) Gastrin-producing islet cell tumors that cause ZES are frequently multicentric. Caused by duodenal wall carcinoid tumors and that resection of these tumors improves the cure rate.

- (3) In families in which there is a high incidence of malignant islet cell tumors that cause death, total pancreatectomy at an early age may be considered to prevent malignancy.

- Mgt of metastatic islet cell carcinoma is unsatisfactory.

- Hormonal abnormalities can sometimes be controlled. eg ZES can be treated with H_2 receptor antagonists or proton pump inhibitors;

- somatostatin analogues eg octreotide or lanreotide, are useful in the management of carcinoid, glucagonoma, and the watery diarrhea syndrome.

- Bilateral adrenalectomy for ectopic ACTH syndrome .

- Islet cell carcinomas frequently metastasize to the liver but may grow slowly.

- Hepatic artery embolization, radiofrequency ablation, or chemotherapy may reduce tumor mass, control symptoms of hormone excess, and prolong life; however, these treatments are never curative.