

The curious case of the recurrent 'perforator'

Aathi.S

C.Pretorius

Mackay Base Hospital

Introduction

- 51 year old ,male presents to the Mackay Base Hospital with an episode of small bowel perforation in Feb 2013 on the background of 3 episodes of hematemesis.
- Multiple admissions over subsequent months for abdominal pain. Ct in August 2013 showed pneumobilia.

- Given these findings the patient went on to undergo an upper gastrointestinal endoscopy which showed extensive ulceration of the esophagus and duodenum (3 ulcers; non-bleeding with clean base). Duodenal papilla was distorted by the degree of inflammation.
- Histology from gastroscopy (Aug 2013):
 - Gastro-esophageal junction: Extensive ulceration of esophageal mucosa; -ve malignancy; +ve for Candida
 - D2: Erosive acute duodenitis with evidence of regenerative atypia

- Multiple duodenal ulcers with clean base.

Images:



1 Lower Third of the Oesophagus



2 2nd Portion of the Duodenum



3 2nd Portion of the Duodenum



4 2nd Portion of the Duodenum



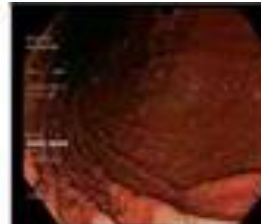
5



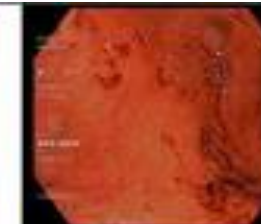
6 Upper Gastrointestinal Tract



7



8 Gastric Body



9 Gastric Body



10 Lower Third of the Oesophagus



11



12 Lower Third of the Oesophagus



13

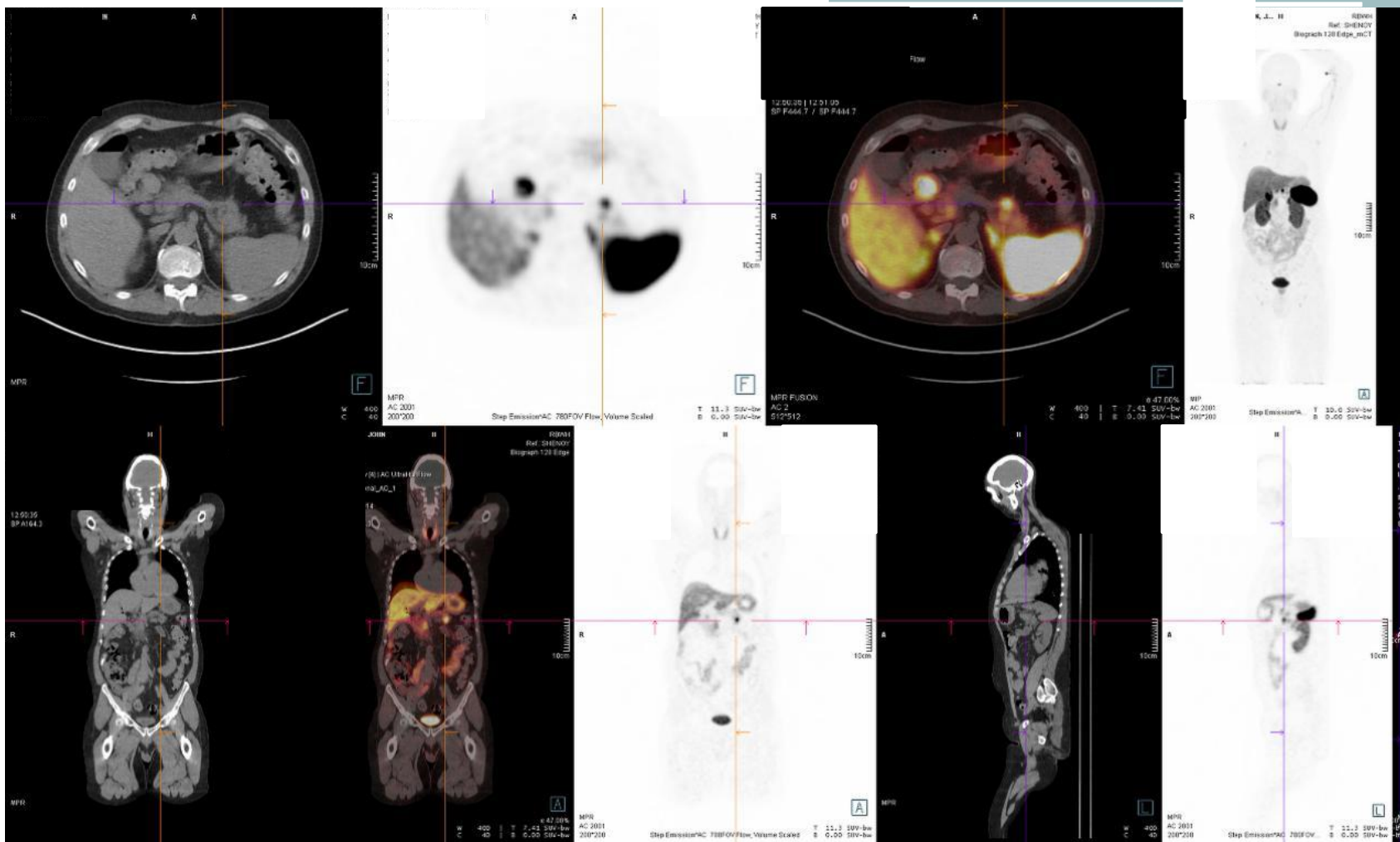
Gastroscopy images carried out on the patient in August 2013

- The patient then presented to Mackay again with a small bowel perforation on the 6th of Jan 2014 and had to undergo resection of the proximal jejunum.
- There was no evidence of Crohn's on histology. Case referred on to Gastroenterology service at referral centre whilst pt on steroids and AZT.
- Represented to MBH March 2014 with contained perforation that was drained percutaneously.

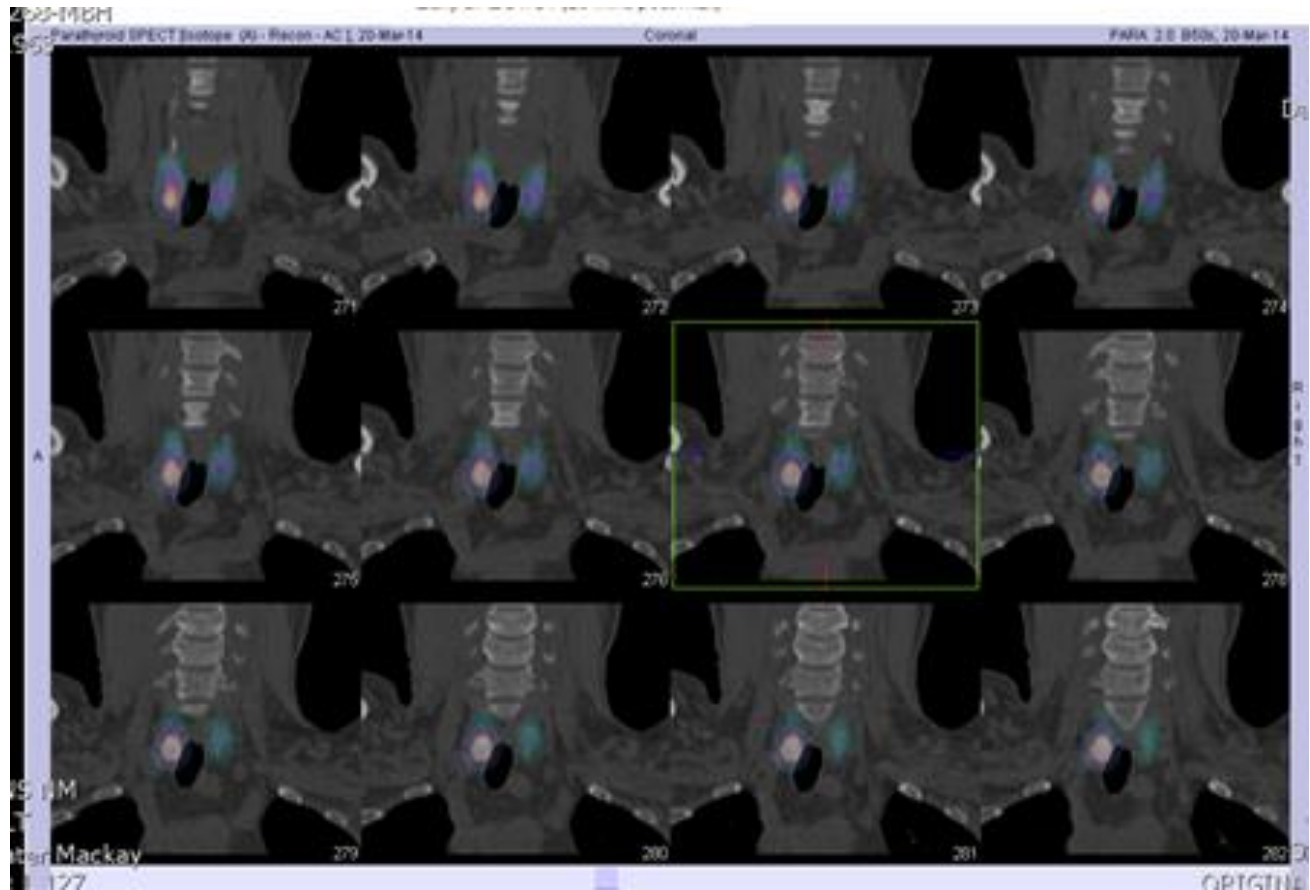
Histology from the laparotomy in Jan 2014(2nd laparotomy):

- Proximal Jejunal biopsy:
 - Heterotopic gastric mucosa
 - Ulceration and possible perforation
- Terminal ileum:
 - Serosal inflammation and fibrosis related to previous perforation
 - Benign gastrointestinal stromal tumour (GIST)

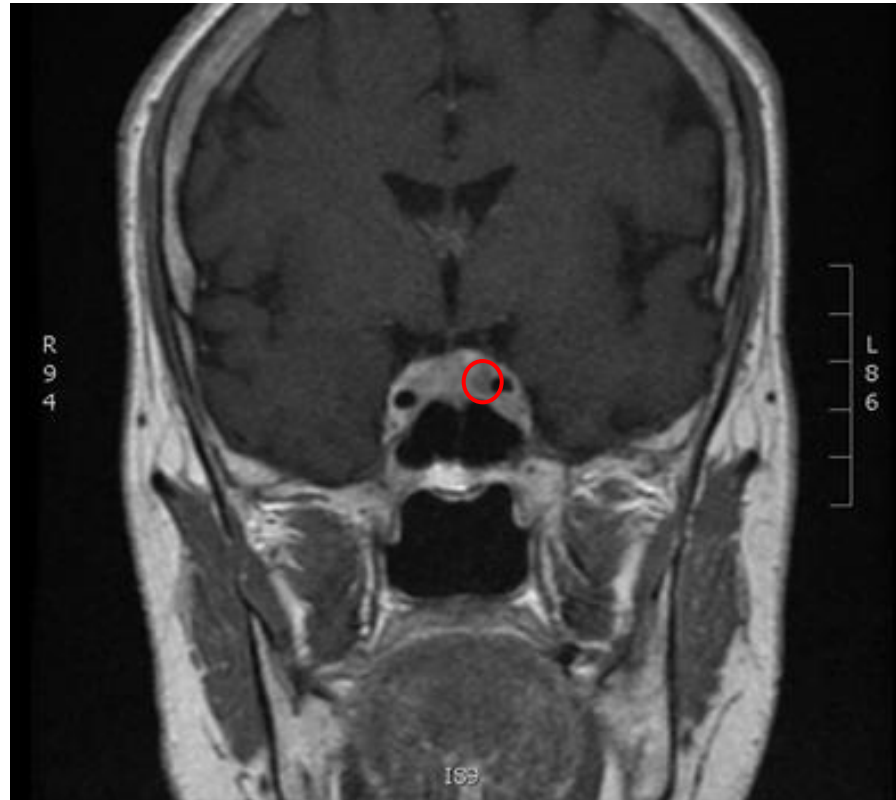
- Subsequent discussion of pt's case at radiology meeting demonstrated a subtle lesion at the tail of the pancreas.
- Pt was happy to inform (in retrospect) that his family had a high incidence MEN 1.
- Subsequent testing demonstrated :
 - Gastrin 6280 ng/L (on ppi)
 - Prolactin 11,400 mU/L
 - Parathyroid level 7.6
 - Normal TSH , T4, insulin and fasting glucose



Ga-Dotatate scan identifying lesion in body and tail of pancreas with 3cm porto-caval node also highlighted on axial scan



Sestamibi nuclear medicine scan identifying functioning parathyroid adenoma in Right lower pole



MRI brain T1 sequence identifying showing microadenoma in the posterior aspect of Left anterior pituitary

- Features on MEN 1 on this patient:
 - Prolactinoma
 - MRI: small microadenoma in post. Left pituitary measuring 5x4x4mm suggestive of micro prolactinoma
 - On Cabergoline treatment
 - Hyperparathyroidism
 - Pt underwent subtotal parathyroidectomy + thymectomy and auto-transplantation into the forearm (14/5)
 - Histology : asymmetrical hyperplasia of parathyroid tissue
 - Gastrinoma
 - 3cm porto-caval node is visible; involvement of body and tail of pancreas
 - Endoscopic US-guided FNA : gastrin secreting neuroendocrine tumour

Approach to complex MEN 1 patients

Management of parathyroid tumours

- Surgery is preferred form of treatment (sub-total parathyroidectomy+thymectomy)⁽¹⁾
- If surgery contraindicated or hyperparathyroidism reoccurs, options:
 - Octreotide and Cinacalcet
 - Ethanol ablation

Management of Zollinger Ellison Syndrome(ZES)

- Medical management first line
- Limits symptoms and complications with high dose ppi (60mg od omeprazole, 120mg od esomeprazole, 120mg od pantoprazole)⁽²⁾
- Surgery: Indications
 - Extra-pancreatic gastrinoma
 - Aggressive growth patterns
 - Exploratory laparotomy to identify lesions larger than 2cm

Management of pituitary tumours

- Prolactinoma – Dopamine agonists form the first line of treatment
- Surgery(transsphenoidal hypophysectomy) is first line treatment for other pituitary tumours and second line treatment for dopamine agonist resistant prolactinoma
- Radiation therapy appropriate after noncurative pituitary surgery (3)

Peculiar features about this case

- Pt is part of a familial type MEN (3 identified first degree family members) whose chromosomal mutation has not yet been characterised.
 - Tested normal for cytogenetic testing in 2002
- Patient presented with proximal bowel perforations ; an uncommon initial presenting feature. (4)

• References:

1. *Schreinemakers et. al.* ; The optimal surgical treatment for primary hyperparathyroidism in MEN1 patients: a systematic review. *World J Surg.* 2011 Sep;35(9)
2. *Imamura M et. al.* ; Biochemically curative surgery for gastrinoma in multiple endocrine neoplasia type 1 patients. *World J Gastroenterol.* 2011;17(10):1343
3. *Pieterman CR et. al.*; Care for patients with multiple endocrine neoplasia type 1: the current evidence base. *Fam Cancer.* 2011 Mar;10(1):157-71
4. *Waxman I et. al.*; Peptic ulcer perforation as the presentation of Zollinger-Ellison syndrome. *Dig Dis Sci.* 1991: 36: 19-24