East Kent Hospitals University
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Extra-pancreatic, Extra-intestinal Pancreatic Polypeptide secreting tumour presenting as a case of diarrhoea.

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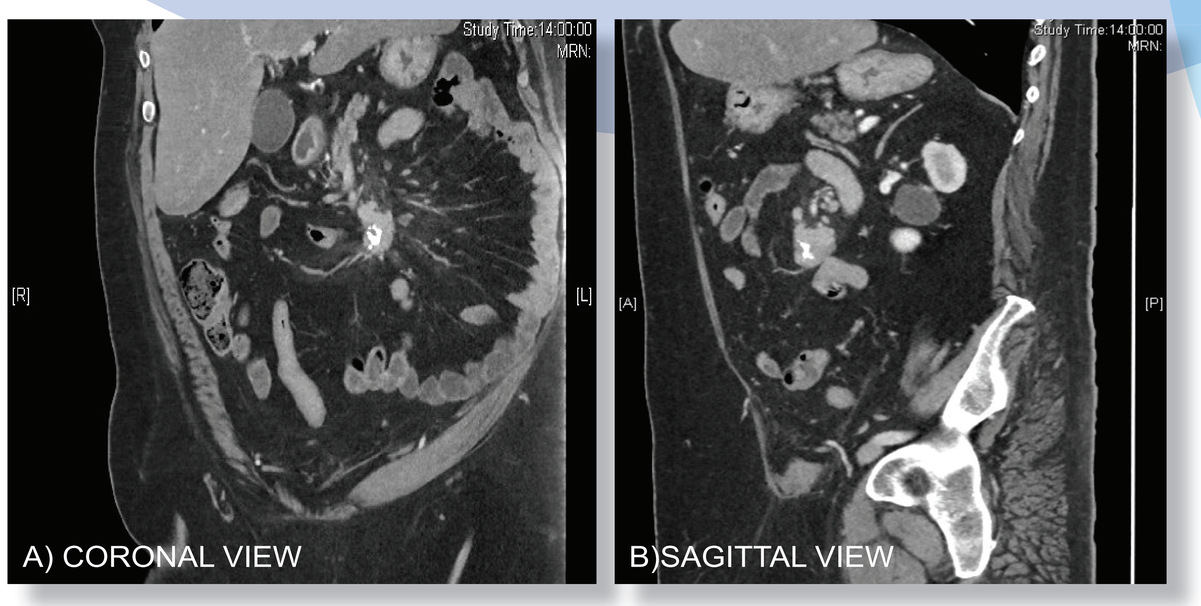
Introduction

- Pancreatic Neuroendocrine tumours, P-NETs, comprise 2-3% of all pancreatic tumours[1].
- Usually classified as functional or non-functional based on whether these secrete biologically active amines, causing specific syndromes or not. Even non-functional P-NETs in majority of cases secrete substances, not associated with any specific syndrome, like Chromogranin A and Chromogranin B and Pancreatic Polypeptide (PP)[2].
- An estimated 2% of all the neuroendocrine tumours secrete Pancreatic Polypeptide exclusively and are sometimes called PPomas[3]. This is a rare group of NETs and in almost all the reported cases, the source, usually a tumour, was originating from the pancreas.

Case History

- The case we describe is unique as the source of excess PP is neither coming from the pancreas nor the intestine.
- This 65-year old gentleman with background of type 2 DM presented with a few weeks' history of explosive diarrhoea with no flushing, pruritus or other symptoms usually associated with carcinoid syndrome.
- CT scan showed a soft tissue mass with calcifications close to the mesentery at the level of the lower poles of the kidneys.
- A full biochemical profile for work up of neuroendocrine tumour was carried out which showed raised Chromogranin A and B and PP levels with rest of profile unremarkable including Gastrin, Glucagon, Somatostatin, VIP and 24hour urine 5HIAA.
- The mass was confirmed as somatostatin avid lesion with no uptake elsewhere on the Octreotide scan.
- Patient was referred to the neuroendocrine oncologist for further management who commenced Sandostatin LAR.

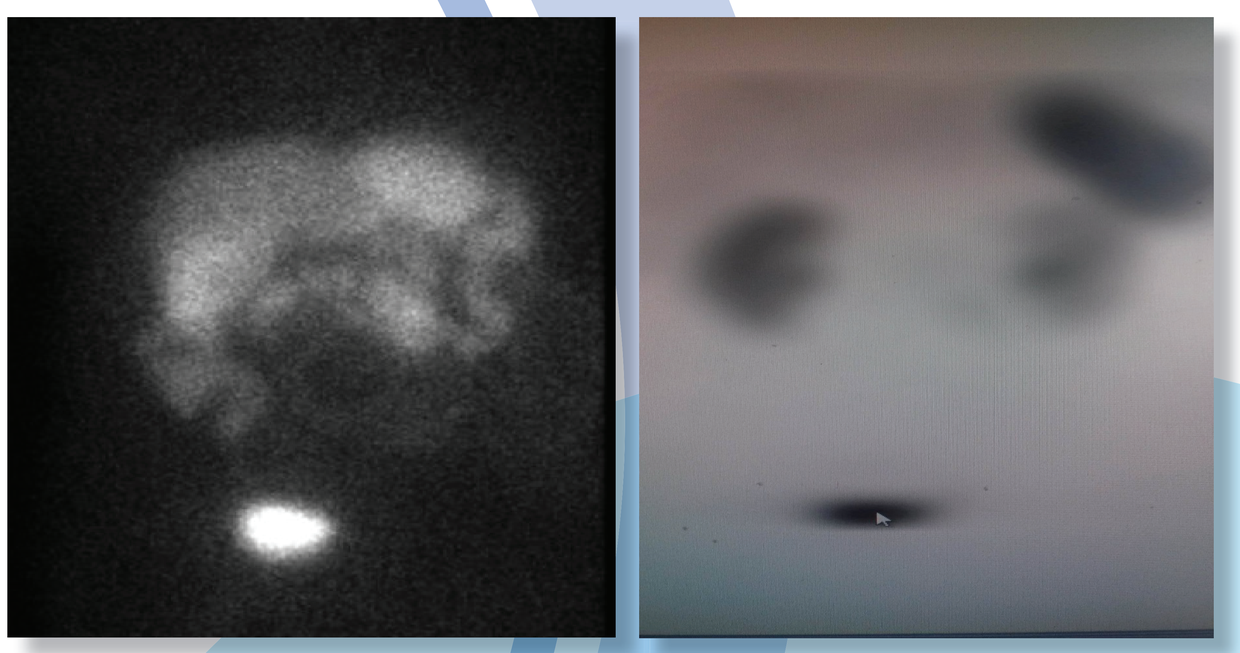
CT Scan: CT scan showing soft tissue mass separate from pancreas and bowel loops



Lab Results

TEST (Normal Range)		RESULT
Chromogranin A	(0-60 pmol/l)	364
Chromogranin B	(0-150 pmol/l)	282
Pancreatic Polypeptide	(0-300 pmol/l)	>500
Gastrin	(0-40 pmol/l)	6
Glucagon	(0-50 pmol/l)	25
Somatostatin	(0-150 pmol/l)	51
VIP	(0-30 pmol/l)	10
Urine 5HIAA	(0-42 umol/24hour)	1

Octreotide Scan: Somatostatin Avid lesion with no uptake elsewhere



Conclusion and recommendation

- A thorough literature search has not shown previously reported extra-pancreatic PPoma causing diarrhoea and no other symptoms.
- Usually PPomas are considered silent but we believe this is no longer true and can arise from outside pancreas.
- Further research however, is warranted into their origin which we now believe can even be extra-pancreatic and extra-intestinal.

References

1. Rizvi SM, Wong J, Saif MW, et al. Pharmacogenetics in neuroendocrine tumors of the pancreas. JOP. 2014 Jul 28;15(4):299-30
2. Granberg D. Biochemical Testing in Patients with Neuroendocrine Tumors. Front Horm Res. 2015;44:24-39.
3. Kasper D, Longo D, Fauci A, Hauser S, Jameson J et al. Harrison's principles of internal Medicine, 19e. New York, NY: McGraw-Hill; 2015.

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