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.
- 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).
- An estimated 2% of all the neuroendocrine tumours secrete Pancreatic Polypeptide exclusively and are sometimes called PPomas. 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.

Lab Results

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

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


Octreotide Scan: Somatostatin Avid lesion with no uptake elsewhere

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

A) CORONAL VIEW

B) SAGITTAL VIEW