Phaeochromocytoma- but where?

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EP48: Case Report- NET

Initial presentation
• M/53, intermittent retrosternal pain and dysphagia
• Gastroscopy (21/01/2014): 2 cm soft sub-pedunculated polypoid mass in lower oesophagus, biopsy: adenocarcinoma
• CT TAP: Distal oesophageal lesion occupying entire lumen, no lymphadenopathy, 20mm right adrenal ‘incidentaloma’ with mild calcification and reassuring imaging characteristics
• EUS T2N0 oesophageal mass
• 18FDG PET CT: High-grade metabolically active lesion in distal oesophagus (SUV-max 9.5); 2.7 cm nodule in right adrenal with some calcification, HU 30 (higher than lipid rich adenoma), intermediate grade increased activity (SUV-max 4.2), unlikely to be a metastasis.
• Ivor Lewis oesophagectomy and mesh repair of incisural hernia a year later without any endocrine assessment.
• Histologically: moderately differentiated adenocarcinoma, pT1b, pN0, R0, V0.

Two years after:
• CT scan for post thoracotomy pain in right side of chest: right adrenal lesion increased slightly in size, appeared necrotic- metastasis from his oesophageal carcinoma or a primary adrenal lesion.

Referred to endocrine team- asymptomatic besides slightly reduced effort tolerance.
• 24 hour urinary free noradrenaline on three separate days were 1318, 1755 and 837 nmol/24 hr (ref: 82-650) with normal adrenaline and dopamine.
• MIBG: Intense uptake within the gastric pull-up, normal uptake in the adrenal glands, no obvious neuroendocrine tumour elsewhere.

Biochemical investigations to confirm catecholamine hypersecretion

<table>
<thead>
<tr>
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<th>Day 1</th>
<th>Day 2</th>
<th>Day 3</th>
<th>Reference range</th>
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<tbody>
<tr>
<td>Plasma metanephrines</td>
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<tr>
<td>Normetanephrine (pmol/L)</td>
<td>2657</td>
<td>3298</td>
<td>3037</td>
<td>120-1180</td>
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<tr>
<td>Metanephrine (pmol/L)</td>
<td>301</td>
<td>333</td>
<td>293</td>
<td>80-510</td>
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<tr>
<td>3-methoxytyramine (pmol/L)</td>
<td>&lt; 180</td>
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<tr>
<td>24hr urinary catecholamines</td>
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<tr>
<td>Free noradrenaline (nmol/col)</td>
<td>1478</td>
<td>2016</td>
<td>-</td>
<td>82-650</td>
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<td>Total normetadrenaline (pmol/col)</td>
<td>8.6</td>
<td>9.2</td>
<td>-</td>
<td>0-4.9</td>
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</tbody>
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Plasma normetanephrine (pmol/L)
• Sitting 3298
• After 30 minute supine rest 3037

• 68Ga-DOTATATE scan: Intense DOTATATE uptake in right adrenal nodule (2.8x2.1cm), higher than expected physiological activity (SUVmax 30, cf left adrenal with normal configuration had SUVmax 16), no DOTATATE avid disease elsewhere, 12mm nodule in right lung oblique fissure.

• Right adrenalectomy was performed after appropriate alpha and beta blockade.
• Histology confirmed phaeochromocytoma (immunohistochemistry positive with chromogranin and synaptophysin) with PASS (Phaeochromocytoma of the Adrenal gland Scoring Scale) score of two.

Discussion:
This case reminds us of certain interesting points:
• Although most adrenal phaeochromocytomas secrete both norepinephrine and epinephrine, about a third exclusively produce norepinephrine and a much smaller proportion exclusively produce epinephrine [1].
• DOTATATE scan is generally found to be more sensitive and specific than MIBG scan for diagnosis of NETs [2]. In a recent meta-analysis of 14 studies, change in management occurred in 44% (range: 16-71%) of NET patients after SSTR PET/CT. In 4/14 studies SSTR PET/CT was performed after an ¹¹¹In-Octreotide scan. In this subgroup additional information by SSTR PET/CT led to a change in management in 39% (range: 16-71%) of patients [3].
• There is some association in literature linking false negative MIBG scans with SDHB mutations, high frequency to develop metastatic disease, extra-adrenal location and hypersecretion of normetanephrine or norepinephrine [4].
• Osophageal uptake in MIBG scan proved to be a red herring. Red herrings in isotope scans are so common that they are no longer red!

References: