

Phaeochromocytoma- but where?

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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 incisional 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.

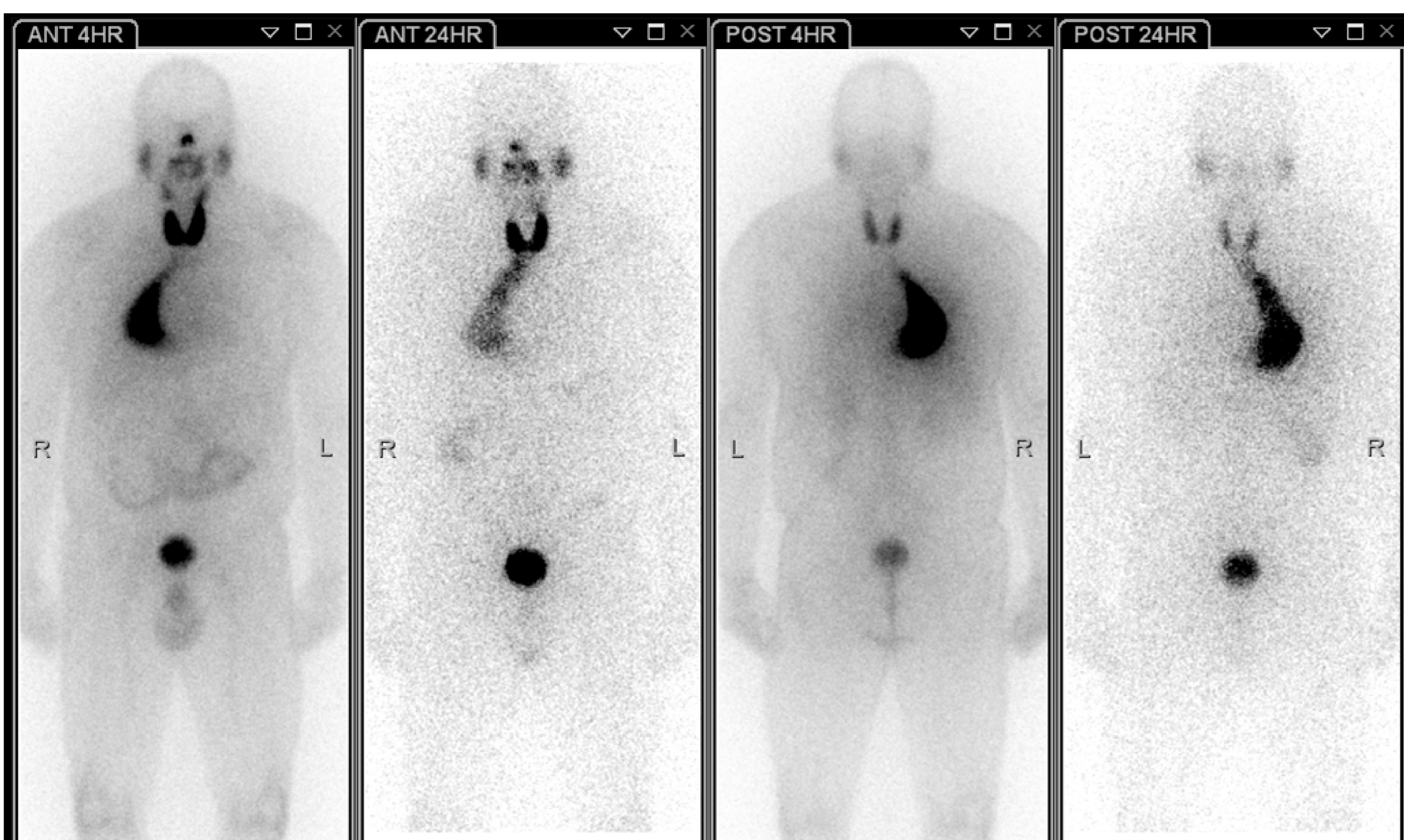


Initial



Two years later

- Referred to endocrine team- asymptomatic besides slightly reduced effort tolerance.
- 24 hour urinary free noradrenalin 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



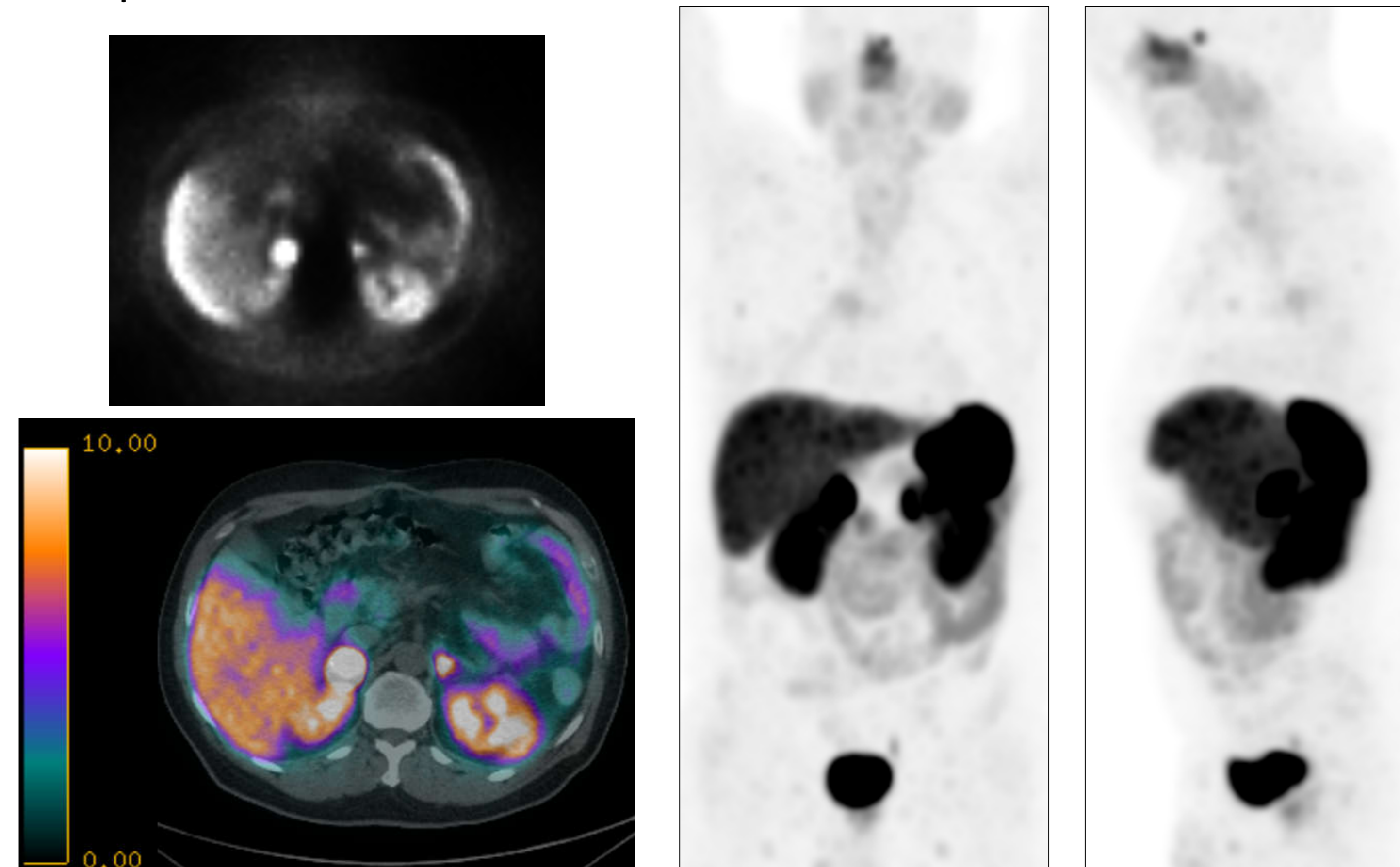
References:

1. Kantorovich V, Pacak K. Pheochromocytoma and paraganglioma. Progress in brain research. 2010;182:343-373.
2. Mojtahedi A et al. The value of 68Ga-DOTATATE PET/CT in diagnosis and management of neuroendocrine tumors compared to current FDA approved imaging modalities: a review of literature. American Journal of Nuclear Medicine and Molecular Imaging. 2014;4(5):426-434.
3. Barrio M et al. The Impact of Somatostatin Receptor-Directed PET/CT on the Management of Patients with Neuroendocrine Tumor: A Systematic Review and Meta-Analysis. J Nucl Med. 2017 May;58(5):756-761.
4. Fonte JS et al. False-negative ¹²³I-MIBG SPECT is most commonly found in SDHB-related pheochromocytoma or paraganglioma with high frequency to develop metastatic disease. Endocr Relat Cancer. 2012 Feb 13;19(1):83-93.

Biochemical investigations to confirm catecholamine hypersecretion

	Day 1	Day 2	Day 3	Reference range
Plasma metanephrines				
Normetanephrine (pmol/L)	2657	3298	3037	120- 1180
Metanephrine (pmol/L)	301	333	293	80- 510
3-methoxytyramine (pmol/L)	< 180	< 180	< 180	< 180
24hr urinary catecholamines				
Free noradrenaline (nmol/col)	1478	2016	-	82- 650
Total normetadrenaline (µmol/col)	8.6	9.2	-	0- 4.9
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].
- Oesophageal uptake in MIBG scan proved to be a red herring. Red herrings in isotope scans are so common that they are no longer red!

I am here to distract you

