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Title: A Case of Pheochromocytoma and Lung Carcinoid

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A 62yr old lady first presented to a District General Hospital in 1997 with confusion on background of hypertension. She was diagnosed as having transient ischaemic attack. Her blood pressure was controlled on Doxazocin. Following a differential diagnosis of Pheochromocytoma by the House Officer, she had 24-hr urinary catecholamines which confirmed the diagnosis - Adrenalin 240(7-82), Noradrenalin 14945(76-561), Dopamine 16280(366-2897) nmol.

MRI scan abdomen showed a 4cm mass between IVC and portal vein and 1.8x1.5cm lesion in left adrenal. There were also 3 lesions seen in the liver, later thought to be haemangiomas on USS and MIBG. Histology of the extra-adrenal tumour confirmed the diagnosis as "Pheochromocytoma but malignancy cannot be excluded". She was followed up until 2001 with serial UFC, and then was "lost" to follow up for 5yrs.

In 2006, she presented back to hospital with "collapse" and febrile illness. CXR and CT thorax showed a left hilar mass. Repeat 24-hr urinary catecholamines and MIBG were negative. Histology from bronchoscopy samples was in keeping with diagnosis of Carcinoid; however 5-HIAA levels were normal. Apart from history of long-term profuse and troublesome diarrhoea, she had no other symptoms to suggest Carcinoid syndrome.

Octreotide scan showed increased focus of metabolic activity within left lung and a 3.8cm mass. There was also a focal activity within the pituitary.

The tumour was excised and post-operative levels of 5-HIAA have remained normal. The diarrhoea persists several months post-operation. Anterior pituitary hormones, serum calcium and 24-hr UFC are all within normal limits. Ophthalmoscopy did not show retinal angiomas. Recently genetic screen for VHL gene mutation was negative whilst results of other genes are awaited. Gut hormones screen was normal.

Our original working diagnosis was either VHL or MEN 2; however after investigations this is unlikely.

Cause of diarrhoea in this lady is still unknown. Could it be still MEN syndrome?

MRI scan of her pituitary is awaited.