A case of Phaeochromocytoma co-secreting ACTH as a rare cause of Cushing syndrome

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Case History:

A 49 year lady was admitted with sudden onset headache secondary to severe hypertension. CT brain and LP were normal. Her urinary catecholamines were found to be elevated. Her hypertension did not settle with alpha/beta blockade and further to this she became clinically Cushingoid and was found to have hypercortisolaemia.

Investigations and method:

Urine catecholamines, CT abdomen, MIBG scan, overnight dexamethasone suppression test (ONDST), 24 hour urine free cortisol (UFC), plasma ACTH level

Results and treatment:

Noradrenaline 709 and 972 nmol/mmol of creatinine (ref range 0-48), Adrenaline 215 and 256 (ref range 0-10). CT abdomen - 4.2 cm left adrenal mass, MIBG scan – confirms phaeochromocytoma. Cortisol following ONDST >1750 nmol/L; 24hr UFC > 1750 nmol/L; ACTH 555 ng/L

Initial treatment involved alpha and beta blockade with dose titration, reaching up to phenoxybenzamine 70mg tds and propranolol LA 160mg. Despite this she remained haemodynamically unstable and clinically started to become Cushingoid. Investigations for cortisol excess revealed ectopic ACTH secretion. Metyrapone was added to her treatment and she was transferred to tertiary centre for adrenalectomy. Her pre-op course was complicated by admission to ITU for suspected respiratory failure due to swine flu associated pneumonia and small PE. After recovery, she successfully underwent laparoscopic left adrenalectomy. Subsequent biochemical testing and imaging confirms complete resolution of catecholamine and cortisol excess. Screening for MEN was negative.

Conclusions and points for discussion:

The diagnosis is left adrenal Phaeochromocytoma cosecreting ACTH. This case is unusual in its acute presentation with highly metabolically active phaeochromocytoma which has probably masked the symptoms and signs of cortisol excess until active blockade of catecholamine action.