A challenging case of Cushing's

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Case history

48 year old patient with 4 months of progressive lower limb weakness and weight gain of 12 kg. Previous history included hypertension and osteoarthritis. On examination she was clinically Cushingoid and hypertensive (170/90) with evidence of proximal myopathy.

Investigations

Potassium was low (2.2 mmol/L). Urinary free cortisol elevated at 7703 nmol/24h with a basal 0900h cortisol and ACTH of 1167mmol/L and 507 ng/L respectively. Cortisol did not suppress after formal low dose dexamethasone testing. MRI: normal pituitary. CT chest/abdomen/pelvis: metastases in the liver with a mass in the pancreas. Octreotide scanning: uptake in the liver only. Echocardiography: reduced ejection fraction (35%). Liver biopsy histology confirmed co-secretion of ACTH and gastrin from a low grade neuroendocrine tumour (NET)

Results and treatment

Commenced on Spironolactone 100mg OD with Metyrapone (increasing to 1g t.d.s) and Ketoconazole (200mg t.d.s.) introduced to control her hypercortisolism. Referred for emergency bilateral adrenalectomy as very symptomatic. She has subsequently been commenced on dexamethasone (1mg and 0.5mg) and Sandostatin LAR 30mg. Interval echocardiographic monitoring has demonstrated improvement.

Conclusions

This is a challenging case of ectopic ACTH secretion from a probable pancreatic NET. Coexisting Cushing’s cardiomyopathy further complicates management.
On-going treatment includes somatostatin analogue therapy, consideration of hepatic embolization of the liver metastases and further surgery