# Reversal of dilated cardiomyopathy in a patient with Cushing's syndrome after a successful adrenalectomy

*N Abbas, J Chambers, J Powrie*
*Department of Endocrinology, Department of cardiology*
*Guys and St Thomas' NHS Foundation Trust, London*

## Introduction

Cushing's syndrome (CS) associated with dilated cardiomyopathy without LVH is rare but important to recognise as treatment of CS can lead to total recovery of heart function.

## Case History

A 30 year old previously fit & well Turkish man presented with chest pain and was diagnosed with NSTEMI & CCF.

An MRI of the heart and CT coronary angiogram showed normal coronary arteries but a **large right adrenal tumour of 11.5cm with extension into IVC**.

The transthoracic echocardiogram showed a globally dilated left ventricle with an estimated ejection fraction 20%. There was moderate to severe functional mitral regurgitation.
Clinically the patient had gross clinical CS and was hypertensive (BP-170/90).

## Investigations & Management

Further investigations revealed

**Na-140, K-3.3**
**cortisol after a 1mg ODST; 509 nmol/L**
**24H urinary cortisol - 4026 nmol/day (normal range 100-379nmol/day)**
**ACTH<5 ng/L**
Aldosterone, renin, urinary metanephrines, DHEA and androstendione all normal.

His heart failure was managed with optimal medical treatment and metyrapone was started. Right adrenalectomy with extraction of intracaval tumour thrombus was performed with evidence of complete excision both on histological assessment and a post operative CT scan. Histology confirmed adrenal cortical carcinoma. Weiss score 5.

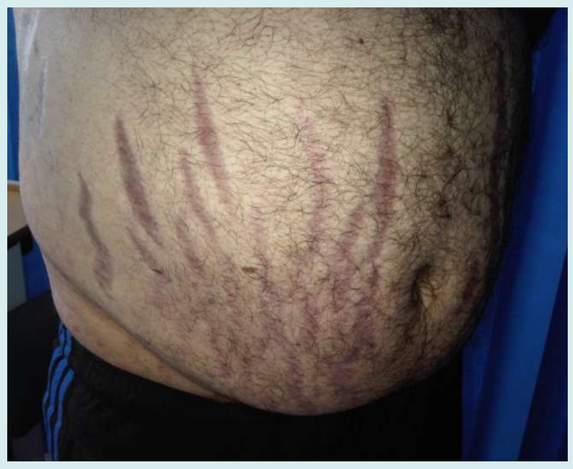
**1 year post surgery, clinical features of Cushing's have almost resolved. Echocardiogram 7 months after adrenalectomy showed a reduction in LV size and an EF of 40-45%. A recent echocardiogram showed EF 56%.**

## Discussion

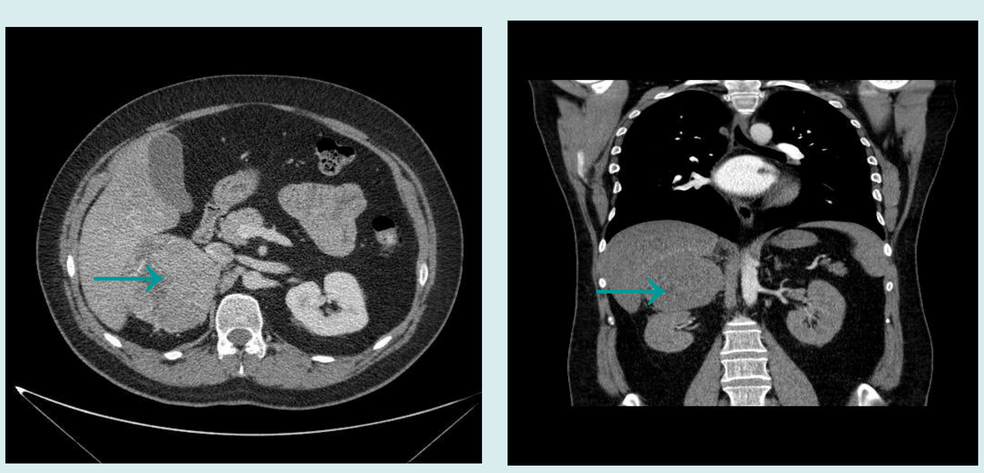
**Studies examining the relationship between hypercortisolism and cardiac dysfunction suggest that excess cortisol is contributory to cardiac re-modelling and dilated cardiomyopathy, independent of hypertension.**

**The pathophysiology of cardiac remodeling involves complex mechanisms including activation of neurohormonal factors, alpha adrenergic and renin-angiotensin-aldosterone systems.**

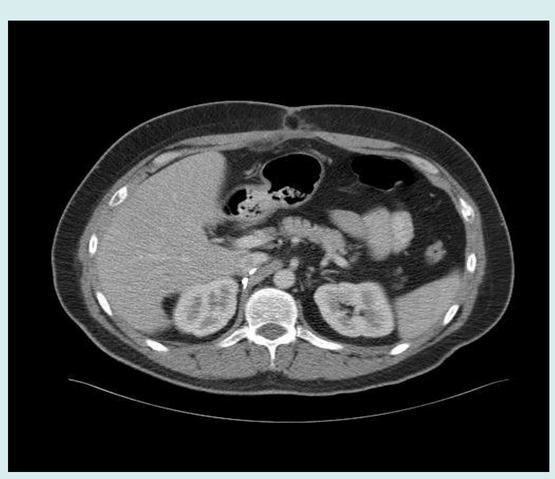
**Experimental models have found that the effects of noradrenaline, angiotensin II, and aldosterone can be heightened by hypercortisolism. The saturation of 11β-HSD2 enzyme resulting in mineralocorticoid receptor activation by cortisol has also been suggested as a possible reason for cardiomyopathy in CS.**



**Central Obesity, Abdominal Striae**



**Large right adrenal corticocarcinoma**



**Post operative scan shows complete resection with no residual tumour tissue**

| Investigation | 17/11/2011 | 24/11/2011 post op results | Normal Values |
|---|---|---|---|
| **18:00hours Cortisol** | **634** | 26 | **64 – 327 nmol/L (4-8 PM)** |
| **11 Deoxycortisol** | **241.1** | 5.2 | **0 – 20.9 nmol/L** |
| ACTH | < 5.0 | 7.0 | < 46 ng/L |
| **Na** | **140** | 138 | 135 -145 mmol/L |
| **K** | **3.3** | 4.4 | 3.5 - 5.0 mmol/L |
| **Androstenedione** | **> 35.0** | < 1.05 | 2.1 – 10.8 nmol/L |
| Dehydroepiandrosterone | 9.6 | 0.6 | 4.3 – 12.2 umol/L |
| 17 Hydroxyprogesterone | 10.0 | 3.6 | nmol/L |
| Testosterone | 8.4 | 0.5 | 9.9 – 27.8 nmol/L |
| Renin | 1.2 | <0.5 | 5.4 – 60 mU/L |
| Aldosterone | 98 | 54 | 100 - 800 pmol/L |
| Metadrenaline | 0.30 | | <0.7 nmol/L |
| Normetadrenaline | 0.76 | | < 1.3 nmol/L |

**Investigation results**

## Histology Report

Tumour composed of eosinophilic cells with large, often pleomorphic nuclei possessing nucleoli.
Diffuse growth pattern and extensive necrosis is identified within tumour.
The tumour appears to be restricted within its capsule.
Vascular invasion into capsular vessels is noted and the tumour also appears to be filling a large vein.

**Scoring:**
Weiss criteria: score of 5
Modified Weiss criteria: score of 3
(both of which are suggestive of malignant behaviour).