A complicated case of Cushing's Syndrome

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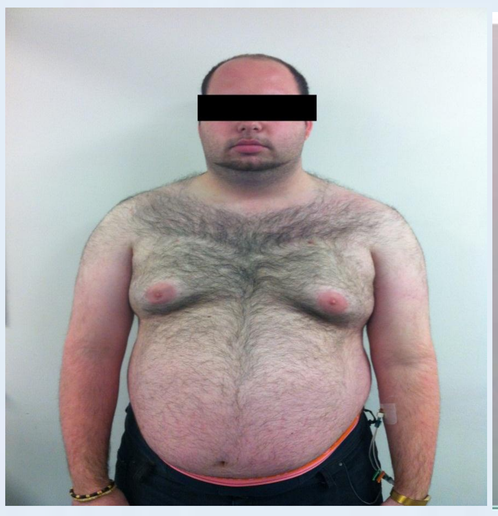
Introduction

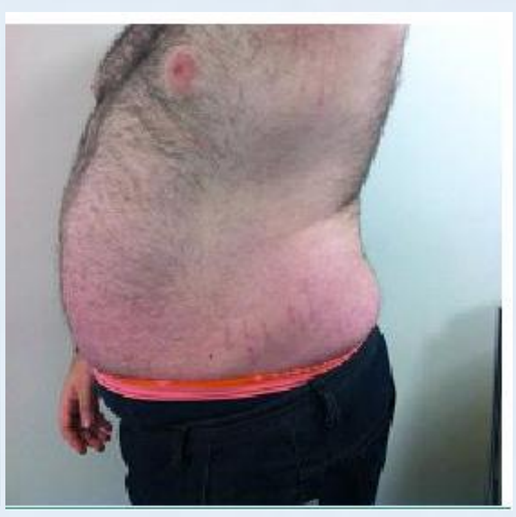
Cushing's syndrome is broadly categorized into ACTH dependent (Pituitary & ectopic source) and ACTH independent (adrenal source). Localizing the source can be a challenging process[1].

Dilated Cardiomyopathy, a rare complication of Cushing's syndrome adds to the difficulty in investigating and managing patients.

Clinical Examination

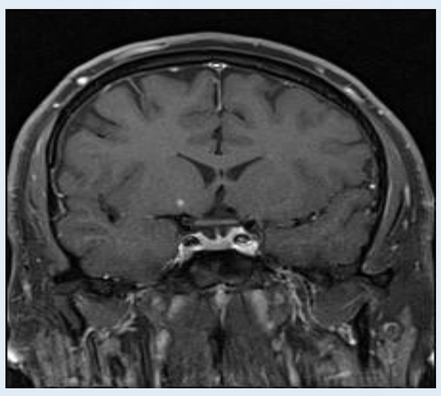
Florid Cushing's appearance:
Round face, Depressed, Central obesity,
Gynaecomastia, Purple striae,
Proximal Myopathy
Signs of heart failure

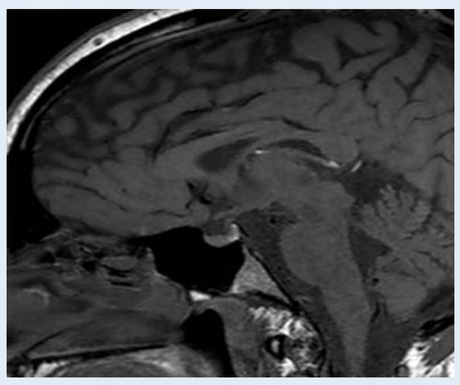




Investigations

Tests	Results 1 North Middlesex Hospital	Results 2 North Middlesex Hospital	Results 3 UCH
9 am cortisol (nmol/l)	328	428	308
ACTH (ng/l)	<5	<5	9.0
24 hrs Urine Cortisol (nmol/24 hrs)	374	354	
LDDST (nmol/l)	28	<28	





CT Adrenal: No focal lesion
Gallium Dotatate scan :No Avid uptake
Echocardiogram : EF 23%
Pituitary MRI x 2 : No focal lesion

Case Summary

25 year old teacher of Greek dancing.
Medications: none
F/H: nil
Non smoker, No illicit drugs
Alcohol: on occasions
Exogenous steroid: none
Presented with 6 months of : Dyspnoea, weight gain, muscle weakness

IPSS

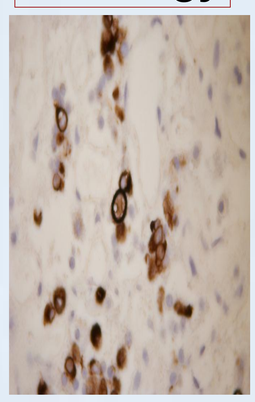
Bilateral Inferior Petrosal Sinus Sampling with CRF stimulation

Time	Ratios	ACTH (ng/l) Peripheral	Right	Left
Basal	**L/P** 1.33 **R/P** 1.01	9.6	9.9	12.8
Basal	**L/P** 11.08 **R/P** 0.92	10.1	9.4	112.0
3 min post CRH	**L/P** 107.4 **R/P** 1.33	10.4	13.9	1117
08 min post CRH	**L/P** 83.2 **R/P** 0.98	27.1	26.7	2250.0
15 min post CRH	**L/P** 10.97 **R/P** 1.40	28.6	28.0	314.0

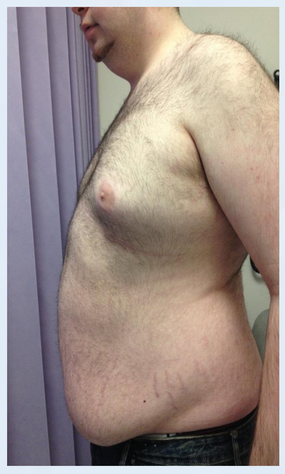
Management & outcome

- Treatment: 2 transsphenoidal surgeries.
- Histology: Crooke's Hyaline changes but no adenoma.
- Urinary free cortisol after second surgery < 28 nmol/l/24 hrs.
- Marked clinical improvement:15 kg weight loss, improved effort of breathing and EF of 41%.
- On replacement with Hydrocortisone, Thyroxine and Testosterone.

Histology



Tests	Baseline	Post 1st surgery 07/12/12	Post 2nd surgery 14/12/12
Cortisol (nmol/l)	378	510	27
Testosterone (nmol/l)	12.5	5.5	<0.4
FSH IU/L	3.8	2.6	2.0
LH (IU/L)	4.7	2.9	2.3
Prolactin (miu/l)	179	52	<10
T4 (pmol/l)	15.8	11.1	11.4
TSH (miu/l)	0.6	0.51	0.51



Conclusions

Diagnosing Cushing's syndrome and identifying the source can be challenging. Dilated Cardiomyopathy is a rare complication which adds to the diagnostic conundrum in the management of Cushing's syndrome. Treatment can reverse the many of the cardiac manifestations of hypocortisolism [2].

References [1]Meier CA, Biller BM. Clinical and biochemical evaluation of Cushing's syndrome. *Endocrinol Metab Clin North Am*. 1997;26:741–62

[2] Marazuela M, Aguilar-Torres R, Benedicto A, Gómez-Pan. A Dilated cardiomyopathy as a presenting feature of Cushing's syndrome. *Int J Cardiol.* 2003 Apr;88(2-3):331-3