New onset Sarcoidosis following treatment of Cushing's Disease

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Introduction

Cushing's Disease (CD) is caused by pituitary adrenocorticotropic hormone (ACTH) secreting tumours. These tumours are almost always benign in nature and are usually microadenomas by classification with the disease characterised by excessive elevation of glucocorticoid concentrations. [1]

In rare cases the treatment of Cushing's Disease, with a subsequent reduction in glucocorticoid concentration, may result in the unveiling or aggravaion of a disease responsive to glucocorticoid therapy. [2][3][4]

We describe a case were a patient was diagnosed with new onset Sarcoidosis following treatment of Cushing's Disease.

Case

- A 47-year-old woman with a past medical history of resistant hypertension and uncontrolled type 2 diabetes was reviewed in our outpatient clinic.
- The patient complained of a 5 year history of significant weight gain with associated oligomenorrhoea, proximal muscle weakness and depression.
- On examination the patient had a strikingly Cushingoid appearance and so underwent evaluation for Cushing's Disease.

Investigations, management and follow up

A diagnosis of Cushing's Disease was made and the patient underwent an uncomplicated left sided transphenoidal hemi-hypophysectomy. The results of the initial biochemical, radiological and subsequent pathology investigations can be viewed in table 1. On postoperative day three the patient's serum cortisol was < 30 nmol/l and therefore replacement hydrocortisone was commenced. Despite an initial tranquil postoperative convalescence the patient was frequently admitted and readmitted to hospital with a systemic illness of unclear aetiology.

Over these hospital admissions the patient complained of progressive myalgia, arthralgia, lethargy and shortness of breath but with no clear evidence of sepsis or overt hypocortisolism. However, the patient was felt to improve clinically with intravenous glucocorticoids and empirical antibiotic therapy. During one of the hospital admissions a CT Thorax was repeated and this showed new bilateral mediastinal and hilar lymphadenopathy. The patient's serum Angiotensin Converting Enzyme (ACE) level was found to be elevated, 107 IU/L, and epitheloid granulomas in keeping with Sarcoidosis were demonstrated on eventual hilar biopsy. The patients clinical condition has since spontaneously improved: her respiratory function tests and chest radiograph showed no evidence of disease profession and the serum ACE had fallen without the need for high dose corticosteroids.

Investigations	Result 1	Result 2
24-Hour Urine Free Cortisol (UFC)	540 nmols/24h	624 nmols/24h
Overnight Dexamethasone Suppression Test (ODST)	671 nmol/L	790 nmol/L
Low Dose Dexamethasone Suppression Test (LDDST)	125 nmol/L	N/A
High Dose Dexamethasone Suppression Test (HDDST)	43 nmol/L	N/A
Morning ACTH concentration	7 mU/L	5 mU/L
Bilateral Inferior Petrosal Sinus Sampling	Failed	Failed
CT Thorax, Abdomen and Pelvis with contrast	No ectopic source for ACTH secretion or other abnormality found	
3T MRI Head	Left sided 9 x 6 x 6 mm inferoposterior pituitary adenoma	
Pituitary Pathology	Pituitary microadenoma with positive immunostaining for ACTH	

Table 1: Investigations for CD

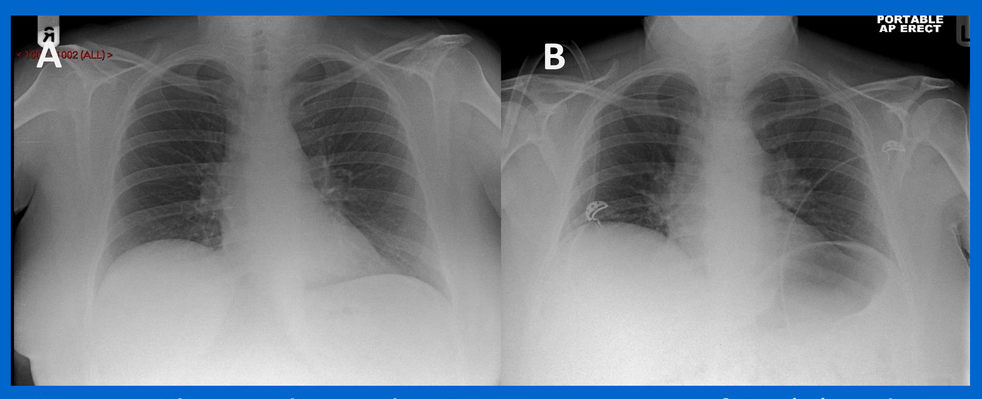

Figure 1: Chest radiograph prior to treatment of CD (A) and afterward (B)

Discussion

- Glucocorticoid responsive illnesses can be unmasked or aggravated following treatment of Cushing's Disease and it is thought that the rebound immunity that occurs plays a significant role. [5]
- Cases reports have described exacerbation and onset of inflammatory arthropathies, vasculitides, thyroid and granulomatous disease. [2] [4]
- Sarcoidosis has been reported as a rare sequelae following treatment of CD but usually presents with skin manifestations such as erythema nodosum. [2]
- This case is unusual in that dermatological signs were absent and that only a generalised systemic illness with hilar and mediastinal lymphadenopathy was seen.

Conclusion

- The unveiling of a glucocorticoid responsive disease post treatment for Cushing's Disease is rare.
- The convalescent period following treatment for Cushing's Disease can be stormy and consideration must occasionally be given to unusual diagnoses.

References

1. Nieman L, Biller B, Findling J, Newell-Price J, Savage M, Stewart P et al. The Diagnosis of Cushing's Syndrome: An Endocrine Society Clinical Practice Guideline. The Journal of Clinical Endocrinology & Metabolism. 2008;93(5):1526-1540.
2. Schaefer S, Meyer S, Brueck C, Weber M, Luedecke D, Wagner H et al. Sarcoidosis Following Cushing's Syndrome: A Report of Two Cases and Review of theLiterature. Experimental and Clinical Endocrinology & Diabetes. 2010;118(03):147-150.
3. Maldonado, MD M, Orlander, MD P, Roshan, MD S, McCutcheon, MD I, Cleary, MD K, Friend, MD K. SARCOIDOSIS AFTER SURGICALLY INDUCED REMISSION OF CUSHING'S DISEASE. Endocrine Practice. 1999;5(1):43-45.
4. Selek A, Baris S, Cetinaslan B, Canturk Z, Tarkun I, Akyay Z. New-Onset Sarcoidosis After Remission of Cushing's Syndrome. Turkish Thoracic Journal/Türk Toraks Dergisi. 2015.
5. Da Mota F, Murray C, Ezzat S. Overt Immune Dysfunction after Cushing's Syndrome Remission: A Consecutive Case Series and Review of the Literature. The Journal of Clinical Endocrinology & Metabolism. 2011;96(10):E1670-E1674.

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