

Isolated central hypoadrenalism as the sole manifestation of presumed neurosarcoidosis



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Introduction:

- Hypothalamic-pituitary sarcoidosis is uncommon and affects less than 10% of patients with neurosarcoidosis.
- It presents a diagnostic challenge.
- We present a case of isolated central hypoadrenalism as the sole manifestation of neurosarcoidosis

Case Progress:

- An empirical trial of steroid therapy for presumed sarcoidosis commenced with symptomatic improvement
- Withdrawal of steroids attempted in view of the inconclusive lymph node biopsies which resulted in a marked deterioration with hypotension, listlessness and obtundation
- Adrenal insufficiency was suspected
- Improved symptomatically after reinitiating glucocorticoids

Case History:

- 76 year old man with recently diagnosed primary hypothyroidism
- Presented with weight loss, lethargy and fatigue
- Physical exam : inguinal lymphadenopathy
- Staging CT thorax: bilateral mediastinal lymphadenopathy, (**Picture 1**) interstitial lung parenchymal changes, and splenomegaly
- Mild hypercalcaemia (Calcium-2.65 mmol/l) and hyponatremia noted
- Initial DD:lymphoma, malignancy, or granulomatous disease

Thoracic CT



Table 1
 Long Synacthen Test

Time	Cortisol (nmol/l)
Baseline	<28
8 Hours	414
24 Hours	1024

Investigations:

- Diagnostic excision biopsies of left inguinal and mediastinal lymph nodes were inconclusive
- ACTH stimulation with tetracosactide: sub-optimal cortisol increment with peak of 224nmol/l
- Adrenal antibodies were negative
- Serum ACTH <5 mg/l
- Secondary adrenal insufficiency confirmed by a subsequent long ACTH stimulation test (**Table 1**)
- CT of pituitary gland: no gross abnormality

Discussion:

- The exact mechanism of injury in hypothalamic-pituitary sarcoidosis is unclear
- Endocrine dysfunction was historically attributed to granulomatous infiltration of the hypothalamus and pituitary though hypothalamic insufficiency and pituitary responsiveness to hypothalamic releasing factors are now thought to be the main cause of hypopituitarism
- Central hypoadrenalism can present with protracted and non-specific symptoms and pose a diagnostic challenge in the absence of any gross structural pituitary abnormality
- A high index of suspicion remains the cornerstone of diagnosis