A case of Hypothalamic-Pituitary Sarcoidosis with Hypothermia

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Introduction

Sarcoidosis is a multisystem noncaseating granulomatous disease with prevalence of 40/100,000(1). 1% of patients with sarcoidosis have isolated neurosarcoidosis, few cases involve the hypothalamus/pituitary (65 reported cases from 2002-2014)(2).

We present a case of hypothalamic-pituitary sarcoidosis with hypothermia (1 case reported between 2002-2014)(2), leptomenigitis, diabetes insipidus and pan-hypopituitarism.

Case Report

Family brought a previously healthy 23-year-old African American male to the hospital due to fatigue, hypersomnolence for 2 weeks, intermittent headache for one month. Physical exam revealed non-focal neurological exam, photophobia.

MRI of the brain showed diffuse leptomeningeval enhancement with increased signal in suprasellar, optic chiasm, optic tracts. Differential diagnosis was TB/pyogenic meningitis, sarcoidosis and diffuse carcinomatosis.

Cerebrospinal fluid showed protein 418 mg/dl (normal 15-45), WBC 230cumm (normal 0-5) (84% lymphocytic), ACE 4U/L (normal 0-2.5), negative gram stain and culture. CT chest showed sub centimetre pre-tracheal lymph nodes.

Pituitary functions showed free T4 0.6ng/ml (normal 0.93-1.7), TSH 1.37 μIU/ml (normal 0.27-4.2), FSH 0.4milliUnit/ml (normal 1-11), LH <0.1 milliUnit/ml (normal 1-8), ACTH 39.5pg/ml (normal 7.2-63), Prolactin 14.3ng/ml (normal 0.5-17), total testosterone <20ng/ml (normal 37-198).

He tested negative for ANA, HIV, Hepatitis C, Mycobacterium TB Interferon-Gamma. He later developed diabetes insipidus and hypothermia (temperature of 33 °C). He underwent transphenoidal biopsy of the Dura (frozen section showed no granuloma), biopsy of inferior portion of pituitary gland (frozen section showed granuloma).

He was started on desmopressin, methylprednisolone, levothyroxine and external warming. Patient had significant improvement following treatment. Repeat brain MRI showed near complete resolution of previous findings. He was discharged home on hydrocortisone, desmopressin and levothyroxine.

Conclusions:

Neurosarcoidosis can present with diverse clinical features depending on the anatomical location of the disease. Common presentations include facial palsy, leptomenigitis, hypopituitarism. Rarely, it can affect thermo regulation centre of hypothalamus. It is important to recognize and treat early as it can be life threatening.

References: