Development of an Empty Sella in Neurosarcoidosis

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Introduction

An empty sella develops when arachnoid herniates into the sella turcica through a congenital or acquired defect in the dura, and cerebrospinal fluid (CSF) fills the sella and compresses the pituitary gland. Radiologically, the expanded sella is defined as partially empty if less than 50% is filled with CSF and the pituitary gland thickness is ≥ 3mm, or total, when more than 50% of the sella is filled with CSF and the gland thickness is ≤ 2 mm. A partial empty sella is a common radiographic finding, and is usually asymptomatic, and found incidentally. Endocrine deficiencies may occur, but are much more common in total empty sella [1].

Empty sella is also divided into two groups clinically, primary and secondary. Patients with primary empty sella are more often female and overweight [2], and some have idiopathic intracranial hypertension [3]. An empty sella may also develop following pituitary injury. Disorders associated with secondary empty sella include wrents in the diaphragm sella, pituitary macroadenomas treated surgically or by radiotherapy or following the spontaneous necrosis of pituitary apoplexy, Sheehan’s syndrome, vasculitis, infection, head trauma, and autoimmune hypophysitis. In several cases, a pituitary mass, believed to represent autoimmune hypophysitis, was documented to convert to an empty sella [4]. For neurosarcoidosis, there are case reports of empty sella by CT [5] and pneumoencephalogram [6] but the condition is rarely mentioned among the causes of empty sella. This case report demonstrates the evolution of a pituitary mass in a patient with neurosarcoidosis into an empty sella.
Case Report

A 56 year-old African American woman with hypertension presented with progressive bilateral vision loss, difficulty walking, vertigo, headache, hearing loss, extreme polyuria and polydipsia, and a 20lb weight loss over 4 months. She had deficits in cranial nerves II, VII and VIII, and a wide based gait.

Results

Endocrine tests indicated pan-hypopituitarism: LH 0.21U/L (reference range in menopause 13.1-86.5), FSH 1.01U/L (reference range in menopause 21.5-131), free T4 0.35ng/dL (reference range 0.78-2.19), TSH 0.44μU/L (reference range 0.46-4.68), GH 0.2ng/ml (reference range <10), IGF-1 118ng/ml (reference range 50-317; -1.4 SD for age), IGFBP3 2.5mg/L (reference range 3.4-6.9), A.M. cortisol 0.83μg/dL (reference range in menopause 21.5-131), free T4 0.35ng/dL (reference range in menopause 13.1-86.5), FSH 1.0IU/L (reference range 0.78-2.19), and ACTH 6pg/mL (reference range 4.5-22.7).

Magnetic Resonance Imaging (MRI) revealed diffuse dural thickening, pituitary enlargement and mild leftward deviation of the pituitary stalk (Figure 1A). The level of angiotensin converting enzyme in serum 40U/L (reference range 9-67), A.M. cortisol 0.83μg/dL (reference range 3.4-6.9), and ACTH 6pg/mL (reference range 6-50). The PRL level was increased at 56ng/mL (reference range 3.0-18.6). The plasma osmolality was 290mOsm/kg while the urine osmolality was 236mOsm/kg which rose to 827mOsm/kg following dDAVP, consistent with central diabetes insipidus.

Outcome and follow-up

With treatment, her weight increased 37kg, her neurological symptoms were unchanged, and pan-hypopituitarism and diabetes insipidus persisted. She continued to complain of polydipsia and polyuria even when the urine specific gravity was maintained at 1.018. After 12 months of treatment, methotrexate was stopped, and prednisone was slowly reduced to a maintenance dose, with addition of azathioprine for steroid sparing.

Discussion

Sarcoidosis is a multisystem inflammatory disorder that is defined by the histological finding of non-caseating granulomas. While the disorder can affect any organ, the lung, liver, eye and skin are most commonly involved. The nervous system is affected in 5-14% of cases, which may occur in isolation [7]. Patients with neurosarcoidosis may present with partial or complete anterior hypopituitarism, diabetes insipidus (DI), and hyperprolactinemia [8]. Excess thirst and polyuria may also occur without DI [9]. The diagnosis of isolated neurosarcoidosis can be challenging, as it can present with a wide variety of symptoms and imaging findings [9,10]. Involvement of the leptomeninges can be diffuse, focal or multifocal. The brain parenchyma and spinal cord may be affected, and in some cases the MRI appears normal. Basilar leptomeningeal disease can involve the cranial nerves and the hypothalamus-pituitary unit where the differential diagnosis includes lymphocytic hypophysitis, histiocytosis, tuberculosis, leukemia and metastasis. Measurement of the angiotensin converting enzyme level in serum and CSF is rarely helpful [11], and biopsy is needed for definitive diagnosis. Neurosarcoidosis patients are generally treated with high dose corticosteroids and immunosuppressive agents, and while the neurosarcoid lesions may regress, endocrine dysfunction is usually irreversible [8]. This case demonstrates that empty sella can be the direct outcome of sarcoidosis affecting the pituitary.

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References


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