Interrupting the Axis: A Case of IgG-4 Related Hypophysitis in a Young Hispanic Male

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Background: Hypophysitis refers to inflammation of the pituitary gland that can lead to the disruption of the hypothalamic-pituitary-adrenal axis. Primary hypophysitis has five variants differentiated by histologic findings: lymphocytic, granulomatous, xanthomatous, IgG4-related, and necrotizing. IgG4-related hypophysitis is very rare and is commonly a manifestation of a multi-organ systemic disorder. We present a case of a 43-year-old man presenting with severe headache and symptomatic hyponatremia in the setting of isolated IgG-4 related hypophysitis.

Case Presentation: A 43-year-old male presented with a two-day history of severe posterior headache associated with several episodes of vomiting, photophobia, generalized weakness and fatigue. Patient had no history of headaches and denied any recent trauma, focal weakness, altered mentation, changes in vision or seizure activity. Vitals on admission were remarkable for BP 179/99 mmHg. Neurological examination was non-focal, cranial nerves were grossly intact. Labs showed unremarkable CBC, sodium 117 mmol/L, potassium 3.3 mmol/L, chloride 84 mmol/L, glucose 244 mg/d, TSH 0.6 uIU/mL, cortisol 4.6 ug/dL, urine osmolality 540, serum osmolality 259 and prolactin level 7.8. CT head without contrast showed a mass-like enlargement of the Sella measuring 1.6 cm x 1.4 cm suspicious for macroadenoma. Patient was started on hypertonic saline, fludrocortisone, hydrocortisone, and salt tablets in the setting of suspected macroadenoma. Once the patient was stabilized, he underwent transsphenoidal resection of pituitary lesion. Pathology of the lesion showed a small area rich in plasma cells with positive IgG4 immunostaining in most of the plasma cells, consistent with IgG4-related pituitary hypophysitiis.

Conclusion: IgG4 hypophysitis usually presents in a context of multiple organ IgG4 involvement, however isolated hypophysitis has been reported. The patients present with mass effect symptoms and/or anterior hypopituitarism. When histologic diagnosis is obtained via biopsy, IgG4 hypophysitis can be managed exclusively with corticosteroids.

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