Visual Vignette

Submitted by
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Case Presentation: A 34-year-old man presented with a 6-month history of polydipsia and polyuria with a daily consumption of 7 to 8 L of fluid with matching urinary output. He had a history of holocranial headache that was not associated with vomiting, seizures, or loss of consciousness. The patient had normal libido and erectile function. Findings from physical examination, which included thorough genital and fundus examinations, were unremarkable. Anterior pituitary functions were normal. Findings from a water deprivation test indicated partial cranial diabetes insipidus. Initial magnetic resonance imaging is shown in Figure 1. After 1 year of therapy, magnetic resonance imaging was repeated (Fig. 2). **What is the diagnosis?**

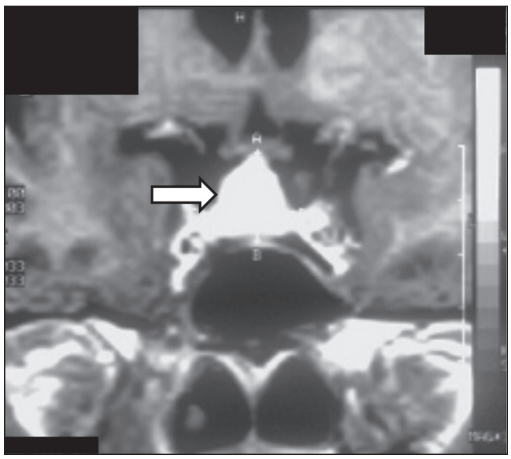

Fig. 1

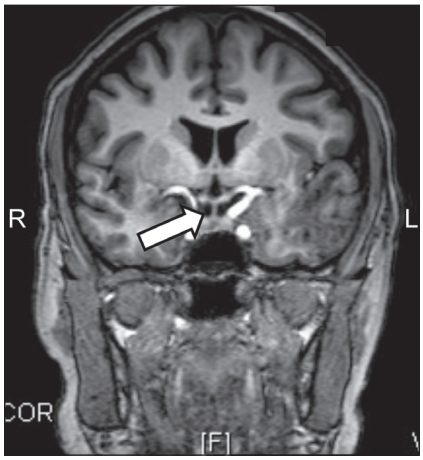

Fig. 2

Answer: Autoimmune hypophysitis (lymphocytic hypophysitis). Figure 1 shows a homogenous contrast-enhancing mass with pituitary stalk thickening. Figure 2 shows complete resolution of stalk thickening after treatment. Diagnosis of lymphocytic hypophysitis was made based on clinical, laboratory, and radiologic features. Our patient had diabetes insipidus and a characteristic finding of a symmetric mass with homogenous enhancement and stalk thickening on magnetic resonance imaging. Imaging also showed an intact sellar floor (1).

Differential diagnoses such as sarcoidosis, histiocytosis, lymphoproliferative disorders, and other granulomatous lesions must be ruled out before a diagnosis of autoimmune hypophysitis is considered. Lymphocytic hypophysitis is a rare inflammatory disorder of the pituitary with a definite female predilection. Most cases are women presenting with hypopituitarism during the peripartum period, but symptoms may also appear in postmenopausal women. Lymphocytic hypophysitis is extremely rare in men, usually presenting with headache and visual disturbances; there may be deficiency of multiple anterior pituitary hormones or it may present as isolated diabetes insipidus. The etiology of the disease is unknown, but it is associated with autoimmune diseases like thyroiditis and adrenalitis, suggesting an autoimmune cause. Assessment for pituitary antibodies has poor sensitivity and specificity for detecting lymphocytic hypophysitis (1).

In lymphocytic hypophysitis, the abnormality is mainly confined to the anterior pituitary, resulting in partial or total hypopituitarism and hyperprolactinemia, with a small proportion of individuals developing diabetes insipidus (2). Our patient presented with diabetes insipidus, did not show any evidence of anterior pituitary hormone deficiency, and had a near-normal prolactin level.

Glucocorticoid therapy has been used successfully for the remission of symptoms and for decreasing lesion size. For patients whose conditions do not respond to medical therapy, transsphenoidal surgery is an option (3). Our patient had a marked resolution of stalk thickening with a normal-sized pituitary after 1 year of steroid treatment. He did not develop deficiency of anterior pituitary hormones during follow-up, but he continued to have diabetes insipidus for which he was treated with desmopressin nasal spray.

REFERENCES

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