Sapporo Medical Journal
Volume 57, Issue 05, May 2023

Lymphocytic Hypophysitis Presenting as Central Diabetes Insipidus: A Case Report

Hani Shalabi[1]
[1]Department of Internal Medicine, college of Medicine, university of Jeddah, Saudi Arabia

Abstract— Lymphocytic hypophysitis, a rare autoimmune disorder causing inflammation of the pituitary gland, often presents diagnostic challenges due to its varied manifestations and rarity. This report discusses a unique case of a 20-year-old female patient presented with polyuria, polydipsia, and fatigue. The initial laboratory evaluation suggested diabetes insipidus (DI), and magnetic resonance imaging (MRI) revealed an enlarged pituitary gland with mild, uniform thickening of the pituitary stalk. The patient was diagnosed with lymphocytic hypophysitis and managed with desmopressin and hydrocortisone. This case report highlights the challenges in diagnosing and treating lymphocytic hypophysitis presenting with central DI. The report emphasizes the importance of considering lymphocytic hypophysitis as a potential differential diagnosis in cases of unexplained DI, particularly in young women. Additionally, it underlines the significance of a comprehensive diagnostic approach, including laboratory tests and imaging studies, to reach a conclusive diagnosis in such rare and complex cases. The management of lymphocytic hypophysitis involves addressing the underlying autoimmune inflammation and associated hormonal imbalances. In this case, the use of desmopressin and hydrocortisone proved successful in managing the patient's symptoms and improving her overall clinical condition. This case report contributes valuable information to the limited body of knowledge surrounding the diagnosis, management, and follow-up of lymphocytic hypophysitis, particularly when presenting with central diabetes insipidus.

Keywords: Autoimmunity, Desmopressin, Hypophysitis, Diabetes Insipidus, Inflammation, Pituitary.

1. Case report

A 20-year-old female patient reported to the endocrine clinic, complaining of polyuria, polydipsia, and fatigue for the past two weeks.The patient reported a daily urine output ranging from 6 to 8 liters, characterized by a clear and dilute appearance. and experiencing constant thirst. The patient denied having headaches, visual disturbances, or neurological deficits, and had no significant past medical or family history. She was not taking any medications and denied illicit drug use. Her vital signs were stable, and her body mass index (BMI) was 27 kg/m2. Her physical examination showed no abnormalities. Initial lab tests showed a serum sodium of 148 mmol/L (normal range: 135-145 mmol/L), serum osmolality of 298 mOsm/kg (normal range: 275-295 mOsm/kg), and urine osmolality of 117 mOsm/kg (normal range: 500-800 mOsm/kg), suggesting DI. Other tests, Serum glucose: 90 mg/dL (normal range: 70-100 mg/dL fasting) ,TSH: 2.5 mIU/L (normal range: 0.4-4.0 mIU/L), Free T4: 1.2 ng/dL (normal range: 0.9-1.7 ng/dL),ACTH: 20 pg/mL (normal range: 7.2-63.3 pg/mL), 8 AM cortisol: 15 µg/dL (normal range: 6-23 µg/dL),Prolactin: 15 ng/mL (normal range: 2-29 ng/mL for non-pregnant females),BUN: 15 mg/dL (normal range: 7-20 mg/dL),Creatinine: 0.95 mg/dL (normal range: 0.84-1.21 mg/dL for females),AST: 25 U/L (normal range: 10-40 U/L),ALT: 30 U/L (normal range: 7-56 U/L),ALP: 80 IU/L (normal range: 44-147 IU/L), Total bilirubin: 0.5 mg/dL (normal range: 0.1-1.2 mg/dL) .

An MRI displayed an enlarged pituitary gland with mild, uniform thickening of the pituitary stalk suggesting a potential underlying pathological condition (figure 1)

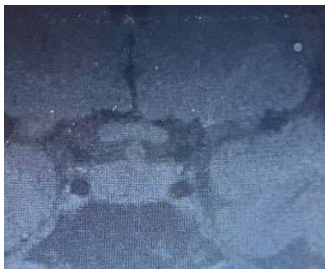

Figure 1a (Pre-Contrast): This image reveals the pituitary gland prior to contrast administration. The gland appears enlarged and exhibits mild, uniform thickening of the pituitary stalk, suggestive of an underlying pathology.

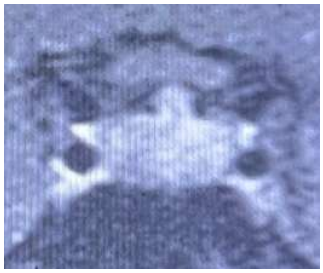

Figure 1b (Post-Contrast)

Due to the patient's unwillingness to undergo surgery and considering other available clinical evidence, a working diagnosis of lymphocytic hypophysitis was made.The patient was admitted for further evaluation and treatment, including a water deprivation test. After 8 hours of fluid restriction, her urine osmolality was 487 mOsm/kg (normal range after fluid restriction: >800 mOsm/kg), indicating an inability to concentrate urine. Desmopressin administration at a dosage of 0.1 mcg intranasally twice daily led to an increase in urine osmolality to 858 mOsm/kg, confirming a central diabetes insipidus diagnosis. She was treated with intravenous fluids, including 0.9% normal saline and 5% dextrose, to address dehydration and electrolyte imbalance, and desmopressin to alleviate her diabetes insipidus symptoms. Her serum sodium and osmolality levels normalized after three days of treatment, with serum sodium at 142 mmol/L and serum osmolality at 290 mOsm/kg. She was discharged with a hydrocortisone tapering dose, starting at 20 mg in the morning and 10 mg in the evening for one week, and then gradually reducing by 2.5 mg per week, and desmopressin prescriptions at a dosage of 0.1 mcg intranasally twice daily. Outpatient follow-ups were scheduled for monitoring her progress.At her six-month follow-up, her polyuria was well managed with desmopressin. A subsequent MRI revealed a decrease in the pituitary gland's size and enhancement (figure 2), indicating a response to hydrocortisone. However, her diabetes insipidus continued, necessitating ongoing desmopressin treatment. The patient attended regular appointments at the outpatient endocrinology clinic, where her symptoms and electrolyte levels remained stable with her desmopressin. Serial MRI imaging showed a gradual reduction in pituitary gland size, indicating an improvement in the inflammatory process. She continues to receive annual follow-up for monitoring.

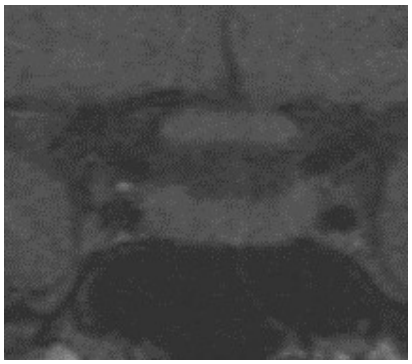

Figure 2a (Pre-Contrast): The pre-contrast image of the pituitary gland shows a noticeable decrease in size compared to the initial MRI.

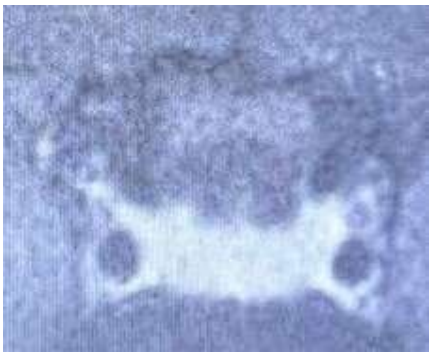

Figure 2b (Post-Contrast)

2. Discussion

Lymphocytic hypophysitis (LYH) is a rare autoimmune disorder that affects the pituitary gland, predominantly in females during pregnancy or postpartum period [1]. Recent epidemiological data suggests an estimated prevalence of 1 in 9 million people [2]. The clinical presentation of LYH can be nonspecific, making diagnosis challenging [3].In this case, the 20-year-old female patient presented with polyuria, polydipsia, and fatigue, along with lab findings indicating central diabetes insipidus (DI). These findings are consistent with other reported cases of LYH. An MRI displayed an enlarged pituitary gland with mild, uniform thickening of the pituitary stalk, raising suspicion for LYH[4] The management of LYH generally involves addressing the underlying inflammation and managing hormonal deficiencies[5]. In this case, the patient was treated with hydrocortisone and desmopressin to target inflammation and manage central DI, respectively. This treatment approach aligns with standard management practices for LYH. The patient's symptoms and radiological findings improved with steroid therapy, supporting the LYH diagnosis [6]. The prognosis for patients with LYH is variable, but most cases show improvement with appropriate treatment [7]. At the six-month follow-up, this patient's polyuria was well managed with desmopressin, and the MRI showed a decrease in the pituitary gland's size and enhancement, indicating a response to hydrocortisone [8]. However, her diabetes insipidus continued, necessitating ongoing desmopressin treatment. The patient attended regular appointments at the outpatient endocrinology clinic, where her symptoms and electrolyte levels remained stable with desmopressin.Differential diagnoses for this case include

other causes of central DI, such as pituitary adenoma, craniopharyngioma, sarcoidosis, histiocytosis, or metastatic lesions to the pituitary gland. Ultimately, the patient's clinical presentation, lab findings, and response to treatment supported the diagnosis of LYH. In conclusion, this case highlights the importance of considering LYH in patients presenting with central DI and pituitary abnormalities on imaging. Early diagnosis and appropriate management are crucial for favorable outcomes. Regular follow-ups are necessary to monitor patients' progress and detect potential complications.

3. Conclusion

This case report underscores the importance of considering lymphocytic hypophysitis in patients presenting with central diabetes insipidus and pituitary abnormalities on imaging. Early diagnosis and appropriate treatment, including hydrocortisone and desmopressin, are essential for achieving favorable outcomes in managing this rare autoimmune disorder. Continuous monitoring through regular follow-ups at an outpatient endocrinology clinic is necessary to ensure symptom stability, electrolyte level maintenance, and early detection of potential complications. Clinicians should be vigilant in their approach to diagnosing and managing patients with LYH to improve overall patient care and long-term prognosis.

4. References

[1] Caturegli P, Newschaffer C, Olivi A, et al. Autoimmune hypophysitis. Endocr Rev. 2005;26(5):599-614.

[2] Buxton N, Robertson I. Lymphocytic and granulocytic hypophysitis: a single Centre experience. Br J Neurosurg. 2001;15(3):242-5.

[3] Imura H, Nakao K, Shimatsu A. Lymphocytic infundibuloneurohypophysitis as a cause of central diabetes insipidus. N Engl J Med. 1993;329(10):683-9.

[4] Sato N, Sze G, Endo K. Hypophysitis: endocrinologic and dynamic MR findings. AJNR Am J Neuroradiol. 1998;19(3):439-44.

[5] Gutenberg A, Bell JJ, Lupi I, et al. Pituitary and systemic autoimmunity in a case of intrasellar germinoma. Pituitary. 2011;14(4):388-94.

[6] Cheung CC, Ezzat S, Smyth HS, et al. The spectrum and significance of primary hypophysitis. J Clin Endocrinol Metab. 2001;86(3):1048-53.

[7] Brown T, Dhillon S, Chung B, et al. Lymphocytic hypophysitis in the elderly: A case presentation and review of the literature. Int J NeurosciBehav Sci. 2016;4(4):67-71. doi: 10.1016/j.inat.2016.10.007.

[8] Honegger J, Buchfelder M, Schlaffer S, Droste M, Werner S, Strasburger C, et al. Treatment of Primary Hypophysitis in Germany. J Clin Endocrinol Metab. 2015;100(9):3460-9. doi: 10.1210/jc.2015-2146.