Adult E-Poster

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LANGERHANS HISTIOCYTOSIS-RELATED HYPOPHYSITIS: A DISTINCT CAUSE OF CRANIAL DIABETES INSIPIDUS

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INTRODUCTION/BACKGROUND

Cranial diabetes insipidus (DI) is a rare manifestation of hypothalamo-pituitary axis disorders. Langerhans Cell Histiocytosis (LCH), a clonal disease of dendritic cells, can infiltrate multiple organs, including the pituitary gland. When the posterior pituitary is involved, LCH can cause secondary hypophysitis, leading to cranial DI. Due to overlapping symptoms with more common conditions, diagnosis can be delayed.

CASE

A 22-year-old female with a one-year history of bullous skin lesions and oral ulcers was initially diagnosed with pemphigus vulgaris and treated with prednisolone. However, a skin biopsy later confirmed LCH. Imaging, including CT of the brain, neck and thorax-abdomen-pelvis, showed multisystem involvement (skin, ear, thyroid, and thymus). Hence, she underwent chemotherapy.

After the first cycle of chemotherapy with vinblastine and high-dose dexamethasone, she developed vomiting and lethargy. Laboratory tests revealed a hyperosmolar hyperglycemic state (serum osmolality 323 mOsm/kg). Despite normalisation of her blood glucose, she developed marked polyuria (>13,000 mL/day). Further testing showed serum osmolality of 294 mOsm/kg and urine osmolality of 54 mOsm/kg. A desmopressin trial resulted in a >50% increase in urine osmolality (398 mOsm/kg at 2 hours; 511 mOsm/kg at 4 hours), confirming cranial DI. MRI revealed a 1.1 × 1.3 × 1.0 cm lesion in the pituitary infundibulum, consistent with LCH-related hypophysitis, along with empty sella syndrome, likely secondary to chronic pituitary involvement and glucocorticoid therapy. She was treated with sublingual desmopressin and high-dose steroids, showing clinical improvement.

CONCLUSION

This case highlights the need to consider cranial DI as a manifestation of systemic LCH, especially in young adults with multisystem disease. Given the overlap of symptoms, early recognition of cranial DI is crucial for timely management.

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DISCORDANT THYROID FUNCTION TESTS: DIAGNOSTIC CHALLENGES IN A PATIENT WITH A TSH-SECRETING PITUITARY ADENOMA

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INTRODUCTION/BACKGROUND

Discordant Thyroid Function Tests (TFTs) may present a diagnostic challenge in clinical practice. Assay interference must be excluded before proceeding to further investigations. Rarely, the underlying diagnosis may be a TSH-secreting pituitary adenoma (TSHoma) or resistance to thyroid hormone (RTH). Making a diagnostic distinction between these two conditions is important as their clinical management varies significantly.

CASE

A 47-year-old engineer was referred to the Endocrine clinic with symptoms of hyperthyroidism and discordant TFTs. These symptoms improved with propranolol and carbimazole. Only his paternal aunt has a goitre on family history. On physical examination, there was no palpable goitre. His TFT was discordant before starting carbimazole [TSH 4.32 mIU/L (NR:0.27-4.20), fT4 34.1 pmol/L (NR: 12.0-22.0), fT3 11.6 pmol/L (NR: 3.10-6.80)]. Discordance persisted despite using two different immunoassays after stopping carbimazole. Alpha-subunit was raised (2.47 IU/L; NR: 0.0-0.7). A pituitary MRI showed pituitary macroadenoma. Pituitary hormones were within normal limits except for an elevated IGF-1. An oral glucose tolerance test was inconsistent with acromegaly as nadir growth hormone was 0.8 mIU/L (0.27 g/L). Several tests distinguishing a TSHoma from RTH were unavailable, so the patient underwent a somatostatin test. Following octreotide, TSH was suppressed with low-normal free thyroid hormone levels, highly suggestive of a TSHoma.

Our patient underwent an endoscopic transsphenoidal hypophysectomy. The histology report confirmed TSH and growth hormone-secreting adenoma.

CONCLUSION

The case illustrates challenges in establishing the diagnosis of a TSHoma with resource limitations and supports using intramuscular octreotide LAR as a diagnostic tool. Appropriate evaluation of discordant TFT is paramount