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BALANCING DISEASE CONTROL AND METABOLIC HARM: A CASE OF IgG4-RELATED HYPOPHYSITIS

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INTRODUCTION

Immunoglobulin G4-related hypophysitis (IgG4-RH) is a rare fibro-inflammatory disorder affecting the pituitary gland. Glucocorticoids remain the first-line therapy, but their use may be complicated in patients with significant metabolic comorbidities. We report a case of suspected IgG4-RH presenting with hyperosmolar hyperglycemic state (HHS), highlighting the challenges of balancing disease control against glucocorticoid metabolic toxicity adverse effects.

CASE

A 32-year-old female with obesity and newly diagnosed diabetes mellitus was admitted with HHS. Prior to admission, she reported weight fluctuations, episodic headaches, progressive visual disturbance, and secondary amenorrhea. Following resolution of HHS, persistent polyuria of 10–16 L/day prompted further evaluation and led to a diagnosis of arginine vasopressin deficiency. Anterior pituitary hormonal work-up revealed hypogonadotropic hypogonadism. Pituitary magnetic resonance imaging demonstrated infundibular thickening measuring 0.5 cm, with concomitant marked bilateral parotid enlargement. Serum IgG4 was elevated at 2.26 g/L (0.63–2.01), raising strong suspicion for IgG4-RH with systemic involvement. Histopathological confirmation from the parotid gland biopsy was consistent with sialadenosis.

She was commenced on sublingual desmopressin and cyclical sex hormone replacement therapy. Given the provisional diagnosis of IgG4-RH, oral prednisolone 40 mg daily was initiated as a reduced induction regimen. However, treatment was poorly tolerated, with rapid weight gain from 93 to 100 kg and worsening glycemic control. Prednisolone was therefore tapered rapidly to 10 mg daily. Repeat imaging demonstrated interval improvement in infundibular thickening, but no functional endocrine recovery was observed.

CONCLUSION

This case illustrates the therapeutic challenge of managing IgG4-RH in the setting of pre-existing metabolic syndrome. Although glucocorticoids are effective for induction, their metabolic adverse effects may significantly restrict

treatment tolerability. Early consideration of steroid-sparing agents, such as rituximab or azathioprine, may be important to achieve remission while minimizing glucocorticoid-related adverse effects.

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A DIAGNOSTIC DILEMMA IN ECTOPIC ACTH SYNDROME: WHEN BIOCHEMISTRY AND IMAGING CONFLICT

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INTRODUCTION

Ectopic ACTH secretion (EAS) is a rare cause of Cushing's syndrome, accounting for 5–15% of cases. Pulmonary neuroendocrine tumors are the most frequent cause. However, primary tumor localization remains a significant diagnostic challenge, delaying effective treatment.

CASE

A 59-year-old female with diabetes mellitus presented with refractory hypertension and hypokalemia. Initial laboratory findings revealed a baseline cortisol of 1,164 nmol/L and morning adrenocorticotrophic hormone (ACTH) of 19.5 pmol/L (normal <10.2 pmol/L), with an overnight dexamethasone suppression test cortisol of 624 nmol/L. Pituitary magnetic resonance imaging (MRI) and computed tomography (CT) scans of the thorax, abdomen, and pelvis were initially unremarkable. A PET scan identified mild hypermetabolism in the left adrenal gland and gastrointestinal tract, though colonoscopy revealed only chronic colitis. An intravenous desmopressin stimulation test demonstrated a 181% rise in ACTH and a 35% rise in cortisol, pointing toward pituitary Cushing's. However, repeat pituitary MRI remained normal.

A high-dose dexamethasone suppression test showed approximately 34% cortisol suppression (cortisol 1,128 nmol/L to 745 nmol/L), suggesting ectopic Cushing's syndrome. Bilateral inferior petrosal sinus sampling demonstrated a peak central-to-peripheral ACTH ratio of 1.8 on the right and 1.6 on the left, pointing toward ectopic Cushing's.

Localization with DOTATATE PET-CT identified a 7 × 9 × 8 mm nodule in the right middle lobe. A wedge resection was performed. Histopathology confirmed a typical carcinoid tumor with clear resection margins. Immunohistochemistry revealed tumor cells positive for CK AE1/