Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS II

Undiagnosed Chronic Eczema as a Presentation of Glucagonoma in MEN 1 Syndrome

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Background: Glucagonomas are pancreatic tumors arising from the islets cell of Langerhans that over secrete glucagon. Necrolytic migratory erythema (NME) is an important feature for the recognition of glucagonomas. Glucagonomas occurring in MEN1 is infrequent and seen in less than 3% of all glucagonomas.

Clinical Case: A 51-year-old male presented to the clinic multiple visits for rash affecting the legs and genital area of two months. His medical history include type 2 DM and HT. The rash was attributed to subacute eczema and treated with topical steroids but showed no improvement. The skin eruption initially appeared on lower extremities progressed to trunk, and face. The skin lesions were associated with weight loss and stomatitis. On physical examination, skin showed ill-defined erythematous plaque exhibiting annular pattern, scale, and erosion on all extremities and perioral area. When the skin lesions healed, the new cutaneous eruptions occurred. Laboratory testing revealed plasma glucose of 185 mg/dL. The skin biopsy reported vacuolated keratinocytes in the epidermis with eosinophil cytoplasm, compatible with NME leading to further workup for pancreatic tumor. CT abdomen revealed tumor mass 9.6x6 cm at the pancreatic tail and multiple nodules in the liver. Somatostatin receptor scintigraphy showed an area of increased radiotracer uptake at the tail of the pancreas and multiple liver nodules corresponding to the previous CT scan. Serum glucagon was 923 pg/mL, confirming the diagnosis for glucagonoma. The patient was treated with distal pancreatectomy, and enucleation of liver metastases. Histopathological reported grade 2 well-differentiated NET. During the admission, the patient was found to have a parathyroid level of 79.3 pg/mL and increased uptake in the left and right lower regions of the thyroid gland from parathyroid MIBI scan, indicating hyperfunctioning parathyroid glands. All pituitary hormones were within normal ranges and no pituitary tumor was detected by the MRI brain. BMD showed osteoporosis at the lumbar spine and left femoral neck. The patient was referred to general surgery for subtotal parathyroidectomy. Pathological of resected parathyroid glands reported parathyroid hyperplasia. Postoperative PTH level and calcium were returned to normal range. Genetic testing focused on MEN1 gene, and the mutation was identified. After four months follow up plasma glucagon decreased to 425 pg/dL, patient had complete resolution of the cutaneous lesions.

Conclusion: Glucagonomas are a rare pancreatic tumors and often difficult to recognize. Chronic eczema may misdiagnose for NME and delay diagnosis. NME can be challenging for physicians to recognize NME which is an important key to diagnose glucagonoma. Even though MEN1 association with glucagonoma is infrequent, awareness of such is important to allow appropriate testing for MEN1 in patients with glucagonoma.

Adrenal

ADRENAL CASE REPORTS II

Case Series of ACTH-Secreting Pheochromocytoma: Diagnosis and Management

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INTRO

ACTH-secreting pheochromocytoma is rare, posing challenges in diagnosis and management. Here we report two cases with different presentations and pre-operative approaches.

CASES

Patient 1 A 59 yo female who presented for basal cell carcinoma resection experienced hypertensive crisis intra-op and NSTEMI due to stress cardiomyopathy. The patient reported spells of palpitations, diaphoresis, tremors and pallor since 2015. She had elevated plasma metanephrine (M) 19.4 (0-0.49nmol/L), normetanephrine (NM) 24.7 (0-0.89nmol/L), 24-hour urine M 18711 (30-180mcg) and NM 11897 (128-484mcg). Imaging demonstrated a 5.6 x 5.5 x 5.8cm left adrenal mass. After pre-operative alpha and beta blockade, patient underwent left adrenalectomy.

Pre-op 24-hour urine cortisol was checked as part of secondary HTN evaluation and found to be elevated at 219 (3.5-45mcg). She did not have any features of Cushing's and this was initially felt to be an appropriate stress response. Due to persistent post-op hypotension and a low 10PM random cortisol level of 3.4mcg/dL, which was inappropriate given patient's clinical status, hydrocortisone was started with subsequent hemodynamic improvement. Our suspicion was that she had an ACTH-secreting pheochromocytoma, which would have resulted in hypercortisolism pre-op, and secondary AI post-op. Pathology report later confirmed ectopic ACTH production with 25% cells positive with immunostaining.

Patient 2 A 48 yo female was admitted for hypertensive urgency with symptoms of palpitations, facial and abdominal swelling, easy bruising and proximal muscle weakness. Her labs showed elevated 24-hour urine epinephrine 115 (0-20mcg), norepinephrine 366 (0-135mcg), plasma M 288 (0-62pg/mL) and NM 475 (0-145pg/mL), and an AM cortisol was 47.9mcg/dL. Aldosterone and renin levels were <1ng/dL and 0.775ng/mL/hr. Imaging revealed 3.4 x 3.1cm right adrenal mass with portal venous phase of 53 HU and no washout. AM cortisol level post 1mg dexamethasone was 62.6mcg/dL with an ACTH level of 222pg/mL. An ACTH-producing pheochromocytoma was suspected. Despite progressive uptitration of alpha and beta blockade, her BP remained poorly controlled. Her blood glucose also increased to 300s range. Ketoconazole 400mg BID was subsequently added to the regimen with rapid and marked improvement in BP and glycemic control. She underwent