BCL-6, leucocytes common antigen (LCA) and Ki67 proliferating index was 60% in the tumour cells. A diagnosis of diffuse large B cell lymphoma (DLBCL) was made. The patient was referred to the haematology team and started on chemotherapy.

CONCLUSION

Primary adrenal lymphoma is a rare entity with a generally poor prognosis. They usually involve both adrenal glands, but unilateral adrenal involvement can occur in about one-third of patients. In patients with large adrenal masses and constitutional symptoms, the initial dilemma is to differentiate between adrenal carcinoma versus other forms of malignancies or chronic infections. Adrenal biopsy is generally avoided in suspected adrenal carcinoma as it may be harmful because it can lead to tumour seeding. In our patient, the presence of multiple lymphadenopathy which was accessible for biopsy helped clinch the diagnosis of adrenal lymphoma. The presence of lymphadenopathy with large adrenal masses, even if unilateral, should prompt suspicion of the diagnosis of adrenal lymphoma.

EP_A013

CT STAGING THAT UNVEILS A MYSTERY – ASYMPTOMATIC PHEOCHROMOCYTOMA ASSOCIATED WITH NEUROFIBROMATOSIS TYPE 1

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Muzhaffar Mokhtar,[1] Masliza Hanuni MA,[1] Siti Sanaa WA,[1] Hussain Mohamad,[2] Nor Hisham Muda[2]
[1]*Endocrinology Division, Department of Medicine, Hospital Sultanah Nur Zahirah, Kuala Terengganu, Malaysia*
[2]*Breast and Endocrine Surgery Unit, Department of Surgery, Hospital Sultanah Nur Zahirah, Kuala Terengganu, Malaysia*

INTRODUCTION/BACKGROUND

Pheochromocytomas and paraganglioma (PPGL) are catecholamine-secreting tumours, derived from chromaffin cells. The classical triad comprises paroxysms of headache, palpitation, and diaphoresis. About 10% of patients are asymptomatic. At least one-third of patients with PPGLs have hereditary disease caused by germline mutations. This includes neurofibromatosis type 1 (NF-1) which may predispose patients to pheochromocytoma and occurs in 0.1-5.7% of cases.

We report a case of a 57-year-old female with hypertension and diabetes who was referred from the surgical team after an incidental finding of a right adrenal mass on abdominal CT performed for rectal adenocarcinoma staging. She denied paroxysms or other symptoms that suggest catecholamine or cortisol excess. Blood pressure was well controlled with a single agent. Examination revealed axillary freckling, multiple cafè au lait spots and generalized cutaneous nodules which were present since adolescence.

CASE

Serial abdominal CT scans showed increasing size of right adrenal mass measuring 4.4 x 5.4 x 5.9 cm (previously 4.4 x 5.1 x 5.6 cm) with presence of fluid-fluid level within, with HU ranging from HU 20 (anteriorly) and HU 70 (posteriorly). Metanephrine 5.42 umol/L (0.33-1.53), normetanephrine 8.0 umol/L (0.88-2.88) and 3-methoxytyramine 1.16 umol/L (0.66-2.60) were elevated on 24-hour urine collection. Thyroid function test and serum calcium were normal. Histopathological examination of the cutaneous nodule confirmed neurofibromas. She underwent open right adrenalectomy and HPE was consistent with pheochromocytoma. Three months post adrenalectomy, urine metanephrines had normalized, and there was no tumor residual or recurrence on CT imaging. She no longer requires any anti-hypertensive drug, and we were able to withdraw insulin therapy.

CONCLUSION

Though rare, the combination of NF-1 with pheochromocytoma in our patient is an offbeat presentation of adrenal incidentaloma in a patient with multiple cutaneous nodules, hypertension and diabetes. Screening for pheochromocytoma should be done in a patient with NF-1 and hypertension. Prompt treatment will alleviate the detrimental effect of catecholamine excess and improve the patient's quality of life.

EP_A014

CORTISOL DAY CURVE TO GUIDE GLUCOCORTICOID REPLACEMENT IN A PATIENT WITH ADRENAL INSUFFICIENCY ON ANTI-TUBERCULOSIS THERAPY

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Jun Kit Khoo, Tharsini Sarvanandan, Ying Guat Ooi, Nicholas Ken Yoong Hee, Quan Hziung Lim, Lee Ling Lim, Sharmila Paramasivam, Shireene Vethakkan, Jeyakantha Ratnasingam
Endocrinology Unit, Department of Medicine, Universiti Malaya, Kuala Lumpur, Malaysia

INTRODUCTION/BACKGROUND

Hydrocortisone in divided doses (typically 15 – 25 mg/day) is the most common form of glucocorticoid replacement regimen in patients with adrenal insufficiency (AI). However, this may be inadequate for patients on CYP3A4 inducers

which affect glucocorticoid metabolism. While there are no specific guidelines on dose adjustment, cortisol day curve (CDC) could be used to guide optimal replacement. We report a case of a female with AI secondary to pituitary tuberculosis requiring hydrocortisone dose adjustment following initiation of anti-tuberculosis therapy (anti-TB).

CASE

A 45-year-old female presented with a worsening headache for a month, without any constitutional symptoms. Imaging revealed a heterogenous sellar mass (1.3 x 1.5 x 2.1 cm) without chiasmal compression or cavernous involvement. Blood investigations showed serum cortisol <14 nmol/L (reference interval 145-619), FT4 10 pmol/L (reference interval 11.5 – 22.7) and TSH <0.01 m IU/L (reference interval 0.55-4.78). Other blood investigations were normal. She was commenced on hydrocortisone 10 mg BD (8 am, 2 pm) and levothyroxine 50 mcg OD. Trans-sphenoidal resection was performed, and histopathology revealed necrotising granulomatous inflammation with caseating necrosis. The diagnosis of pituitary tuberculosis was made, and first line anti-TB drugs (rifampicin, isoniazid, pyrazinamide, and ethambutol) were initiated. Despite pre-emptively increasing hydrocortisone to 20 mg BD, she experienced postural hypotension, lethargy, and nausea soon after initiation of anti-TB medications, especially in the early afternoon and evening. Cortisol day curve was done by measuring serum cortisol hourly from 8 am – 8 pm while she took her regular hydrocortisone 20 mg BD. Serum cortisol levels were <14 nmol/L (8 am), 1009 nmol/L (9 am), 664 nmol/L (10 am), 386 nmol/L (11 am), 217 nmol/L (12 pm), 88 nmol/L (2 pm), 761 nmol/L (3 pm), 857 nmol/L (4 pm), 521 nmol/L (5 pm), 256 nmol/L (6 pm), and 85 nmol/L (8 pm). Hydrocortisone was adjusted to 20 mg (8 am), 10 mg (1 pm) and 5 mg (6 pm) to counter the trough levels. This led to a marked improvement in her symptoms.

CONCLUSION

CYP3A4 inducers (anti-TB) affect glucocorticoid metabolism and replacement in patients with AI. The cortisol day curve could be used as a guide to tailor therapy in situations where adequate replacement doses and timing are not easily identified.

EP_A015

A PERPLEXING MIXED CORTICAL AND MEDULLARY ADRENAL NEOPLASM PRESENTING WITH INDETERMINATE ADRENAL MASS

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Wan Mohd Hafez WH,[1] Masliza Hanuni MA,[1] Siti Sanaa WA,[1] Hussain Mohamad,[2] Nor Hisham Muda,[2] Norsyahinaz Hashim,[3] Navarasi A/P S Raja Gopal[4]

[1]*Endocrinology Unit, Medical Department, Hospital Sultanah Nur Zahirah, Kuala Terengganu, Malaysia*

[2]*Breast and Endocrine Surgery Unit, Department of Surgery, Hospital Sultanah Nur Zahirah, Kuala Terengganu, Malaysia*

[3]*Pathology Department, Hospital Sultanah Nur Zahirah, Kuala Terengganu, Malaysia*

[4]*Pathology Department, Hospital Putrajaya, Putrajaya, Malaysia*

INTRODUCTION

A mixed cortical and medullary adrenal neoplasm originating in the adrenal gland is an exceedingly rare tumour characterized by the coexistence of cell nests from both the adrenal cortex and medulla within a singular mass, which results in the production of adrenocortical steroid hormones and catecholamines (CA). We report a case of mixed cortical medullary adrenal neoplasm presenting with indeterminate adrenal mass.

CASE

A 51-year-old female, with a known case of left breast mucinous carcinoma, underwent a right mastectomy and axillary clearance and completed 6 cycles of systemic chemotherapy. The computed tomography of the thorax, abdomen and pelvis (CT TAP) surveillance post-treatment revealed left adrenal indeterminate incidentaloma. Computed tomography adrenal protocol showed that the left adrenal mass was heterogeneous with a size of 1.8 x 2.2 x 2.4 cm, with a plain HU of 18 HU, absolute contrast washout of 21%, and relative contrast washout of 15%. Biochemically, both the overnight dexamethasone suppression test (ODST) and low dose dexamethasone suppression test (LDDST) were not suppressed with values of 81 nmol/L and 119 nmol/L, respectively. A 24-hour urine metanephrine collection demonstrated results within the normal range: metanephrines 0.4 umol/day (0.1-6.2), normetanephrine 1 umol/day (0.2-1.3) and 3-methoxytyramine 0.8 umol/day (0.1-1.79). Given that the left adrenal indeterminate incidentaloma had autonomous cortisol secretion, a left adrenalectomy was performed. Remarkably, the histopathological examination (HPE) unveiled a mixed corticomedullary adrenal tumour.