

was administered, followed by oral isavuconazole and itraconazole. Computed tomography (CT) scan demonstrated enlarged bilateral adrenal glands with the right side measuring 4.9 x 2.6 x 6.8 cm, and the medial limb of the left adrenal gland measuring 4.3 x 2.5 cm. The lateral limb of the left adrenal gland was 3.6 x 2.1 cm. Ten weeks after antifungal therapy was started, CT scan revealed a smaller left adrenal lesion, but the right adrenal lesion remains unchanged. Short synacthen test showed PAI with peak cortisol 246 nmol/L, ACTH 20.3 pmol/L (1.6-13.9). He is awaiting adrenal biopsy pending urinary metanephrines. Glucocorticoid replacement was initiated. Antifungal therapy would be continued for no less than one year.

CONCLUSION

Adrenal histoplasmosis is common and histopathological analysis is crucial in managing such cases. It is important to be vigilant about infections like histoplasmosis as a potential cause of PAI. Delay in treatment could result in life-threatening consequences.

EP A011

MALIGNANT PARAGANGLIOMA IN AN ADOLESCENT

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INTRODUCTION/BACKGROUND

Pheochromocytomas (PCC) and paragangliomas (PGL) (PPGL) are rare neuroendocrine tumours occurring in children and adolescents. Nevertheless, they are the most common endocrine tumours in the paediatric population and account for 0.5–1% of paediatric hypertensive cases. We describe a 16-year-old female with malignant paraganglioma.

CASE

A previously healthy 16-year-old female presented with a one-month history of intermittent headaches associated with palpitations and presyncopal attacks. The first blood pressure reading revealed that she was hypertensive, with a BP of 159/116. She had no chest pain, shortness of breath, diaphoresis, abdominal pain, or diarrhoea. There is no family history of hypertension in the young or endocrine disorder. The patient is lean with a BMI of 14.2 kg/m². No goitre, cushingoid or acromegalic features were present. The hormonal workup done was consistent with phaeochromocytoma (normetanephrine: 55.30 umol/day) (35.5 X ULN)). Other forms of work-up for secondary hypertension were unremarkable. Adrenal CT imaging

revealed an enhancing mass at the left pararenal space measuring 4.1 x 4.7 x 4.7 cm with local infiltration to the tail and body of the pancreas complicated by a left renal infarct. Therefore, she was diagnosed with left paraganglioma with local infiltration. Preoperatively, she was started on oral prazosin 1 mg, 6 hourly and oral bisoprolol 2.5 mg daily and successfully underwent open resection of the left paraganglioma. Unfortunately, she remained hypertensive post-surgery, indicating a possible malignant paraganglioma. Thus, she was restarted on antihypertensive medications. Gallium-DOTATE scan and genetic testing have been arranged to aid further management.

CONCLUSION

Diagnosis of pheochromocytoma and paraganglioma is paramount during the evaluation of secondary hypertension in the paediatric population. Although they are uncommon, possible curative surgery can be offered. All children should be subjected to genetic testing given the high rate of inheritance of these tumours. Subsequently, all patients with genetic mutations ought to be under lifelong surveillance in view of the risk of recurrence and malignancy.

EP_A012

A RARE CASE OF UNILATERAL ADRENAL LYMPHOMA WITH LYMPHADENOPATHY

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INTRODUCTION/BACKGROUND

Adrenal lymphoma is an extremely rare and highly invasive malignant disease. We report a rare case of unilateral adrenal lymphoma with lymphadenopathy.

CASE

A 68-year-old male presented with abdominal discomfort, polyuria and weight loss of 15 kg over 6 months. Physical examination revealed a thin elderly man with fullness over the left lumbar and inguinal lymph nodes. Laboratory tests showed markedly elevated lactate dehydrogenase (LDH) levels of >690 UI/L (<248) and hypercalcemia. A computed tomography (CT) scan revealed a large left adrenal mass (11.6 x 8.3 x 9.6 cm) with multiple matted abdominal lymph nodes, raising a suspicion of adrenal malignancy. Following this, hormonal profile was done which showed normal cortisol and catecholamines. An ultrasound-guided trucut biopsy of the right inguinal lymph nodes was performed. The microscopic examination showed a malignant tumour composed of mononuclear cells with pleomorphic nuclei with high mitotic figures. On immunohistochemistry, the tumour cells were positive for vimentin, CD20, CD10,



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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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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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