CR-GE-21

A CASE REPORT OF VON HIPPEL-LINDAU DISEASE WITH METASTATIC FOLLICULAR THYROID CARCINOMA AND RECURRENT PHEOCHROMOCYTOMA: ASSOCIATION OR COINCIDENCE?

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INTRODUCTION
Von Hippel-Lindau (VHL) disease is a rare autosomal dominant syndrome caused by germ line mutations in the VHL tumor suppressor gene. VHL manifestations encompass a spectrum of tumors, namely retinal and central nervous system hemangioblastomas, renal cell carcinoma, pheochromocytoma, pancreatic endocrine tumours and endolymphatic sac tumours. However, the association between VHL and follicular thyroid carcinoma (FTC) is unknown.

CASE
A 25-year-old lady with unremarkable family history was diagnosed with metastatic FTC and pheochromocytoma 10 years ago. She underwent a right adrenalectomy and was in remission postoperatively confirmed by normal 24-hour urinary catecholamines. As for the FTC, she underwent total thyroidectomy and 2 sessions of radioactive iodine ablation therapy for lymphovascular invasion and bone metastases. Post-ablation whole body scans and thyroglobulin level were normal. Unfortunately, she was lost to follow-up and presented 5 years later with headache, palpitation and uncontrolled hypertension. 24-hour urinary catecholamines revealed elevated noradrenaline and CT scan showed a left adrenal mass, suggestive of a recurrent pheochromocytoma and was treated with left adrenalectomy. Molecular genetic testing demonstrated c.467A>G (p.Tyr156Cys), which is confirmatory of VHL disease.

CONCLUSION
This is the first reported case of bilateral pheochromocytoma and FTC in VHL disease. The association of VHL gene alteration and thyroid carcinoma particularly FTC from literature is scarce. More studies are needed to evaluate the association between VHL and FTC to determine the surveillance strategy, prognosis and appropriate treatment.

KEY WORDS
von hippel-lindau, pheochromocytoma, follicular thyroid carcinoma

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A CASE OF BENIGN PARAGANGLIOMA WITH TRANSIENT HYPERPARATHYROIDISM

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INTRODUCTION
The co-existence of phaeochromocytoma with hypercalcemia is attributed to various factors including MEN2 (Multiple Endocrine Neoplasia), catecholamine-stimulated bone resorption and PTH secretion by the parathyroid glands, ectopic production of PTH by pheochromocytoma and PTH-related protein (PTHrP). In contrast to other causes, serum PTH is suppressed in PTHrP besides elevated urinary cyclic AMP and detection of PTHrP immunoreactivity in the tumor tissue. Irrespective of the cause, hypercalcaemia resolved in all reported cases after removal of the phaeochromocytoma lesions.

CASE
A 37-year-old Chinese lady with 2 years of hypertension developed hypertensive emergency with intracranial bleeding in 2017. She did not have any paroxysmal symptoms or features of other hereditary syndromes. Work-up for secondary causes of hypertension noted raised 24-hour urine norepinephrines at 290 micrograms. CT adrenals showed a left para-aortic lesion measuring 2.5x1.6x3.7 cm which was confirmed by Gallium 68 PET CT at L3 vertebrae level. She was incidentally noted to have hypercalcaemia with cCa 2.7 mmol/l and iPTH 108.7 pg/ml with normal phosphate, alkaline phosphatase and vitamin D levels. Neck ultrasound and sestamibi scan were negative for parathyroid lesion and abdominal ultrasound didn't reveal stones. Negative serum calcitonin and CEA ruled out medullary thyroid carcinoma as part of MEN2. However, the calcium and PTH levels normalised within a few months, even before laparoscopic excision of the benign left paraganglioma.

CONCLUSION
The likely reason for hypercalcaemia in this case could be due to stimulation of PTH secretion by the catecholamines which was transient as the hypercalcaemia resolved before removal of phaeochromocytoma.

KEY WORDS
paranganglioma, hyperparathyroidism, hypercalcaemia