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REMISSION OF SURGICAL HYPOPARATHYROIDISM POST ADRENALECTOMY FOR CUSHING'S SYNDROME

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BACKGROUND
Adrenal insufficiency (AI) is a well-recognized cause of hypercalcemia. In patients with hypoparathyroidism, the effect of coexisting AI on calcium level has not been well-described.

CASE
We report a case of a patient with hypoparathyroidism who was able to discontinue her long-term calcium and calcitriol replacement after adrenalectomy for Cushing's syndrome.

A 63-year-old female with post-operative hypoparathyroidism after total thyroidectomy for papillary thyroid carcinoma (in 2005) was maintained on a stable dose of calcium carbonate 1 g BID and calcitriol 0.5 mcg BID. In 2015, she developed ACTH-independent Cushing's syndrome and underwent right adrenalectomy for a 3.6 cm right adrenal adenoma. Post-operatively, she was diagnosed with AI [serum cortisol 1.04 μg/dl (NV: 5.27–22.45)] and was symptomatic of glucocorticoid withdrawal syndrome despite up-titration of hydrocortisone replacement to 35 mg daily. Interestingly, she was found to have hypercalcemia (highest corrected calcium level 2.97 mmol/L), necessitating down-titration and eventual total discontinuation of her calcium carbonate and calcitriol replacement. She remained normocalcemic despite being off calcium and calcitriol for 4 months. As her symptoms improved, oral calcium and calcitriol were slowly resumed at lower doses (calcium carbonate 500 mg BID and calcitriol 0.25 mcg OD) to maintain her calcium level. On follow-up, she still has AI [8 am serum cortisol 25.4nmol/L (NV: 133 – 537)] and requires 15 mg daily hydrocortisone replacement.

CONCLUSION
The pathophysiology of hypercalcemia in AI is incompletely understood. Our case supported a parathyroid-independent mechanism. In rare cases of AI with concurrent hypoparathyroidism, close monitoring of calcium levels is needed for medication dose adjustments to achieve normocalcemia.

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RARE CASE OF EXCLUSIVELY DOPAMINE-SECRETING PARAGANGLIOMA IN MULTIPLE ENDOCRINE NEOPLASIA TYPE 2A (MEN 2A)

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BACKGROUND
Pheochromocytomas in MEN2A are usually intra-adrenal. Rarely, they may manifest as paragangliomas. Predominantly or exclusively dopamine-secreting pheochromocytomas and paragangliomas (PPGL) are rare with only 33 cases reported in the literature.

CASE
We report a case of exclusively dopamine paraganglioma in the context of MEN 2A.

A 72-year-old male was diagnosed with MEN2A following family screening in 1996. Genetic analysis revealed a mutation in codon 634 of the RET proto-oncogene (C634Y). He underwent total thyroidectomy for medullary thyroid carcinoma in 1996 and total parathyroidectomy for primary hyperparathyroidism in 1997. His yearly 24-hour urinary catecholamines had been within the normal ranges. However, in August 2019, his urinary dopamine was raised at 1033 μg/day (normal range: 64.0-400). Urinary adrenaline and noradrenaline were not elevated. Repeated 24-hour urinary metanephrines in August 2020 yielded an elevated 3-methoxytyramine level of 21.8 μmol/day (normal range: 0.10-1.79). Urinary metanephrines and normetanephrines remained within normal ranges. He has hypertension which is well-controlled on two agents. He is otherwise asymptomatic with no paroxysmal attacks of catecholamine excess. Iodine-131 meta-iodobenzylguanidine (I-131 MIBG) imaging revealed an avid lesion in the mediastinum with no tracer uptake at the adrenal glands. The patient declined further interventions.

CONCLUSION
Our case highlights the importance of measuring urinary or plasma dopamine even in MEN2A. Dopamine-secreting PPGL typically lacks the classical presentation of paroxysmal attacks and are often extra-adrenal. Exclusively dopamine-secreting PPGL is rare. To our knowledge, this represents the first case in a patient with MEN2A.