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LETTERS TO THE EDITOR

Clinical-scientific notes

Cutaneous lichen amyloidosis in multiple endocrine neoplasia

A non-English-speaking 40-year-old woman presented with severe hypertension and multiorgan failure. No past medical history could be elicited. In the first 24 h of her admission, she developed cardiogenic shock with a left ventricular ejection fraction of 25%, labile blood pressure, acute kidney injury, hyperglycaemia, metabolic acidosis, disseminated intravascular coagulation, ischaemic hepatitis and acute limb ischaemia. A unifying diagnosis was not evident.

With the acquisition of medical records from overseas, it was subsequently discovered that the patient had a background of bilateral phaeochromocytoma and adrenalectomy 10 years prior. She was then found to have large masses in both adrenal beds on CT abdomen (Fig. 1) with elevated urinary and plasma catecholamines and metanephrines (Table 1). Catecholamine crisis was diagnosed as the cause of her dramatic presentation and staged bilateral thoracolumbar adrenalectomy confirmed recurrent bilateral phaeochromocytoma. Calcitonin was later found to be raised at 553 ng/L (<20) and thyroid ultrasound revealed two nodules. Biopsy was consistent with medullary thyroid cancer (MTC) with immunohistochemistry positive for calcitonin, carcinoembryonic antigen, amyloid, chromogranin and TTF-1. Total thyroidectomy and lymph node dissection demonstrated multifocal MTC and lymph node metastasis.

Family history was remarkable for her mother and five of six siblings having bilateral phaeochromocytoma. One of the siblings also had MTC. The patient's daughter was later found to have MTC and possibly an early phaeochromocytoma. Multiple endocrine neoplasia type 2A (MEN2A) was suspected based on the clinical and family history and Sanger sequencing of the *RET* proto-oncogene in the proband identified a pathogenic missense mutation, p.C634F.

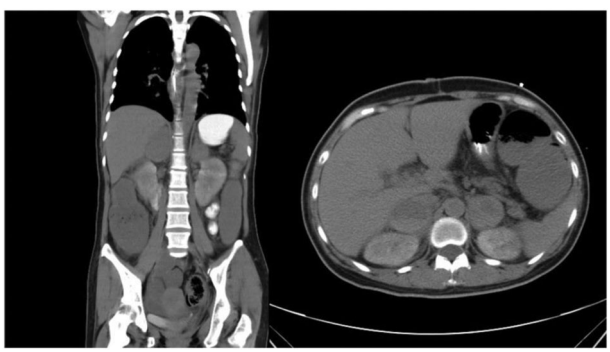

Figure 1 CT abdomen demonstrated bilateral adrenal tumours measuring 7.0 × 4.8 × 4.3 cm on the right and 3.9 × 3.6 × 3.5 cm on the left.

Table 1 Urine and plasma biochemistry showed predominant adrenaline secretion consistent with adrenal phaeochromocytoma

Test	Result	Reference range
Urine adrenaline (nmol/d)	409*	0–150
Urine noradrenaline (nmol/d)	644	0–785
Urine metanephrine (μmol/d)	8.9*	0.0–2.1
Urine normetanephrine (μmol/d)	3.8	0.0–5.6
Plasma adrenaline (nmol/L)	127.0*	0.0–1.5
Plasma noradrenaline (nmol/L)	164.7*	0.1–6.3
Plasma metanephrine (nmol/L)	29.1*	<0.4
Plasma normetaphrine (nmol/L)	15.4*	<0.9

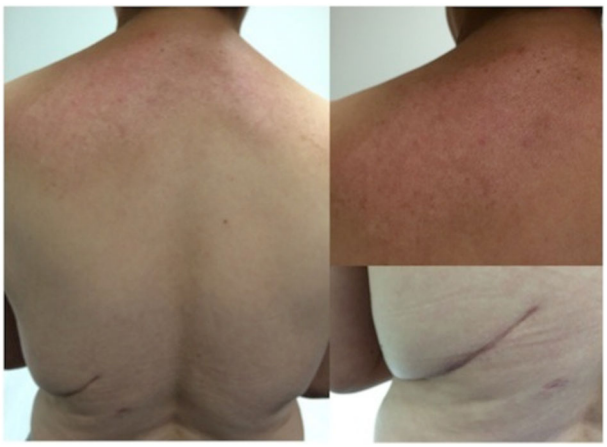

Figure 2 Inspection of the patient's back revealed cutaneous lichen amyloidosis in the interscapular region and surgical scars across the flanks consistent with previous bilateral adrenalectomy.

At follow up, the patient reported interscapular pruritis that began at age 12 years and led to a scaly rash with patchy depigmentation around age 20 years. Of her seven family members with clinical MEN2A, three had an identical rash. The patient was diagnosed with cutaneous lichen amyloidosis (CLA), a condition found in up to one-third of patients with MEN2 and particularly in association with mutations involving codon 634 of the *RET* gene as observed in our patient.[1]

Cutaneous lichen amyloidosis is classically dermatomal affecting dermatomes C8-D3, and it has been hypothesised to be secondary to notalgia paraesthetica, a neuropathy involving the dorsal rami nerves.[2] However, the pathogenic role of the mutated RET product remains unclear. It is possible that there is a defect in neural development directly related to the *RET* proto-oncogene as certain *RET* mutations have been found to cause both endocrine neoplasia and Hirschprung disease characterised by congenital absence of ganglion cells in the gastrointestinal tract.[3]

Whilst CLA is an important marker of MEN2, it does not appear to predict the clinical course of disease in affected subjects and it has been observed in a patient with familial MTC alone.[1,4] Why it affects some and not all subjects in MEN2 kindreds remains unclear with evidence limited to case reports and case series. This poor evidence base also implies that the link between CLA and codon 634 mutations in *RET* may simply reflect the overall predominance of codon 634 mutations observed in MEN2.[5]

Herein, we demonstrate the importance of this easily identifiable but little known dermatological sign in the early diagnosis of MEN2. Recognition of CLA along with the patient's pre-existing surgical scars (Fig. 2) at initial presentation to the local institution may have expedited the diagnosis of MEN2 in this patient. Patients with thyroid nodules, suspected phaeochromocytoma or primary hyperparathyroidism should accordingly be examined for this sign, which should in turn prompt MEN2 phenotypic and genotypic screening. Awareness of CLA may help circumvent episodes of catecholamine crisis due to unrecognised phaeochromocytoma especially at times of surgery.

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