Uncontrolled hypertension: hints from the skin

CASE PRESENTATION

A 59-year-old woman presented with 4 months history of paroxysmal episodes of palpitations and headache. She was recently detected to have hypertension and was on two antihypertensive medications. Her blood pressure (BP) at presentation was 160/110 mm Hg and optic fundus showed grade 2 hypertensive retinopathy. She had features of neurofibromatosis-1 (NF-1) (figure 1 showing multiple neurofibromatoses on the posterior aspect of the trunk and figure 2 showing café-au-lait macules on forearm). In view of uncontrolled hypertension and findings of NF and café-au-lait macules, a possibility of an underlying phaeochromocytoma was considered. Venous sampling was done to assess renal function, serum electrolytes and metabolic profile. The results of these tests were normal. Urine tests did not show the presence of proteinuria or active sediments. Sonogram of the abdomen showed normal-sized kidneys. Biochemical evaluation showed elevated 24 hours urine normetanephrine of 1902 µg (N<600 µg) and metanephrine of 692 µg (N<350 µg). CT scan of the abdomen (figure 3) showed 3.5×2.8 cm sized intense and heterogeneously enhancing right suprarenal mass, diagnostic of phaeochromocytoma.

As part of preoperative preparation, this patient was admitted under our care for a period of 2 weeks. She was initiated on an alpha blocker, namely, extended-release prazosin. She was started on a low dose of 2.5 mg per day; this was gradually up-titrated to a dose of 10 mg two times per day. As postural fall in BP and tachycardia are expected with alpha-blockade, she was encouraged to hydrate herself adequately with 4–5 litres of water. She was also started on a high sodium diet to ensure that she was volume replete. Her intake and output were assessed daily; heart rate and BP were recorded in the supine and upright posture. Three days prior to surgery, she was initiated on a beta blocker, namely, extended-release metoprolol. Prior to her being taken up for surgery, her blood pressure was 110/70 mm Hg, with no postural fall, and heart rate was recorded to be 70–80 beats per minute.

She underwent laparoscopic excision of the tumour and histopathology was consistent with phaeochromocytoma. Following surgery, the patient was stable and was able to be weaned off all antihypertensive medications.

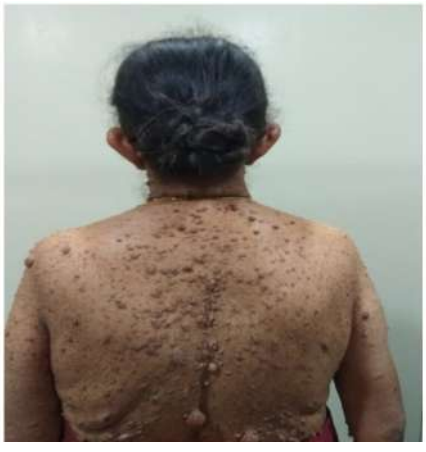

Figure 1 Multiple neurofibromatoses on the posterior aspect of the trunk.

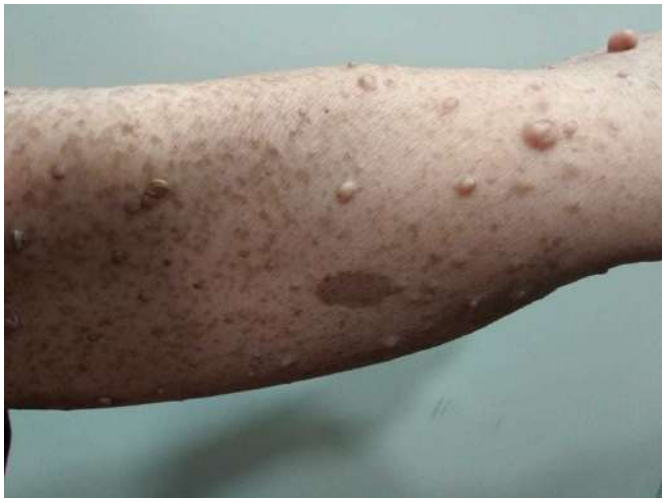

Figure 2 Café-au-lait macules on the forearm.

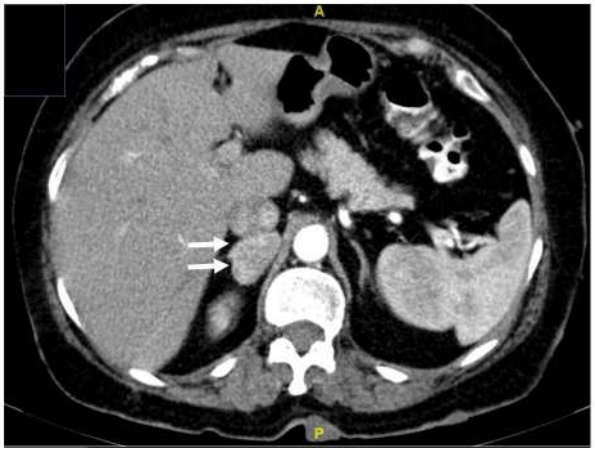

Figure 3 CT scan of the abdomen showing 3.5×2.8 cm sized intense and heterogeneously enhancing mass in the right suprarenal region.

DISCUSSION

Phaeochromocytoma is a rare cause of secondary hypertension, accounting for about 0.2%–0.6% of cases.[1] NF-1 affects approximately 1 in 3500 individuals worldwide and it has an autosomal dominant inheritance. Phaeochromocytoma occurs in 0.1%–5.7% of patients with NF-1.[2] Uncontrolled hypertension in the presence of neurocutaneous markers, such as café-au-lait macules and neurofibromas, should alert the clinician to the possibility of an underlying catecholamine-secreting tumour, as missing this crucial diagnosis could prove detrimental to these subjects.[3] It is also prudent to counsel the family members about disease inheritance, its varied manifestations and the need for lifelong follow-up.

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