Case Report

An unusual cause of toe necrosis

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ABSTRACT

Peripheral vascular disease is a rare feature of pheochromocytoma. This potentially catastrophic but curable tumor should be suspected in combination of distal necrosis with hypertension and palpable pulses. We report such an unusual case of pheochromocytoma presenting as toe necrosis.

Key words: Catecholamine, peripheral vascular disease, pheochromocytoma, toe necrosis

Introduction

Peripheral vascular disease (PVD) is a common vascular disease. PVD is commonly associated with absent and/or feeble peripheral pulses. The common causes of PVD are Buerger's disease, atherosclerosis, diabetes, and Raynaud's disease. Pheochromocytoma is associated with various cardiovascular problems like hypertension, cardiomyopathy, myocardial infarction, stroke,[1] and rarely associated with peripheral vascular disease, acute limb ischemia,[2-4] toe necrosis,[5] and intermittent claudication.[6-8] If no other signs and symptoms of pheochromocytoma are present, diagnosis is easily overlooked.

We report such a case of pheochromocytoma presenting as toe necrosis.

Case Report

A 50-year-old male presented with history of pain in both lower limbs and ulcer in partially amputated left great toe since 8 months. He had history of pain in both lower limbs with blackish discoloration of left great toe for which he had undergone partial left great toe amputation by local practitioner before 8 months. He was a non-smoker. He had been treated for limb pain symptomatically before being referred to our institute. Before referral to the vascular surgery unit, CT angiography was done which incidentally detected left hypervascular suprarenal mass.

On examination, the patient was found to have tachycardia and hypertension. There were no peripheral signs of pheochromocytoma. Abdominal examination was unremarkable. Pulses were weak in both lower limbs as compared to hand, and great toe was partially amputated with ulcer [Figure 1].

Laboratory data showed hemoglobin (Hb) 11 gm/dl and random blood glucose 121 mg/dl. Coagulation profile and lipid profile were normal. Urine vanillylmandelic acid (VMA) was 10.3 (2–8 mg/24 h), urine epinephrine 16.8 (0–20 mg/24 h), urine norepinephrine 133.4 (15–80 mg/24 h), and serum cortisol after overnight dexamethasone suppression test was 46.72 (171–536 nmol/l).

CT angiography revealed relatively delayed runoff in lower limb arteries with marginally reduced caliber in right lower limb arteries and hypervascular left suprarenal mass suggestive of pheochromocytoma. Contrast-enhanced computed tomography (CECT) abdomen revealed a 5.2 × 3.5 × 5.8 cm hypervascular mass lesion replacing left adrenal, suggestive of pheochromocytoma [Figure 2].

Patient was started on antihypertensive. After adequate alpha and beta blockade, he underwent left adrenalectomy

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[Figure 3]. Histological examination showed a well-circumscribed and encapsulated tumor with monomorphic cells arranged in alveolar and nesting pattern, separated by thin vascularized septae. Individual cells were monomorphic with round nuclei, granular chromatin, inconspicuous nucleoli, and moderate to abundant granular cytoplasm. The features were suggestive of a pheochromocytoma.

Postoperatively, his BP was normal. On follow-up, his toe ulcer healed completely.

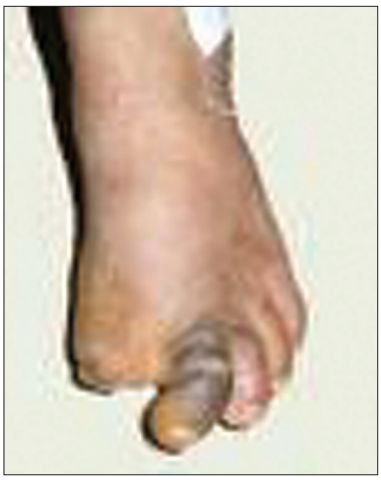

Figure 1: Left great toe partially amputated with an ulcer

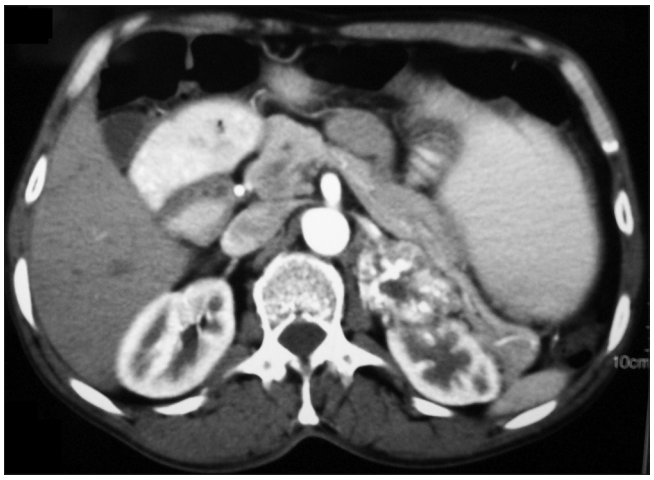

Figure 2: Contrast-enhanced computed tomography abdomen revealed a 5.2 × 3.5 × 5.8 cm hypervascular mass lesion replacing left adrenal, suggestive of pheochromocytoma

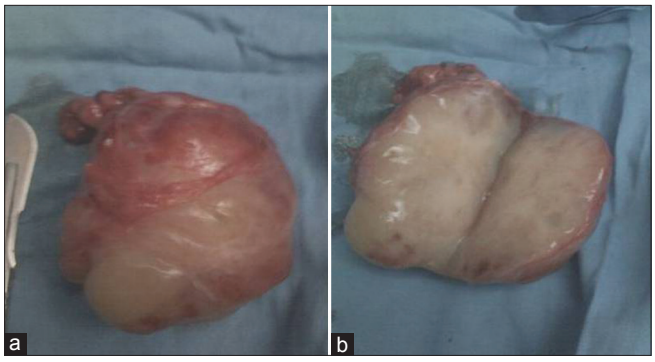

Figure 3: (a) Gross appearance of left suprarenal mass and (b) its cut section

Discussion

Classical clinical triad of pheochromocytoma is formed by tachycardia, sweating, and headache. In addition to the classical triad, pheochromocytoma is commonly associated with hypertension, cardiomyopathy, myocardial infarction, and cerebrovascular accident.[1] Rarely, pheochromocytoma is also associated with peripheral vascular disease with intermittent claudication, non-healing ulcers, and necrosis of toes. Our patient presented with non-healing ulcer over partially amputated left great toe.

High levels of catecholamine production may be the pathologic mechanism causing extreme vasoconstriction or diffuse arterial vasospasm and critical peripheral ischemia.[1,3,5,8-10] Significant negative correlation between plasma catecholamine concentration and skin blood flow has been demonstrated in pheochromocytoma.[10,11] Resolution of ischemia and wound healing after removal of pheochromocytoma confirms the suggestion that excess catecholamine was responsible for the development of critical toe ischemia in our patient.

High index of suspicion and timely diagnosis is extremely important, as these tumors are curable by surgical removal, and any surgery in a patient with unsuspected pheochromocytomas carries high risk of morbidity and mortality.

In conclusion, we report an unusual case of pheochromocytoma presenting as toe ischemia and necrosis, and hypertension that was unsuspected because of rarity of the tumor presenting as toe necrosis. Diagnosis is mainly clinical. This potentially catastrophic but curable tumor should be suspected in combination of distal necrosis with hypertension and palpable pulses.

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