A 24-year-old man presented with 2 years of treatment-resistant hypertension, headaches, palpitations, and sweats. One year earlier at an outside hospital, he was found to have elevated urine metanephrines, although an abdominal computed tomography scan and $^{123}$I-metaiodobenzylguanidine scintigraphy were unrevealing. He was lost to medical follow-up until he presented to this clinic with similar symptoms as well as a fixed livedoid rash on his back and limbs (Figure 1A), which had developed several months prior. Initial labs revealed elevated total serum metanephrines of 4302 pg/mL (normal, <206 pg/mL) and normetanephrines of 4258 pg/mL (normal, <149 pg/mL), as well as total 24-hour urine metanephrines of 9256 g (normal, <600 g). An abdominal computed tomography scan revealed a 3.5-cm right adrenal mass with no evidence of metastases, and repeat $^{123}$I-metaiodobenzylguanidine scintigraphy now demonstrated decreased right adrenal uptake consistent with necrosis. As thyroid ultrasound showed no suspicious lesions and the patient had no suggestive family history, testing for multiple endocrine neoplasia syndromes was not pursued. He underwent right adrenalectomy with resolution of his symptoms, improvement in his rash (Figure 1B), and normalization of metanephrine levels (total serum metanephrines 256 pg/mL, normetanephrines 256 pg/mL). Pathology demonstrated neoplastic neurosecretory cells characteristic of pheochromocytes.

Livedo reticularis, a rash that arises when arteriolar vasoconstriction leads to pooling of blood in surrounding venules, has rarely been described in conjunction with pheochromocytoma (1, 2). Indeed, the typical rash of pheochromocytoma is intermittent facial flushing associated with symptomatic catecholamine release. There is a growing body of literature describing fixed lesions that resolve with treatment of pheochromocytoma. In addition to livedo reticularis, these lesions include livedo racemosa, Raynaud’s phenomenon (1, 2), nodular and macular rashes, Addisonian discoloration (3), and hypochromic lesions or necrotic ulcerative lesions (4, 5). Of note, several of these conditions are physiologically akin to livedo reticularis (specifically livedo racemosa, Raynaud’s phenomenon, and possibly ulceration) in that they arise from vasospasm with or without local hyperviscosity and thrombosis. It is possible that intermittent catecholamine release results in increased vasomotor tone, viscosity, and hypercoagulability, leading to these protean cutaneous manifestations.

Acknowledgments

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The authors report no grants or fellowships supporting this work.

Disclosure Summary: The authors report no conflicts of interest.

References

Figure 1. On presentation to the Endocrinology Clinic, the 24-year-old man had a diffuse livedoid rash (A) covering his arms, legs, and back that improved after adrenalectomy (B).