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Preoperative Evaluation for Parathyroidectomy—Rule Out Pheochromocytoma

Rubin Bahuva, MD; Sudhir Manda, MD; and Saurabh Kandpal, MD
Cleveland Clinic Foundation, Cleveland, OH

Case Presentation: A 71-year-old woman presented for preoperative evaluation for parathyroidectomy. Her past medical history was significant for hypertension, diabetes mellitus, osteoporosis, and paroxysmal SVT. Labs had shown hypercalcemia and hyperparathyroidism, and ultrasonography revealed a 0.98 × 0.64 × 0.46 cm lesion typical of left lower parathyroid adenoma. Further questioning revealed that she had flash pulmonary edema and severe hypertension during her previous surgery. Her blood pressure (BP) control was suboptimal on multiple medications. Her BP was 182/110 with normal exam. Due to a history of intraoperative severe hypertension, pheochromocytoma was considered in the differential. Labs revealed elevated free plasma metanephrines and elevated epinephrine and norepinephrine levels of 3,934 and 1,824, respectively. CT of the abdomen revealed a 9.4 × 11.4 × 11.3 cm necrotic left adrenal mass. Since the patient had findings consistent with hyperparathyroidism and pheochromocytoma, multiple endocrine neoplasia syndrome type 2A (MEN-2A) was considered a possibility; however, there was no evidence of medullary thyroid carcinoma. She was started on phenoxybenzamine and scheduled for the pheochromocytoma surgery before the parathyroidectomy.

Case Discussion: Pheochromocytoma occurs in ~50% of patients with MEN-2A and hyperparathyroidism in 15% to 20%. About half of the pheochromocytomas are bilateral, and > 50% of patients who have had unilateral adrenalectomy develop a pheochromocytoma in the contralateral gland within a decade. Most clinicians recommend removing only the affected gland during primary surgery. If both adrenal glands are removed, glucocorticoid and mineralocorticoid replacement is mandatory.

From the internist's perspective, preoperative patient preparation is essential for safe surgery. Alpha-adrenergic blockers (phenoxybenzamine) should be initiated at low doses and titrated up. Because patients are volume-constricted, liberal salt intake and hydration are necessary to avoid orthostasis. Adequate alpha blockade generally requires 10 to 14 days, with a typical final dose of 20 to 30 mg phenoxybenzamine three times daily. Before surgery, the BP should be consistently below 160/90 with moderate orthostasis. Beta-blockers can be added after starting alpha-blockers. BP can be labile during surgery, particularly at the onset of intubation or when the tumor is manipulated. Nitroprusside infusion is useful for intraoperative hypertensive crises, and hypotension responds to volume infusion. Atraumatic endoscopic surgery has now become the method of choice. It may be possible to preserve the normal adrenal cortex, particularly in patients with hereditary disorders, in whom bilateral pheochromocytomas are more likely.

Conclusion: Physicians should evaluate for pheochromocytoma in patients undergoing parathyroidectomy, especially if patients have severe hypertension, as there is a well-known association between the two. These patients need initiation of alpha-blockers and surgery for pheochromocytoma before parathyroidectomy.