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Quieting the Catecholamine Storm: A Preoperative Approach to Managing the Pheochromocytoma Patient

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Introduction

The Who: A pheochromocytoma, a rare catecholamine-secreting tumor derived from chromaffin cells. Catecholamines released from the tumor stimulate both alpha- and beta-adrenergic receptors.

The Where: Typically an adrenal tumor, also associated with syndromes including von Hippel-Lindau (VHL) syndrome, multiple endocrine neoplasia type 2 (MEN 2), and neurofibromatosis type I (NF1).

The What: Secretes epinephrine and norepinephrine which can precipitate life-threatening hypertensive crises and cardiac arrhythmias. About 10% are malignant, which is defined as the presence of metastases.

The How: Surgical resection remains the standard treatment of choice.

The Why: A preoperative approach to the management of the pheochromocytoma patient has yet to be standardized. Here we provide an anecdotal preoperative outline to the management of a hemodynamically unstable patient suffering from pheochromocytoma.

Case Report

A 33-year-old Asian female presented to the emergency department with headaches, flushing, palpitations, diaphoresis, and insomnia. She was tachycardiac to 144 and hypertensive to 220/118. She had no previous history of hypertension. Orthostatic measurements were also positive as her standing blood pressure was 89/61. Creatinine and troponins were mildly elevated. EKG showed sinus tachycardia with 1st-degree AV block and right axis deviation. An abdominal CT revealed a 6 cm right adrenal mass contiguous with the liver and superior pole of the kidney. 24-hour urine catecholamine measurement revealed markedly elevated epinephrine and norepinephrine levels, confirming the diagnosis of pheochromocytoma. Surgery, nephrology, and endocrinology services were consulted. The patient was admitted to the intensive care unit due to the labile nature of her blood pressure. Treatment was initiated with phenoxybenzamine 10mg BID along with maintenance intravenous fluids of NSS 100 cc/hr. Her blood pressure remained labile on day two, with 74-179 systolic and 65-140 diastolic. She was still tachycardic to 110. Fluids were increased to 150cc/hr. On day three, the phenoxybenzamine dose was doubled to 20mg BID. Her tachycardia did not resolve and on day four, metoprolol 12.5mg BID was added. The patient's blood pressure normalized and remained stable. She was asymptomatic, creatinine returned to baseline, and she was transferred to lowlevel monitoring. Her blood pressure was stable at 90-110 systolic and 50-70 diastolic. After achieving hemodynamic stability, surgery was scheduled. On day 5, metoprolol was doubled to 25mg BID. This regimen was continued until day 9 when metoprolol was taken back down to 12.5mg BID in preparation for surgery. This treatment regimen was continued for 1 more day before a successful laparoscopic right adrenalectomy.



Image 1.

Transverse view of CT scan showing large right suprarenalmass contiguous with the liver. The mass was a pheochromocytoma after confirmation by elevated 24-hour urinary metanephrines.



Image 2.
Coronal view of CT scan showing a 6 cm pheochromocytoma.

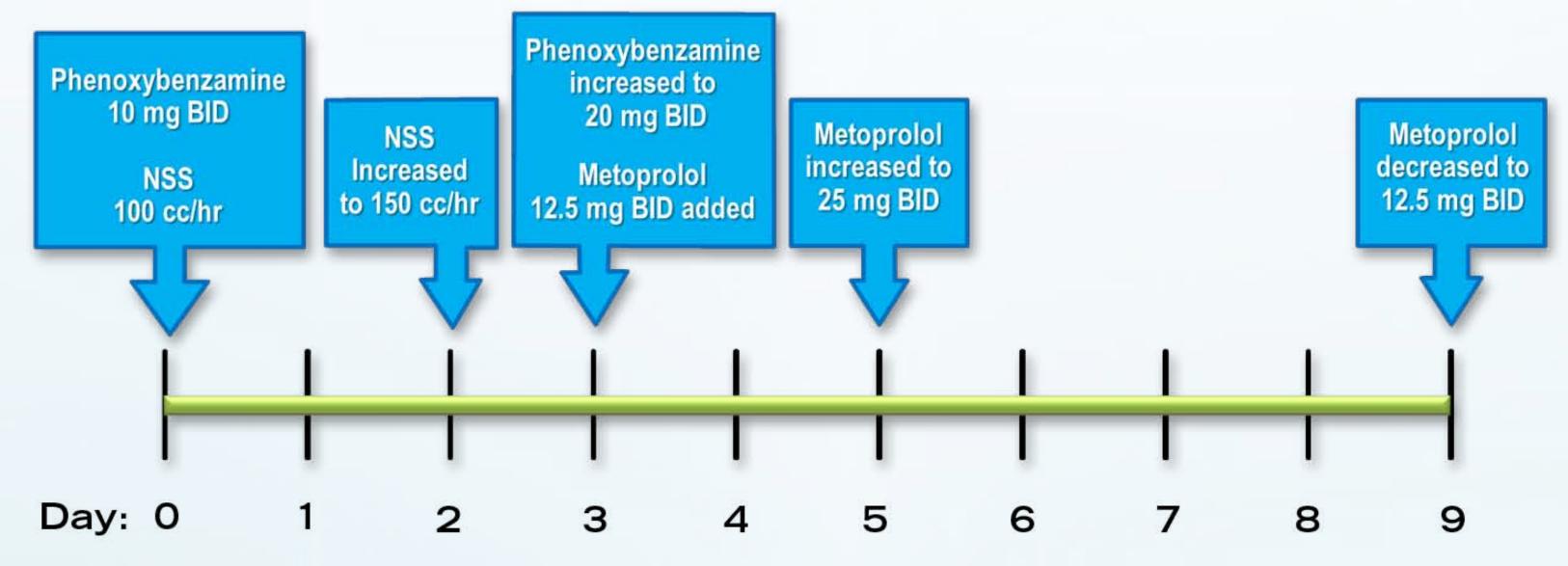


Image 3.

Timeline of preoperative treatment before successful removal of a pheochromocytoma.

Discussion

Preoperative management of the pheochromocytoma patient is not well elucidated. This case describes a successful sequence of steps taken to ensure preoperative hemodynamic stability.

- If beta-blockers are used alone, unopposed alphamediated vasoconstriction will occur, thus exacerbating hypertension to dangerous levels. Alpha-blockade should occur first.
- Volume re-expansion is important. Since accelerated hypertension will cause pressure natriuresis, large volume re-expansion with normal saline solution was achieved. However, volume balance should be kept even.
- It is important to initiate beta-blockage to stabilize heart rate, which may lead to tachyarrhythmias if uncontrolled.
- Beta-blockade was then titrated down the day before surgery to avoid protracted hypotension as vasodilation would follow pheochromocytoma removal.
- With the concerted efforts of multiple teams including endocrinology, nephrology, critical care medicine, and surgery, the pheochromocytoma patient may be appropriately cared for preoperatively.

Reference:

Pacak, K. Approach to the patient: preoperative management of the pheochromocytoma patient. The Journal of Clinical Endocrinology and Metabolism, 2007. 92(11):4069-4079.

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