A Rare Case of an Exclusive Dopamine-Secreting Cardiac Pheochromocytoma

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Introduction: Most functional pheochromocytomas produce norepinephrine and/or epinephrine. Tumors that produce predominantly or exclusively dopamine and found involving the heart are exceedingly rare (1). We present a case of a dopamine-secreting paraganglioma to highlight distinguishing features and identify specific anesthetic challenges.

Case Presentation: A 65 year old male initially presented with conversational dyspnea and fatigue. There was no history of hypertension. CT/MRI imaging revealed a 6 x 7 x 3 cm intrapericardial mass infiltrating the main pulmonary artery and right ventricle causing mild narrowing of the right ventricular outflow tract. TTE findings were unremarkable. Biopsy of an enlarged AP window lymph node was positive for malignant paraganglioma. Plasma catecholamine levels showed excess dopamine secretion but normal levels of norepinephrine and epinephrine. Prior to scheduled surgical resection, alcohol ablation and coiling of feeder vessels to the tumor was performed, followed by multiple rounds of chemotherapy, with some reduction in tumor size and dopamine levels. Preoperative alpha-blockade was not administered. Following placement of a brachial arterial line and standard ASA monitors, anesthesia was induced with midazolam, fentanyl, propofol, and rocuronium. A transient hypertensive episode without tachycardia was noted prior to cannulation and was treated with large boluses of nitroglycerin and nitroprusside. Upon mobilization of the tumor, after initiation of cardiopulmonary bypass, it was determined to be unresectable. The post-CPB course was uneventful, without need vasodilator or vasopressor therapy. The patient was extubated later that day and discharged from the hospital on POD #5.

Discussion: Pheochromocytomas, most commonly found in the adrenal glands, are rare tumors derived from chromaffin cells. Approximately 1% of pheochromocytomas are located within the thoracic cavity, with only about 70 reported cases in the literature involving the heart (2). Tumors that produce predominantly or exclusively dopamine are yet more rare. The preferential excretion of dopamine over norepinephrine/epinephrine is due to the reduced activity of dopamine-beta-hydroxylase which converts dopamine to norepinephrine (3). As a result, the clinical presentation is often atypical and patients are usually normotensive, causing the diagnosis often to be delayed until the tumor becomes large enough to produce a mass effect. Dopamine-secreting tumors also tend to have higher malignancy and recurrence rates (1). En bloc surgical resection is the definitive treatment. Unlike most pheochromocytoma resections, preoperative alpha-blockade is not routinely recommended for predominantly dopamine-secreting tumors due to its association with potential cardiovascular collapse. This may be due to an unopposed vasodilatory action of dopamine when vasopressor catecholamines are blocked (3). This may also be further supported by the tendency towards elevated blood pressure after these tumors are resected. While normotension is usually seen, perioperative hemodynamic instability is not uncommon and should still be anticipated (1).

References:
1) Singapore Med J 2010; 51(5)
2) J Anesth 2007; 21:504-506
3) South Med J 2003; 96(9)