

PERIOPERATIVE HEMODYNAMIC CRISIS IN UNDIAGNOSED PHEOCHROMOCYTOMA PATIENT

- Undergoing Incidental Surgery -

ANIS BARAKA*

The classic symptomatic triad of pheochromocytoma consists of headache, palpitation and excessive sweating. However, pheochromocytoma may go unrecognized, when induction of anesthesia and surgery may precipitate catastrophic hemodynamic crisis and even multiple organ failure in pheochromocytoma patients undergoing incidental surgery. In this situation, mortality is close to 80%.

The factors triggering intraoperative pheochromocytoma crisis can be attributed to excessive release of catecholamines from the undiagnosed tumor secondary to anxiety of the awake patient, or secondary to light general anesthesia during surgery.

Also, it may be drug-induced due to histamine release, dopamine receptor blockade, or sympathomimetic action. In addition, excessive release of catecholamines from the pheochromocytoma may be attributed to mechanical factors such as squeeze of the tumor during straining, positioning of the patient, by scrubbing, by intraperitoneal carbon dioxide insufflations during laparoscopy, or by direct manipulation of the tumor.

In the pregnant patient, having pheochromocytoma, excessive uterine contractions or fetal movements, as well as normal vaginal delivery or Cesarean section may precipitate the crisis; the symptoms and signs may mimic that of severe preeclampsia. However, preeclampsia is associated with hypertension and proteinuria usually after the 20th week of gestation, while pheochromocytoma is rarely associated with proteinuria and may cause hypertension throughout pregnancy.

The perioperative pheochromocytoma crisis may mimic other conditions such as thyroid storm or malignant hyperthermia. Dramatic changes in heart rate and/or blood pressure should alert the anesthesiologist to consider pheochromocytoma crisis as a possible cause. However, circulatory shock and pulmonary oedema may be the first manifestation of undiagnosed pheochromocytoma. Echocardiography revealed global cardiac hypokinesia secondary to cardiomyopathy which is seen in 25% to 50% of pheochromocytomas as a result of sustained exposure of the myocardium to high levels of catecholamines.

Multiple organ failure (MOF) may be the initial presentation and is called pheochromocytoma multisystem crisis. MOF may result from the high levels of circulating catecholamines, which can trigger excessive vascular spasm, volume contraction, platelet aggregation and thrombosis. The splanchnic vessels are highly susceptible to catecholamine-induced vasoconstriction, and, hence, the ischemic gut mucosa may allow bacterial translocation or the passage of endotoxins across the intestinal barrier to extra intestinal sites including the lung which is the first organ to fail. Also, acute renal failure can be attributed to acute tubular necrosis because of the combination

* MD, FRCA (Hon). Emeritus Professor of Anesthesiology, American University of Beirut, Lebanon.

of cardiogenic shock reducing the renal perfusion, associated with renal vasoconstriction induced by a surge of catecholamines.

Management of intraoperative pheochromocytoma crisis consists of elimination of the triggering factors, as well as controlling hypertension by the administration of short-acting vasodilators such as the alpha-adrenergic blocker phentolamine, or sodium nitroprusside infusion. Whenever hypertension is associated with severe tachycardia, beta-adrenergic blocker can be administered; the short-acting and selective beta₁ blocker esmolol is preferred to the long-acting and nonselective propranolol.

The administration of beta adrenergic blocker without prior α -adrenergic blockade may be complicated by cardiac failure and pulmonary edema secondary to its negative inotropic effect associated with an increased after load. Nonselective beta blockade leads to loss of beta₂ receptor-mediated vasodilation, leaving the unopposed effects of alpha receptors to cause vasoconstriction, resulting in

increased afterload, myocardial dysfunction and pulmonary oedema. Nonselective beta blockers should be avoided in any patient who could conceivably have a pheochromocytoma,

When the pheochromocytoma is surgically accessible during incidental surgery as laparotomy, the surgeon may be tempted to excise the tumor. However, tumor handling may result in dramatic increases in arterial blood pressure followed by intractable hypotension after tumor excision. A safer option is planned resection of the pheochromocytoma after confirmation of the diagnosis and optimal preoperative pharmacologic preparation. However, in patients with undiagnosed pheochromocytoma urgent adrenalectomy is recommended whenever multisystem injury deteriorates despite maximal medical therapy.

In conclusion, patients with undiagnosed pheochromocytoma undergoing incidental surgery may develop intraoperative hemodynamic crisis complicated by postoperative multiple organ failure. In this situations, mortality is close to 80%.

References

1. SIDDIK-SAYYID SM, DABBOUS AS, SHABAAN JA, DAABOUL DG, BARAKA AS: Catastrophic cardiac hypokinesia and multiple organ failure after surgery in a patient with an undiagnosed pheochromocytoma emergency excision of the tumor. *Journal of Cardiothoracic and Vascular Anesthesia*, vol 21, no 6, 2007, pp. 863-866.
2. DABBOUS A, SIDDIK-SAYYID S, BARAKA A: Catastrophic hemodynamic changes in a patient with undiagnosed pheochromocytoma undergoing abdominal hysterectomy. *Anesthesia and Analgesia*; 2007, 104:223-224.
3. BRAVO EL, GIFFORD RW: Current concepts/ Pheochromocytoma: diagnosis localization and management. *N Engl J Med*; 1984, 311:1298-1303.
4. O. RIORDAN JA: Pheochromocytomas and anesthesia. *Int Anesthesiol Clin*; 1997, 35:99-127.
Newell KA, Prinz RA, Pickleman J, et al: Pheochromocytoma multisystem crisis: A surgical emergency. *Arch Surg* 123:956-959,1988.
5. FUKUSHIMA R, KOBAYASHI S, OKINAGA K: Bacterial translocation in multiple organ failure. *Nippon Geka Gakkai Zasshi* 99:497-503, 1998.
6. GILSANZ FG, LUENGO C, CONEJERO P, et al: Cardiomyopathy and pheochromocytoma. *Anaesthesia*; 1983, 38:888-891.
7. DUGAS G, FULLER J, SINGH S, WATSON J: Pheochromocytoma and pregnancy: a case report and review of anesthetic management. *Can J Anesth*; 2004, 134-138.
8. GOLDSHEVSKY JR, KAREL K, TEALE G: Pheochromocytoma causing acute pulmonary oedema during emergency caesarean section. *Anaesth Intensive Care*; 2007, 35:423-427.
9. SIBAL L, JAVANOVIE A, AGARWALT SC, PEASTON R, ET AL: Pheochromocytoma presenting as acute crisis after beta blockade therapy. *Clinical endocrinology*; 2006, 65:186-190.
10. SLOAND EM, THOMPSON BT: Propranolol induced pulmonary oedema and shock in a patient with pheochromocytoma. *Archives of Internal Medicine*; 1984, 144:173-174.