**Título:** Unexpected intra-operative hypertension in patient with Von Hippel Lindau disease: what else? A case report

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**Área Terapêutica/Tema:** Segurança dos Doentes (Patient Safety)

**Resumo:**

Background: Von Hippel Lindau (VHL) syndrome is a rare autosomal dominant disease characterized by the presence of hemangioblastomas, renal cell carcinoma and neuroendocrine tumors.

Case report: We report a case of a 52-year-old woman suffering from VHL disease proposed for an open partial nephrectomy due to a renal cell carcinoma. She had history of retinal, intracranial and intramedullary hemangioblastomas and adrenal pheocromocytoma surgically removed. Although no complications throughout pre-operative period and anesthesia induction were reported, during the procedure there were several moments of unexpected systolic and diastolic hypertension (Fig.1). The first on the surgical incision and moreover as the peri-renal tissue was dissected. The possibility of an undiagnosed pheochromocytoma was suggested to the surgical team, thus the procedure continued avoiding adrenal gland manipulation and no anti-hypertensive medication was needed. The presence of a mass reported during surgery and on post-operative image supported the diagnose.

Discussion: Pheochromocytoma is a tumor of chromaffin cells characterized by the production of catecholamines. Up to one third are inherited in syndromes. Those associated with VHL disease have some singularities. Unlike others these are bilateral in half of cases and rarely metastize. They produce almost exclusively noradrenaline, which explains why they are asymptomatic, causing sustained systolic and diastolic hypertension and are less associated with tachycardia. In a series of incidental intraoperative catecholamine-producing tumors most hemodynamic instability was related to mass manipulation and during induction. The peri-operative mortality associated with these tumors was 8%, however a under reporting bias of poor prognosis cases was pondered.1 Other comorbidities comprise cardiac failure, pulmonary edema, arrythmias and myocardial ischemia. It should be emphasized that some drugs can indirectly spur a catecholaminergic crisis in the presence of pheochromocytoma.

Learning points: Patients with undiagnosed neuroendocrine tumors may be a challenge to the intraoperative care. Pheocromocytoma must be considered in patients with abnormal BP behavior, particularly in VHL disease. Early invasive monitoring should be pondered.

Reference: J Clin Anesth. 2009;21(3):220–9; J Anaesthesiol Clin Pharmacol. 2013 Jan

Fig  1: Vital  signs trends during intra-operative period. Bottom symbols represent beginning and end of surgery, respectively. Noninvasive blood pressure (dark red), invasive arterial pressure (light red); heart rate (black), peripheral oxygen saturation (green), end tidal carbon dioxide (blue).



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