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Letter to Editor

Anesthetic management of a challenging case of pheochromocytoma with dilated cardiomyopathy

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Pheochromocytoma is a catecholamine secreting tumor of chromaffin cells, found in the medullary portion of adrenal gland.Excessive secretion of catecholamines causes symptoms like palpitations, sweating, headache, tremors, weight loss and constipation. Pheochromocytoma is the cause behind hypertension in about 0.05-0.2% of the total patients and labile hypertension is its commonest manifestation.[1] Complications of pheochromocytoma are usually due to high blood pressure and consequent sequelae like heart diseases, stroke and kidney failure. Surgical resection of the tumour is the treatment of choice and this usually corrects the hypertension. Careful preoperative treatment with alpha and beta blockers is required to control the blood pressure and prevent intraoperative hypertensive crisis.[2]

A 24-year-old female patient presented with a history of pain in the left side of abdomen, sweating and palpitations for one month. There was associated loss of appetite, constipation and history of weight loss. On examination her vitals were; heart rate (HR):140/min, blood pressure (BP):190/140 mm of hg, respiratory rate (RR): 14-16/min and SpO_2: 98%. Her laboratory investigations revealed high levels of fractionated metanephrines [2944 ug (normal value: 74-297)] and nor-metanephrines [>10000 ug (normal value: 73-808)] in 24-hour urine sample. Ga-PET-CT scanning confirmed the presence of an 11.8 x 9.1 x 9.7 cm somatostatin receptor expressing left sided adrenal mass. Patient's blood sugar levels (RBS) was persistently high with values ranging from 250-350 mg/dl. Her echocardiography showed moderate dilated cardiomyopathy (DCM) with severe diastolic dysfunction with left ventricular ejection fraction of 30-35%. Other investigations were found to be within normal limits. Patient was showing high fluctuations in BP (156/120 mm hg- 190/130 mm hg in supine position and 138/96 mm hg- 170/110 mm hg in standing position) and HR (110-150/min), and thus, was started on tab. prazosin, tab. metoprolol and other medications. Inj. Insulin-R and Insulin glargine were also started to achieve the glycemic control. The medications were up-dosed over next 3-weeks for optimization of BP and HR. Patient was shifted to ICU and put on dexmedetomidine infusion 12 hours before the surgery.

Her pre-operative opening vitals were; HR-104/min, BP-116/76mmhg and SpO_2-99%. Peripheral venous access was insured with 16-G and 18-G cannula. Central venous cannulation was done in the left internal jugular vein for continuous central venous pressure monitoring and administration of drugs. Left radial artery cannulation was done for beat-to-beat measurement of BP and epidural catheter was inserted in the T8-T9 position for the management of pain. Pre-medication was done with Inj. midazolam (1.5 mg), Inj. fentanyl (150 ug), continuous dexmedetomidine infusion (@ 0.5 ug/kg/hr) and

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a combination of paracetamol (1gm), magnesium (2gm) and lignocaine (60 mg) infusion. Induction was done with i. v. inj. etomidate (12 mg) and inj. vecuronium (8 mg). Patient's vitals remained stable during the process of intubation. Maintenance of anesthesia was achieved with O_2:N_2O: isoflurane in the ratio of 33%:66%:0.7-1.2%, and vecuronium infusion.

Patient was showing wide fluctuations in the BP before ligation of adrenal vein. Infusion of sodium nitroprusside (0.5 to 2 ug/kg/min), esmolol (25–50 μg/kg/min) and dexmedetomidine (0.5ug/kg/min) was started to maintain the haemo-dynamics. After tumour resection and ligation of adrenal vein, there was sudden fall in the blood pressure of the patient. Mean arterial pressure (MAP) was maintained with the help of fluid resuscitation and use of vasopressors like nor-adrenaline (0.2- 1 u/kg) and vasopressin (0.01-0.07u/min). Dobutamine (2.5 to 5.0 ug/kg/min) infusion was also started with the aim to keep MAP at least 65 mm hg. Patient was shifted to ICU where she could be weaned off from vasopressors after 24 hours and was extubated thereafter.

Major peri-operative challenges in this case were the stress of intubation, massive surge of catecholamines during handling of the tumour which may precipitate hypertensive crisis and worsen the cardiac function, and profound hypotension with shock which occurs after tumour resection because of sudden decrease in catecholamines. These patients require optimization of blood pressure and heart rate before undergoing surgical resection with medication like alpha blockers, beta blockers, calcium channel blockers for at least 2-3 weeks. Our patient was also given above drugs for 3 weeks for optimization but her haemo-dynamics were still not under much control due to high secretory nature of the tumor, as was depicted from very high urine nor-metanephrine levels. Addition of dexmedetomidine one day before surgery helped us in achieving the haemo-dynamic control in this patient. Hegde et al also used dexmedetomidine in a similar patient of pheochromocytoma with poor left ventricular function and continued the infusion peri-operatively.[3] Pre-induction combination infusion of paracetamol, lignocaine and magnesium was also effective in preventing the stress response to intubation and provide better cardiac stability.[4] There is down-regulation of alpha and beta receptors in patients of pheochromocytoma because of chronically elevated levels of catecholamines. This poses a considerably high risk of hypotension after pheochromocytoma removal and thus, higher than normal values of vasopressors are needed to maintain BP.[5] Our patient also had refractory hypotension after resection of the tumour. The MAP was maintained with the fluid loading and addition of vasopressors like nor-adrenaline, vasopressin and dopamine. Additional challenge in our patient was the presence of dilated cardiomyopathy with poor left ventricular function. As there is limited cardiac reserve, there are more chances of cardiac complications like arrhythmia and cardiac failure peri-operatively. Therefore, care must be taken to maintain normovolemia and avoid medications which can cause myocardial depression. The presence of dilated cardiomyopathy in our patient might also be the reason for higher than usual requirement of vasopressors.

Anaesthetic management of surgical resection of pheochromocytoma is quite challenging because of continuous haemodynamic instability both pre-operatively and intra-operatively. Complications like DCM further add to the risk of peri-operative mortality. Dexmedetomidine can serve as an important weapon in the armamentarium to prevent intraoperative hypertensive crises. A planned approach may serve to deal with intra-operative and post-operative challenges.

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