Perioperative management of combined surgery for phaeochromocytoma and double outlet right ventricle: A rare combination

INTRODUCTION

Anaesthetic management of phaeochromocytoma always remains a challenge.[1] This becomes more when congenital heart disease is associated with phaeochromocytoma.[2,3] We report the perioperative anaesthetic management of a patient of phaeochromocytoma with double outlet right ventricle (DORV) who underwent combined phaeochromocytoma excision and corrective cardiac surgery.

CASE REPORT

A 25-year-old, 52 kg man presented with a history of abdominal pain since 1 year. He also complained of recurrent episodes of sweating since 7 years. His heart rate (HR) was 112/min, blood pressure (BP) 184/110 mmHg and oxygen saturation (SpO_2) 82% on room air. Ultrasonography abdomen showed a nodular retroperitoneal mass of 5 cm × 4.5 cm size posterior to the pancreatic head. Contrast-enhanced computerised tomography (CT) showed 5 cm × 4.5 cm right retroperitoneal mass in suprarenal location displacing the inferior vena cava (IVC) and renal veins. Metaiodobenzylguanidine scan revealed increased uptake of radiotracer in the right suprarenal area and 18 F-fluoro-l-dihydroxyphenylalaninepositron emission tomography scan revealed two phaeochromocytomas [Figure 1]. Total urinary catecholamine was 115 μg/24 h (normal 14–110 μg/24 h) and plasma normetanephrine was 1.4 nmol/L (normal 0.01–0.89 nmol/L) and plasma metanephrine was 1.3 nmol/L (normal 0.01–0.49 nmol/L). Patient was started with prazosin 0.5 mg orally twice daily which was gradually increased to 6 hourly doses. Subsequently, metoprolol 25 mg orally once daily was added. BP settled down in the range of 120/70–130/80 mmHg.

He was a diagnosed case of cyanotic congenital heart disease with DORV, ventricular septal defect (VSD) and pulmonary stenosis (PS). Patient was having progressive cyanosis, increased hypoxemia on exertion

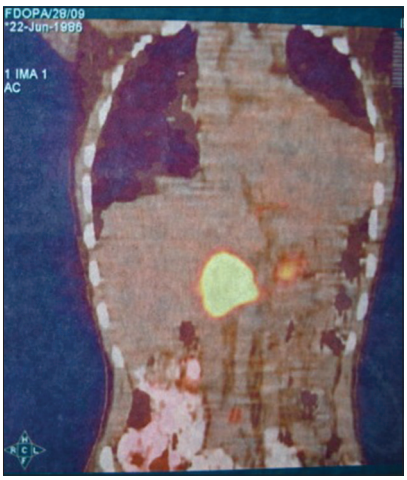

Figure 1: 18F-fluoro-l-dihydroxyphenylalanine positron emission tomography scan showing the two phaechromocytoma at right suprarenal region with increased uptake

and reduced exercise tolerance with heart failure and left ventricular ejection fraction of 25%.

Patient was scheduled for excision of tumour and correction of cardiac defects under cardiopulmonary bypass (CPB) in single surgical setting. Prazosin and metoprolol were continued. He was pre-medicated with alprazolam 0.25 mg and morphine 5 mg and promethazine 25 mg intramuscular injection 1 h before surgery. In the operating room, baseline HR was 110/min, BP 130/86 mmHg and SpO_2 85%. Patient was induced with fentanyl 200 μg, midazolam 1 mg, etomidate 14 mg and intubated with rocuronium 50 mg. Anaesthesia was maintained with oxygen in air, isoflurane and boluses of fentanyl, midazolam and vecuronium. Monitoring included routine monitors (electrocardiogram, pulse oximetry, non-invasive blood pressure) invasive BP, central venous pressure, bispectral index (BIS^R) and transesophageal echocardiography. Suprasternal notch to suprapubic midline incision was used. Hypertension (BP > 140 mmHg systolic) and tachycardia (HR > 100/m) were controlled with titration of anaesthetic depth, boluses of 2–5 mg esmolol and sodium nitroprusside 0.5–2 μg/kg/m infusion. Magnesium 2 g and lignocaine 100 mg were administered over 30 min from a Burette set to control arrhythmias during tumour manipulation. Two encapsulated right suprarenal masses between left renal vein and IVC were removed. After ligation of the right suprarenal vein, BP dropped to 80/34 mmHg, which was managed with discontinuation of nitroprusside, Trendelenburg position, fluid administration and noradrenaline at 0.1 μg/kg/m infusion. After tumour resection, intra-cardiac repairs in the form of VSD closure with routing for both ventricular outflow tract and resection with patch augmentation of right ventricle (RV) outflow tract were performed under CPB. On release of the aortic cross clamp, patient regained normal sinus rhythm with poor ventricular contraction. Adrenaline 0.1 μg/kg/m infusion was started, and CPB was terminated. Nitroprusside 0.5 μg/kg/m and noradrenaline 0.1 μg/kg/m infusion were continued to achieve BP of 110–130 mmHg systolic. Patient was shifted to the intensive care unit (ICU) and extubated after 24 h.

In the ICU, patient remained stable, and inotropes were gradually tapered. Post-operative analgesia consisted of fentanyl, tramadol and ketorolac. On 3rd day, patient was shifted to ward and discharged from the hospital on 7th day. Histopathology report confirmed the tumour as phaeochromocytoma.

DISCUSSION

Double outlet right ventricle is a rare congenital disease with an incidence of 0.09/1000 live births.[4] Whereas phaeochromocytoma is seen in 1 in 1000 people with hypertension.[5] The combination of these two diseases is extremely rare. In DORV, both great arteries arise from the morphologic RV and VSD represents the only outlet from the left ventricle. Classic symptoms of phaeochromocytoma are headache, palpitations, sweating and hypertension. Phaeochromocytoma was incidentally detected in our patient by abdominal ultrasonography and CT while investigating abdominal pain.

Our main concern was the risk of uncontrolled vasodilatation with decreased venous return and decreased systemic vascular resistance (SVR) resulting in shunting of blood from right to left ventricle causing further desaturation from pre-operative use of prazosin and metoprolol. Hence, SVR was maintained with titration of anaesthetics, vasodilators and adequate blood volume, as increased SVR might increase pulmonary blood flow and improve SpO_2.

The catecholamine surge from phaeochromocytoma might increase the degree of arterial hypoxemia and cyanotic spell by infundibular spasm.[2] Development of atrial and supraventricular arrhythmias are high after DORV repair. Inadequately treated and non-operated phaeochromocytoma would create more hemodynamic alterations with arrhythmias in cardiac patients.[2,3] Bleeding would be more with hypertension induced by catecholamines. Induction of anaesthesia was opioid-based to minimise negative inotropic effects as these patients were at increased risk for ventricular dysfunction due to long-standing cyanotic heart disease and catecholamine-induced cardiomyopathy. Within seconds of ligation of venous drainage to phaeochromocytoma, the high concentration of circulating norepinephrine falls. The same situation happened in our patient with fall in BP to 80 mmHg. Drastic fall in BP may be avoided by prior adequate volume replacement. Infusion of vasopressor like phenylephrine or norepinephrine may be started as a stop gap measure until the adequate volume is replaced.[2,3]

Combined surgery for phaeochromocytoma and DORV may reduce the morbidities related to the two surgeries and length of hospital stay. Surgery of only

phaeochromocytoma might have led to poor outcome with LVEF 25%, right to left intra-cardiac shunt and hypoxic cardiac arrest from infundibular spasm at RV outflow. Surgery of DORV and PS without resection of phaeochromocytoma might not be wiser with a chance of catecholamine surge in the perioperative period.

CONCLUSION

The present case management demonstrates the feasibility of a combined procedure for phaeochromocytoma and DORV. We recommend tumour resection to precede intra-cardiac repair. The perioperative anaesthetic management has to be planned with balanced use of alpha-blockers, beta-blockers, magnesium, vasodilators, vasoconstrictors and adequate blood volume loading, to counter the haemodynamic fluctuations.

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