Case Report

Anaesthetic management of a patient with Cushing's syndrome and non-compaction cardiomyopathy for adrenal tumour resection

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ABSTRACT

We describe the anaesthetic management of adrenalectomy in a patient with Cushing's syndrome due to adrenal mass with coexisting non-compaction cardiomyopathy. The problems due to hypersecretion of cortisol in Cushing's syndrome were compounded by the association of a rare form of genetic cardiomyopathy with very few guidelines regarding the perioperative management. The knowledge about the pathophysiological changes, clinical presentation and complications in non-compaction cardiomyopathy is essential for planning the anaesthetic care, and the aim of this presentation is to highlight the issues crucial for management of such challenging patients.

Key words: Cushing's syndrome, non-compaction cardiomyopathy, perioperative care

INTRODUCTION

Cushing's syndrome is characterized by excessive exposure to glucocorticoids, resulting in significant morbidity and mortality when left untreated. The removal of adrenal gland and resection of pituitary adenomas are the options available for the treatment of Cushing's syndrome.[1,2] The problems related to airway management, electrolyte imbalance and cardiovascular complications in Cushing's syndrome need meticulous assessment and planning of anaesthetic care.[3-5] Non-compaction cardiomyopathy is a rare and distinct clinical entity, with multiple diagnostic criteria, few guidelines and scientific data regarding the perioperative management.[6,7] Patients with non-compaction cardiomyopathy present with symptoms of heart failure, arrhythmias and thromboembolism.[6] The perioperative management of these patients involves formulation of a safe anaesthetic plan and strategies for the prevention and treatment of expected untoward events based on the clinical features and complications present in non-compaction cardiomyopathy and Cushing's syndrome.

CASE HISTORY

A 42-year-old, 70 kg male patient was scheduled to undergo radical open adrenalectomy for a right-sided adrenal mass. He presented with history of increased weight gain and facial swelling of 3 months duration. The patient also had symptoms of exertional dyspnoea NYHA class III, palpitation and fatiguability of 6 months duration with effort tolerance of <4 METS. He was diagnosed as cardiomyopathy with heart block in a private hospital and received medical treatment with T. Envas 2.5 mg bd, T. Spironolactone 25 mg od and T. Amiodarone 200 mg bd. Pre-operative cardiac evaluation was done in our centre. ECG showed second degree AV block with ventricular ectopics. Echocardiography revealed global hypokinesia of the ventricle with EF-42%, trabeculations in the left ventricle, non-compaction of LV, structurally normal

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valves and frequent venticular ectopics noted during the study. The cardiologist opined as non-compaction cardiomyopathy, with increased risk for arrhythmias in the perioperative period, and advised continuation of the cardiac drugs.

CT KUB showed right-sided adrenal mass. Dexamethasone suppression test revealed ACTH-independent Cushing's syndrome with S. cortisol of 63.70 mcg/dL. The blood routine and the biochemical parameters were unremarkable, except serum potassium value of 3.0 mmol/L. The coagulation profile, which included prothrombin time, partial thromboplastin time, international normalised ratio, clotting and bleeding time, were within normal values. The chest X-ray showed cardiomegaly with normal lung fields.

On examination, the pulse rate was 78/min, with missed beats every 4th beat, and the blood pressure was 140/90 mmHg. Airway examination revealed mouth opening of 4 cm, Mallampatti grade III, thyromental distance of 5 cm and cushingoid facies with normal neck movements. The breath holding time was 12 s. Examination of the other systems was unremarkable. The cardiac status was stabilised with medications and potassium supplements prior to surgery. He was explained about the anaesthetic plan and accepted for the procedure under ASA class III. Intravenous potassium chloride 60 meq in 0.9% normal saline was given overnight and the serum potassium after correction was 2.9 mmol/L. The patient received the morning dose of T. Amiodarone 200 mg and T. Ranitidine 150 mg on the day of surgery.

Temporary pacemaker was inserted with a preset rate of 70/min in the cardiac cath lab prior to induction. The defibrillator, pre-loaded infusion of Dopamine, Dobutamine and injection Amiodarone, was kept ready in the operating room. The monitors included electrocardiogram, pulse oximetry, capnography and invasive blood pressures through a 20 G cannula in the radial artery. Under ultrasound guidance, the right internal jugular vein was cannulated with a 7 F triple-lumen catheter and central venous pressure monitoring instituted. An epidural catheter was placed at the L1, L 2 space in sitting position for post-operative analgesia. Inj. KCl 40 meq in 200 mL of normal saline was given prior to induction as the baseline arterial blood gas potassium was 2.5 mmol/L. Anaesthesia was induced with fentanyl 2 mcg/kg, propofol 2 mg/kg in graded doses, followed by succinylcholine 2 mg kg for endotracheal tube placement. Anaesthesia was maintained with sevoflurane 1–2% in 33% oxygen–nitrous oxide and intermittent doses of vecuronium. Bolus doses of fentanyl 50 mcg were given to a total of 250 mcg in the intraoperative period. The surgical removal of mass was done in the left lateral position. The saturation dropped to 90% 30 min from the surgical incision time. At the time of desaturation, the heart rate was 88/min, BP was 104/70 mmHg and central venous pressure was 18 mmHg, with increased airway pressures in the ventilator and decreased compliance of the reservoir bag on manual ventilation. Inj. Frusemide 20 mg was given intravenously and ventilation was continued with 100% oxygen and positive end expiratory pressure of 10 cm of water, which improved the saturation to 99%. There were no further episodes of desaturation. Intravenous potassium replacement was done with Inj. KCl 20 meq in 100 mL of NS to a total of 100 mq over 4 h, titrated to measured K^+ levels in arterial blood gas analysis [Table 1], The ventricular ectopics resolved completely after potassium supplementation. The blood loss of 1500 mL was replaced with 2 units of packed red cells. Intraoperatively, 3000 mL of crystalloids and 500 mL of tetrastarch were infused and the urine output was 1200 mL. The central venous pressure was maintained at 12–15 mmHg. The duration of the surgical procedure was 5 h with stable haemodynamics, except for one episode of desaturation. Following surgical resection and haemostasis, the patient was transferred to the critical care unit and extubated at 24 h. The patient received continuous epidural infusion of bupivacaine with fentanyl and paracetamol infusion for post-operative analgesia. Enoxaparin 60 mg once daily was started from the first post-operative day. Intravenous KCl supplementation 40 meq/day continued for 48 h till the potassium value was more than 3.5 mmol/L. The patient developed atrial fibrillation with fast ventricular response on the third post-operative day, which reverted to sinus rhythm with amiodarone bolus, and the infusion was continued for 24 h. The patient was transferred to the ward and discharged home 10 days after surgery with an uneventful post-operative course. The steroids were not supplemented in the immediate post-operative phase as unilateral resection of the tumour was only done and the patient is under regular follow-up of the primary endocrinologist.

DISCUSSION

Non-compaction of the left ventricle has been considered as a distinct form of genetic

Table 1: Arterial blood gas values at different time intervals in the intraoperative period

Time	Pre-induction	30 m	1 h	1 h 30 min	2 h	3 h	4 h	5 h
pH	7.5	7.5	7.54	7.49	7.44	7.45	7.4	7.4
PCO_2 (mmHg)	36.4	35.0	35.8	38.5	40.9	36.0	33.2	34.2
PO_2 (mmHg)	90	111	77.1	145	173	193	216	213
Na^+ mmol/L	136	135	132	133	134	135	135	136
K^+ mmol/L	2.5	2.9	3.0	3.3	3.4	2.4	3.9	3.4
Ca+ mmol/L	96	102	97	97	100	107	105	109
HCO_3^- mmol/L	31	28	30	29	27.4	22.9	24	20.6
FiO_2	0.21	0.33	1	0.33	0.33	0.33	0.33	0.33

cardiomyopathy by the American Heart Association.[6] It is characterized by the presence of prominent trabeculations on the luminal surface of the ventricle associated with deep intertrabecular recesses that extend into the ventricular wall. The diagnosis is based on the morphological features of the left ventricle in Echocardiogram, colour Doppler and magnetic resonance imaging, with multiple diagnostic criteria.[8,9] Hence, the true prevalence rate is unknown and there is no specific therapy for this type of cardiomyopathy. They are more common in males, and present with symptoms related to systolic ventricular dysfunction, arrhythmias and thromboembolism.[10] The rarity of this condition with poor prognosis has limited case reports and perioperative guidelines in the literature.[11] Our patient also presented with symptoms of heart failure and arrhythmias initially, for which he underwent treatment, and the adrenal tumour was identified subsequently on evaluation for the weight gain. Hence, the anaesthetic implication in this case was centred towards management and prevention of fatal arrhythmias, cardiac failure and embolism in the perioperative period.

Arrhythmias, which include ventricular tachyarrhythmia, supraventricular tachycardia, atrial fibrillation and complete heart block, have been reported in non-compaction. Sudden death has occurred in 50% of the adult patients with this condition.[6,12-14] Because our patient had pre-existing block with systolic dysfunction, temporary intravenous pacemaker was inserted prior to induction of anaesthesia. In our patient, there were no events of bradyarrhythmia in the intraoperative period that demanded activation of pacemaker. The external defibrillator and antiarrhythmic drugs for treatment of fatal arrhythmias were kept ready as a preparative strategy. The presence of hypokalaemia was an additional trigger in this patient for intraoperative arrhythmias. The high cortisol levels contributed significantly for the mineralocorticoid activity, resulting in resistant hypokalaemia that demanded continuous correction with IV potassium in the intra- and post-operative periods. The mineralocorticoid activity of cortisol is negligible due to conversion to cortisone by the action of 11 beta hydroxyl steroid dehydrogenase.[1,15] The high cortisol levels cause substrate saturation of this enzyme, manifesting the mineralocorticoid activity. This could be the reason for refractory hypokalaemia in our case, which resolved following resection of tumour.

The association of neuromuscular disorder in non-compaction limits use of succinylcholine for securing the airway.[7] Because our patient did not have associated neuromuscular disease, succinylcholine was preferred considering the low potassium levels and the anticipated difficult airway status. The problem of systolic ventricular dysfunction, as evidenced by increased central venous pressure, airway pressures and desaturation, was managed at an earlier stage itself with diuretics and positive-pressure ventilation with PEEP. The use of epidural analgesia was avoided in the intraoperative period to prevent hypotension, which can have serious consequences, due to systolic dysfunction and ventricular arrhythmias that were present in our patient.

The incidence of new post-operative arrhythmias following open cardiac and non-cardiac surgeries in non-compaction is 21% and 3%, respectively.[16] This patient developed new-onset atrial fibrillation 24 h post-operatively, which was managed with pharmacological measures effectively, emphasizing the need for cardiac monitoring in the post-operative period. As the incidence of embolism is related to the severity of systolic dysfunction and newer arrhythmias in the perioperative period, the patient received low molecular weight heparin to prevent systemic embolism. The availability of improved imaging techniques has resulted in diagnosing this form of cardiomyopathy at an earlier stage itself, and knowledge about their clinical presentation and

complication is imperative for the anaesthesiologist for perioperative optimization and planning.

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