Case Report

Concomitant Takayasu arteritis and Cushing syndrome in a child undergoing open adrenalectomy: An anaesthetic challenge

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ABSTRACT

Takayasu's arteritis (TA) is a rare, chronic progressive panendarteritis involving the aorta and its main branches. Anaesthesia for patients with TA is complicated by their severe uncontrolled hypertension, end-organ dysfunction, stenosis of major blood vessels, and difficulties encountered in monitoring arterial blood pressure. In a patient with Cushing's syndrome (CS), the anaesthesiologist needs to deal with volume overload, hyperglycaemia, hypokalaemia, difficult airway and ventilation. Anaesthetic management of a patient with concomitant TA and CS undergoing adrenalectomy has hardly ever been reported. We present the successful anaesthetic management of a 15-year-old child with coexisting TA and CS undergoing open adrenalectomy.

Key words: Adrenalectomy, anaesthesia, Cushing's syndrome, Takayasu's arteritis

INTRODUCTION

Takayasu's arteritis (TA) is a progressive inflammatory vasculitis, potentially affecting all large and medium calibre arteries, leading to a varying degree of narrowing, occlusion or dilatation with specific predilection for young Asian women.[1] This disease is also referred to as pulseless disease, aortic arch syndrome, young female arteritis, idiopathic aortitis, and Martorell syndrome.[2,3] The major clinical finding is loss of palpable pulses in the upper limbs and neck. The unsuspected ischaemia in vital regional vascular beds may be associated with high anaesthetic risks for these patients. This case was particularly challenging because of the coexisting Cushing's syndrome (CS), which required the anaesthesiologist to be geared up to solve the additional problems of volume overload, hypertension, glucose intolerance and hypokalaemic metabolic alkalosis.

CASE REPORT

A 15-year-old male patient presented to our hospital 6 months before with a history of headache, swelling over face and body and episodic numbness and tingling in both upper limbs and clumsiness of the right hand while working for last 6 months. He had a history of uncontrolled hypertension and steroid intake for last 4 months. Magnetic resonance angiogram revealed narrowing of distal thoracic aorta above diaphragm, dissection above diaphragm extending below up to bifurcation, occlusion of left renal artery with distal reformation, subtle narrowing of right renal artery and occlusion of right subclavian artery with reformation of the brachial artery. Computed tomography (CT) angiography also supported the findings, and diagnosis of TA type V[4] with left adrenal mass(? myelolipoma) was established. Patient underwent digital subtraction

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angiography, and a stent was placed in proximal left renal artery 4 months back. A repeat angiography showed a good dilatation of the concerned artery. Since then patient was on clopidogrel along with antihypertensive medications.

During the present visit, the patient was scheduled for left open adrenalectomy. During pre-anaesthetic evaluation, patient showed features of Cushing's disease (CD). His weight and height were 35 kg and 137 cm respectively. Pre-operative blood pressure (BP) recorded on left upper limb was 150/92 mmHg and correlated well with lower limb recordings. BP was non-recordable on right arm as his right radial, brachial and axillary arterial pulsations were non-palpable. Both lower limb pulses were feeble. Both carotid pulsations were palpable, and bruit could be heard over right carotid artery. Chest radiography showed cardiomegaly and standard 12-lead electrocardiogram (ECG) showed sinus tachycardia and left axis deviation suggestive of left ventricular hypertrophy (LVH) with repolarization abnormality. Echocardiography revealed concentric LVH with grade I diastolic dysfunction. He was on nifedipine, enalapril and prazosin. Serum cortisol level (morning) was 577.0 nmol/L (normal 110-520 nmol/L) and was not suppressed with dexamethasone. Serum adrenocorticotropic hormone (ACTH) was <11.40 pg/ml (normal 4-80 pg/ml) and 24 h urinary cortisol was 461.03 mcg/day (normal range 20-100 mcg/day). His other laboratory and clinical parameters were within normal limits. There was no history suggestive of any focal neurological deficit or visual disturbance and no features suggesting cerebral involvement or ischaemia with head and neck extension. His airway was graded as Mallampatti class I.

Patient was pre-medicated with alprazolam 0.25 mg orally, and all anti-hypertensive medications were continued until the day of surgery. In the operating room, all routine monitors were attached, and peripheral venous access was established. A 20G epidural catheter was inserted at T_{10-11} interspace, and 12 ml of 0.25% bupivacaine with 50 mcg fentanyl was given as a bolus dose. Approximately, 10-12 min after the bolus dose, patient was induced with fentanyl 100 mcg, midazolam 1 mg and thiopentone 150 mg and trachea was intubated after giving vecuronium 5 mg. The lungs were ventilated with 50% oxygen in air and sevoflurane 1 minimum alveolar concentration. The anaesthesia was maintained with sevoflurane, propofol infusion and intermittent boluses of fentanyl and vecuronium as needed. Intravenous infusion of hydrocortisone was started pre-operatively at the rate of 5 mg/h and continued with tapering doses till second post-operative day. The left radial artery was selected for direct arterial pressure monitoring as the pulse was strong and was easily accessible. Right internal jugular vein was cannulated for central venous catheter placement. Intraoperative monitoring included ECG, pulse oximetry, body temperature, capnography, blood sugar level, blood gases, urine output and central venous pressure (CVP). Once arterial blood gas analysis was available, ventilation was adjusted to maintain normocarbia. Epidural infusion of bupivacaine 0.125% with fentanyl 2 mcg/ml was used for intraoperative and post-operative analgesia. Intra-operatively, CVP was maintained between 6 and 8 cm of water, and the patient remained haemodynamically stable. The surgery lasted 4 h and the blood loss was 300 ml. A total of 1.5 L of crystalloids and 500 ml of colloids (6% hydroxyl ethyl starch) were given intra-operatively. The patient was extubated at the end of the procedure. The entire perioperative course was uneventful. Patient was observed in high-dependency unit for 24 h and then shifted to the ward. He was discharged on the 8th post-operative day and referred to an endocrinologist for further care.

DISCUSSION

Takayasu's arteritis is characterised by a focal stenosis process preferentially involving the aorta and the proximal segments of its main branches. As per the new angiographic classification,[4] it is divided into six types (I, IIa, IIb, III, IV and V) and the involvement of coronary or pulmonary arteries is designated as C (+) or P (+), respectively.

Hypertension in TA is commonly renovascular, and TA is the most common cause of renovascular hypertension in India.[5] Hypertension in our patient may be related to CS. Besides calcium channel blocker and alpha-adrenergic blocker, our patient was on an angiotensin converting enzyme inhibitor, enalapril, which was started only after stenting of left renal artery.

Previous reports of the anaesthetic management of TA as well as that of CS have greatly emphasized the importance of adequate cardiovascular monitoring.[6] Fortunately, our patient had good pulses in his left arm for reliable monitoring of arterial BP. Meikle and

Milne[7] had suggested that monitoring of both upper and lower limb BP should be considered in order to have some measure of overall perfusion. In our case, since left upper limb pressures correlated very well with the lower limb pressures pre-operatively, monitoring of BP in the leg was not deemed necessary. We did not insert pulmonary artery catheter as there were no signs or symptoms of pulmonary hypertension. CVP measurement provided adequate information about cardiac preload in this patient.

An increase in BP in the perioperative period should be avoided. We used epidural blockade as part of the anaesthetic management to achieve this without recourse to potent intravenous vasodilator agents.

Hypertensive episodes during general anaesthesia can lead to cerebral haemorrhage and infarction in a patient with TA so cerebral perfusion pressure should be maintained and procedures that diminish carotid blood flow including the hyperextension of the head should be avoided.[8] No specialised neurological monitoring was used perioperatively. However, this patient recovered without any neurological deficits. There is only one report of post-operative cerebral infarction in a patient with TA.[9]

Cushing's syndrome has a multifactorial aetiology.[10] Apart from steroid supplementation (given in TA patients) other causes of CS are unilateral or bilateral adrenal hyperplasia, pituitary adenoma, CD and ectopic ACTH production. Our patient had unilateral adrenal hyperplasia with all the clinical features of CS. After testing the serum cortisol and ACTH, and the dexamethasone suppression test, patient underwent CT angiography and ultrasound examinations, which further confirmed the diagnosis.

It is well-known that patients with CS tend to suffer from volume overload, hypertension, glucose intolerance and hypokalaemic metabolic alkalosis. In our patient, anaesthesia induction was unremarkable, and there was no difficulty in airway management. Special attention was paid to patient positioning, in order to avoid fractures and/or skin damage. There were no significant changes in the electrolytes and blood glucose level, as well as in the acid-base status.

Perioperative steroid supplementation is needed in patients with CS and TA who are on steroids to prevent the occurrence of Addisonian hypotensive crises in such patients.[8] The steroid infusion was administered to the patient in the perioperative period in the present case.

Simeoli *et al.*[11] have reported two patients with concomitant TA and CD, in whom TA was exacerbated after normalisation of cortisol secretion following treatment. Similarly, there can be reactivation of TA activity after adrenalectomy due to the loss of the immunosuppressive effect of endogenous glucocorticoids and hence we should follow-up and monitor these patients for this possibility.

CONCLUSION

Patients with rare association of Takayasu's arteritis with the coexisting Cushing's syndrome can be encountered in daily practice. Careful pre-operative evaluation and proper intraoperative planning and management can lead to a fruitful outcome in these patients.

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