

Perioperative anaesthetic management of a child with cushing's syndrome for bilateral adrenalectomy

Sabina Yeasmeen^{1*}, Debasish Banik², Quamrul Huda², Sayeed Mahmud Ali Reza¹, Abdul Hye³, Anwar Ahmed⁴

¹Assistant Professor, Associate Professor, Professor, ³ Consultant, Department of Anaesthesia, Analgesia and Intensive Care Medicine, BSMMU, Intensive Care Medicine, BSMMU, Dhaka

* Corresponding author: e-mail: yeasmeens@yahoo.com

Abstract:

Nearly twenty five percent of the cases of cushing's syndrome are due to adrenal hyperplasia without an ACTH secreting tumour. Twenty percent of patients with endogenous cushing have adrenocortical tumour about half of which are benign adenoma. Surgical intervention done due to failed medical therapy and in case of adrenal adenoma. A child aged 4½ years, weighting 29kg with features suggestive of cushing's syndrome was admitted under paediatric surgery unit in Bangabandhu Sheikh Mujib Medical University. On investigation serum cortisol levels were raised. blood pressure was controlled by ACE inhibitor, calcium channel blocker and beta-blocker. He was scheduled for resection of adrenal cortical tumour. Electrolyte imbalance was corrected, steroid replacement was done. Patient was haemodynamically stable preoperatively. Surgery was completed uneventfully. Postoperatively patient was kept in ICU, ventilation maintained by control mode (CMV). After 24 hours the patient was extubated. When the patient found haemodynamically stable he was sent to the recovery room.

Key words: Cushing's syndrome, anaesthetic management, bilateral adrenalectomy.

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Introduction:

Cushing's syndrome is a clinical entity resulting from adrenocortical hyperfunction. The most common cause is iatrogenic resulting from administration of corticosteroids. Twenty five percent of the cases are due to adrenal hyperplasia without an ACTH secreting tumour.¹ Other causes of cushing's syndrome because bodies produce too much cortisol. Normally, the production of cortisol follows a precise chain of events. First, the hypothalamus, a part of the brain about the size of a small sugar cube, sends corticotrophin-releasing hormone (CRH) to the pituitary gland to secrete adrenocorticotropin hormone (ACTH), which stimulates the adrenal glands. When the adrenal gland receive the ACTH. They respond by releasing cortisol into blood stream.² Most causes of cushing's syndrome are not inherited. young adults develop small cortisol producing adrenals tumour with multiple endocrine neoplasm type I (MEN) hormone secreting tumours of the parathyroid glands. Pancreas and pituitary develops cushing's syndrome

due to pituitary ectopic or adrenal tumour.^{4,5} This case reports described the perioperative anaesthetic management of child who is undergoing bilateral adrenalectomy.

Case Report:

A 4½ years old male child presented with complaints of excessive weight gain for last 2½ years and increased appetite for same time was seen in picture 1.

On General Examination: Patient was obese, weighting 29kg, Body Mass Index (BMI) was- 35.8kg/m², moon shaped face, truncal obesity, buffalo hump, facial acene, hypertrichosis, pulse- 140/min, blood pressure 170/110 mmHg, respiratory and cardiovascular system was normal, per-abdominal striae were visible. Haematological and Biochemical investigation were normal, X-ray chest normal and ECG shows sinus tachycardia. X-ray skull- Sella turcica was normal. 2D Echocardiography revealed left ventricular ejection fraction (LVEF) 60%, CT brain – Mild cerebral

atrophy, 24 hr urinary cortisol was – 427 $\mu\text{g}/24\text{hrs}$, serum cortisol (after 1 mg Dexamethasone at mid night)- 39.2 $\mu\text{g}/\text{dl}$. USG of adrenal gland – bilateral enlarged suprarenal gland.



Fig. 1: Moon shaped face and truncal obesity are suggestive of cushing's syndrome

Patient was scheduled for bilateral adrenalectomy as ASA grade III. Difficult intubation was due to limited mouth opening, Mallampati grade IV, short neck so adequate and difficult intubation cart was kept ready. Patient was optimized preoperatively, hypertension was controlled with tab Nifedipine (10mg) three times daily, tab. Captopril (25mg) 6.5mg at night. tab. Propranolol (20mg) two times daily. Patient received antibiotic prophylaxis. Preoperative sedative and premedication was avoided. Routine antihypertensive medication were given in the morning tab. Ranitidine (150mg) was given at night. Inj. Hydrocortisone 60mg intravenous bolus was given preoperatively and peroperatively Inj. Hydrocortisone infusion 100mg in 500ml normal saline was started at 3-5mg/hr. In operation theatre patient's heart rate was 114/min, blood pressure was 150/78 mmHg, SpO_2 –92%. Non invasive blood

pressure, ECG and SpO_2 , temperature was continuously monitored.

Intravenous cannulation was done with 22 G canula. Preoxygenation was done with 100% O_2 for 5 minutes. Anaesthesia was induced by . fentanyl- 30 μg ., thiopental sodium – 100mg, intubation was done by rocuronium 25mg intravenously. Intubated the patient with cuffed endotracheal tube 4.5mm size internal diameter. Anaesthesia was maintained with halothane 0.5%, 50% oxygen, 50% N_2O . Incremental doses of . fentanyl and atracurium were given.

Intraoperatively, due to meticulous surgical dissection and adequate depth of anaesthesia, there were no major blood pressure fluctuation during adrenal manipulation except slight decrease in blood pressure after resection of bilateral adrenal gland which was controlled by . 0.9% normal saline 500ml mixed with dopamine (400mg) I/V at 10 microgram/kg / min. Duration of surgery was 3 hours.

Though vasodilator like sodium nitroprusside, glycerintrinitrate were kept ready for blood pressure control, it were not required. Fluid supplementation was given as 500ml crystalloid solution. Blood loss was minimal. Urine output was maintained throughout the procedure. At the end of surgery neuromuscular blockade was reversed with neostigmine 1.2mg and atropine 0.18mg. As patient could not maintained spontaneous respiration then patient was sent to the intensive care unit for mechanical ventilation. for next 24 hours.

Postoperatively arterial blood gas and blood sugar, serum electrolyte were monitored. The patient was kept in ICU and electively ventilated in controlled mode (tidal volume 300ml, respiratory rate-14/min). FiO_2 –0.6. Postoperative pain relief was done by pethidine (1mg/kg) 6 hourly, inotropic support was given by dopamine (200mg) in 50 ml in N/S at 10. microgram/kg /min Hypokalemia was corrected by potassium containing syrup through nasogastric tube. Hyperglycaemia was controlled by 30unit insulin and 50ml normal saline at 1 dial/min (according to the blood sugar). Patient received steroid supplementation with. hydrocortisone succinate by infusion 100mg in 500ml normal saline Extubation was done 24 hours after surgery in ICU. When the patient vital signs were stable patient then send to recovery room.



Fig.-2: Patient was in controlled mode ventilator in ICU.

Discussion

Cushing's syndrome is clinical entity resulting from adrenocortical hyperfunction. The signs and symptoms of cushing's syndrome are related to excess glucocorticoids. Patients present with increased body weight, truncal obesity with buffalo hump, cutaneous striae, odema, glucose intolerance.^{1,3} All these presentation make these patients a challenge to anaesthetists. The most common cause of cushings syndrome is iatrogenic administration of corticosteroids. Approximately 40% of endogenous causes are ACTH producing tumours and ACTH producing non pituitary tumour such as tumours of the lung, prostate, testis, parotid or pancreas. Nearly 25% of cases are due to adrenal hyperplasia without ACTH secreting tumours, 20% of patients have adrenocortical tumour, about half of which are benign adenomas. This patient was a case of bilateral adrenal hyperplasia. The clinical presentation with cushinoid feature.²In this case, hormonal levels showed gross elevation of cortisol, MRI did not show pituitary or adrenal neoplasm. While CT abdomen and brain yielded normal study,

ruling out multiple endocrine neoplasm (MEN) which is frequently associated with higher incidence of bilateral pheochromocytoma in children (20%) than in adults.^{6,7,8}

Hypertension due to adrenal hyperplasia in children tends to be more sustained along with salt retention and increased intravascular and interstitial volume.⁷ Patients may present with paroxymal symptoms like palpitation, trembling, sweating due to dopamine and epinephrine secretion. This patient was on tab Nifedepine and tab Captopril two times daily preoperatively. Surgical intervention is indicated for failed medical therapy and in cases of adrenal adenoma.⁹

The key factor is to remember when preparing patient for surgery is to get medically stabilized. Hyperglycemia is best controlled with regular insulin if needed. In this patient preoperative sugars were controlled (FBS=5.5mmol). The patient scheduled for bilateral adrenalectomy should be treated as addisonian crisis intraoperatively and postoperatively because normal adrenal tissue is suppressed by high level of circulating corticosteroids.¹⁰ Therefore, steroid therapy was instituted by giving . hydrocortisone 60 mg I/V half an hour prior to surgery, followed by 3-5mg/hr infusion peroperatively and continued postoperatively. Goals of anaesthetic management were aimed at suppressing response to endotracheal intubation. Surgical stimulation, adrenal handling and devascularization plus providing optimal surgical condition as in case of pheochromocytoma.^{8,11} Histamine releasing sedative premedicants are avoided in view of difficult airway. The anaesthetic agents preferred are thiopental sodium, propofol or inhalational agents as sevoflurane. Tracheal intubation may be facilitated by succinylcholine but due to fasciculation cortisol secretion may increase. So intubation was done by rocuronium. Anaesthesia is maintained by midazolam, fentanyl , sevoflurane and N₂O. Adequate muscle relaxation is necessary for good exposure of surgical field. Any of nondepolarizing muscle relaxants may be employed. Trans-abdominal resection is recommended in children, sodium nitropruside, phentolamine can be used for BP fluctuation during adrenal handling or Metoprolol I/V bolus to control tachycardia. In this case, meticulous surgical handling did not cause much fluctuation. Steriod cover is mandatory for

patients undergoing bilateral adrenalectomy. Our patient was discharged on oral prednisolone and fludrocortisone. After one week of surgical intervention this patient body weight reduced and patient able to walk.

Conclusion

Diagnosis and management of cushing's syndrome in paediatric patients is as challenging as in adult. Early involvement of anaesthesiologist is essential with better understanding of pathophysiology of adrenal hyperplasia, necessary monitoring and diagnostic modalities, availability of rapid acting drugs which can alter BP. Sophisticated and skilled anaesthesia and surgery all of these have given success to management of bilateral adrenalectomy in a 4½ years old child with hypertension and cushinoid features.

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