



Anaesthetic Management of Laparoscopic Adrenalectomy for Adrenocortical Tumour in a Paediatric Patient – A Case Report

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Abstract: An 8 months, 8kg child with cushingoid features with persistent severe hypertension was admitted in paediatric surgery ward. Blood pressure was controlled by ACE inhibitors & calcium channel blockers. Diagnosis of adrenal tumour was confirmed by abdomen CT scan and normal brain CT scan ruled out any pituitary tumour. We share our experience in successfully managing a challenging paediatric case of Cushing's Syndrome with childhood hypertension and difficult airway due to cushingoid features for unilateral adrenal resection by laparoscopy.

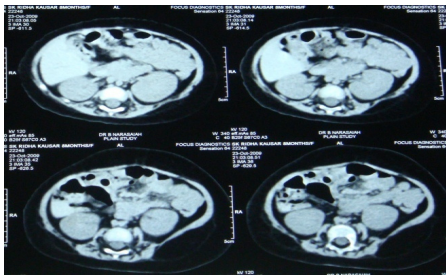
Introduction: Corticosteroid excess may be primary, due to an adrenal adenoma/hyperplasia, or secondary, to an ACTH secreting tumour, pituitary or "ectopic" origin (Cushing's disease), or to exogenous steroid usage. 10 to 20% cases of spontaneous Cushing's syndrome are caused by ACTH independent process, either an adrenal carcinoma or adenoma. Most of these adenomas are incidental findings (85% incidentalomas), discovered during screening CT Scans in a variety of clinical settings. A minority (about 15%) of adrenocortical adenomas are "functional" and produce glucocorticoids, mineralcorticoids, and/or sex steroids. Functional adrenocortical adenomas are surgically curable. Adrenocortical carcinoma (ACC) is a rare, highly aggressive cancer of adrenal cortical cells, which may occur in children or adults. Adrenocortical tumours are rare in children¹ and occur most commonly in females and in children less than four years. Eighty to ninety percent of patients have functional tumours with endocrine manifestations at diagnosis and the majority present with virilisation alone or in combination with overproduction of other adrenal hormones. Isolated Cushing syndrome is rare^{2,3}.

Case Report: An 8 month old female child came with complaints of rapid weight gain (mostly truncal), puffiness of cheeks, development of pubic and facial hair and acne, and clitoral hypertrophy within 4 months. She was suspected to have Cushing's syndrome. The diagnosis was confirmed by a raised serum Cortisol of 35.5mcg/dl (normal 5 to 23 mcg/dl) and CT scan abdomen showing left adrenal mass (41x37mm). CT brain was done to rule out pituitary adenoma and a normal study was

obtained. Urinary adrenaline levels were normal ruling out any abnormality of adrenal medulla. Her BP on admission was 210/118mm Hg and she was put on Tab. Nifedipine 5mg twice a day, Tab. Enalapril 2mg twice a day, Tab. Methyl Dopa 125mg 6 hourly, Tab. Spironolactone 12.5mg twice a day. A week before surgery her general examination revealed a pulse rate of 130/min and blood pressure of 130/100mm Hg. Other investigations like blood count, blood urea, serum creatinine were within normal limits. Her random blood sugar was 90mg% and serum potassium was 4.2meq/l. Patient was accepted for unilateral laparoscopic adrenal resection.



8Months old Child with Cushing's Syndrome



CT Scan Abdomen



Adrenal
Mass

Anaesthetic Management: Patient was kept on maintenance drip of Isolyte P 300ml over 12hrs in the ward. Morning doses of antihypertensives were given. A 24 G intravenous cannula was in situ in the right lower limb. Inj. Midazolam 0.3mg and Inj. Hydrocortisone 16mg IV were given 20 minutes before taking the patient in the operation theatre. In the operation theatre Inj. Glycopyrrolate 0.04mg was given IV. Monitors included pulse oximeter, NIBP, ECG, precordial stethoscope and EtCO₂. A towel roll was placed under her shoulders to aid intubation. Preoperative heart rate was 136/min, BP 140/90 mm Hg, SpO₂ 98%. She was preoxygenated for 3min and induced with Inj. Thiopentone 40mg and Inj. Succinylcholine 15mg IV. Intubation was done with a non-cuffed Portex endotracheal tube no. 4.5 under direct vision laryngoscopy. In spite of cushingoid features and short neck, the intubation was easy. There was no major pressor response to intubation. After confirming bilateral equal air entry, the endotracheal tube was fixed. Inj. Fentanyl 16 mcg IV was given. Another intravenous cannula was secured in right upper limb and since internal jugular venous cannulation was difficult owing to short neck and indistinct landmarks, right femoral vein was cannulated with 20G central venous catheter. A 12 G Ryle's tube was inserted nasally. An 18 G epidural catheter was inserted in L2-3 space with 4cm of catheter in epidural space and 6ml of 0.25% Inj. Bupivacaine was given after giving test dose. Anaesthesia was maintained with Inj Atracurium for muscle relaxation,



N₂O:O₂ (50:50) and Isoflurane (0.5%-1%). Epidural top up dose of 6 ml of 0.125 % Inj. Bupivacaine was repeated 2 hrs after the first dose.

Intraoperatively pneumoperitonium of 12cm of H₂O was created with CO₂. Inj hydrocortisone 20mg in 50 cc NS was started in an infusion @ 4mg/hr. BP fluctuations due to tumour manipulations and dissection were controlled by 0.01% of SNP @ 4mcg/kg/min, in addition 0.2mg boluses of Inj. Metoprolol were given thrice to control heart rate in excess of 150/min.

Time Interval	Heart Rate (beats/min)	Blood Pressure (mmHg)	SpO ₂ (%)	BSL* (mg/dl)
0 min	140	140/80	98	80
20 min	140	118/80	99	
40 min	156	138/70	99	
60 min	160	150/90	99	102
80 min	136	128/86	98	
100 min	130	120/70	99	
120 min	150	113/70	98	100
140 min	130	124/70	99	
160 min	134	140/90	99	
180 min	146	134/78	97	110
200 min	154	132/88	98	
220 min	142	138/90	99	
240 min	146	122/80	99	100

Table 1: Intraoperative Parameters (*BSL Blood Sugar Level)

There was no major blood loss (only about 60ml) and no blood transfusion was done. Fluid supplementation was given with 300 ml of Ringer Lactate. Urine output was 80ml at the end of 4 hours of surgery. Intraoperatively the other adrenal was found to be of normal size. Arterial blood gas analysis and blood sugar level done before extubation were within normal limits. At the end of the surgery patient was reversed with Inj. Neostigmine 0.4mg and Inj. Glycopyrolate 0.06 mg iv, followed by extubation.

Postoperatively, the patient was kept in the Intensive Care Unit. During immediate post operative period, Inj. Hydrocortisone 4mg/hr infusion and 0.01% Sodium Nitroprusside @ 4mcg/kg/hr (owing to raised BP) was continued. The dose was tapered and Sodium Nitroprusside stopped after 24 hrs when the BP stabilized to 130/90 - 120/90 mm Hg. Inj Hydrocortisone was also tapered over next 72 hrs. She was started on oral antihypertensives in same doses as in preoperative period from the next day. Epidural top up dose of 4 ml of 0.125% Inj. Bupivacaine was given 6 hourly for the next 3 days.

Day	Heart Rate (beats/min)	BP (mmHg)	SNP* Infusions
1	150	150/90 – 150/100	SNP reduced from 3 ml/hr to 0.5 ml/hr
2	152	150/100 – 120/90	SNP tapered and stopped
3	160	140/80 -110/70	
4	158	150/90 – 130/80	

Table 2: Postoperative Parameters (*SNP (Sodium Nitroprusside) 0.01%)



Day	Sr. Na (mEq/lit)	Sr. K (mEq/lit)	BSL*(mg/dl)	Others
1	146	4.8	150	
2	131	3.1	167	Sr. ACTH 10.2 (N 0 – 46) Sr.Cortisol 71.5 mcg/dl (N 5 – 25)
3	144	4.5	116	
4	134	3.6	146	

Table 3: Postoperative Investigations (*BSL Blood Sugar Level)

Hydrocortisone replacement was done at the rate of 4mg/hr infusion on day one followed by 12.5 mg 6 hourly on day two to four.

On 5th day, there was sudden fall in BP from 130/80 to 90/60 mmHg. The pulse rate was 210/min. Oral antihypertensives were immediately withheld, 160 ml Isolyte P was pushed through IV cannula, Inj. Hydrocortisone 80 mg was given IV stat and an infusion of 4mg/hr was started. The BP continued to fall to 80/60 mmHg, following which 80cc IV fluid was pushed. The patient responded with gradual rise in BP to 130/88 mmHg. On day 6, the Sr. K⁺ was 2.6 mEq/lit and SpO₂ on oxygen supplement was 92%. She gained 2 Kg weight within 5 days. Inj. Furosemide 4 mg IV eight hourly was started after which the saturation rose to 97% on oxygen supplement. Syrup potassium chloride 3 ml twice a day was started.

On day 7, patient's general condition suddenly deteriorated. She became tachypneic and the SpO₂ dropped to 92% on O₂ by nasal cannula. Her heart rate went up to 200-240/min. She was put on O₂ with ventimask, Inj. Furosemide was repeated. Patient did not respond. She started having spontaneous bleeding from nose and mouth. Her platelet count was 29000/dl. The CBC dropped to 2600/ μ L. PT was >30 sec (control 12 sec) and INR 3.8. She was suspected to be having sepsis and DIC (Disseminated Intravascular Coagulopathy), hence Piperacillin-Tazobactam 100 mg/kg/dose, eight hourly was started along with platelet and fresh frozen plasma transfusion. However patient did not respond to treatment, after which she was intubated and put on ventilatory support. Her SpO₂ did not improve, tachycardia continued in the range of 220-250/min, platelet count did not rise and she finally succumbed to sepsis. The histopathology report obtained on 10th postoperative day was adrenocortical carcinoma

Discussion: Cushing's syndrome is a clinical entity resulting from adrenocortical hyperfunction. The signs & symptoms of Cushing's syndrome are related to excess glucocorticoids. Patients present with increased body weight, truncal obesity with buffalo hump, easy bruisability, cutaneous striae, oedema, hypokalemia, glucose intolerance. All these presentations make these patients a challenge to anaesthetists⁴.

The most common cause of Cushing's syndrome is iatrogenic administration of corticosteroids. Approximately 40% of endogenous causes are ACTH producing pituitary tumours and ACTH producing non pituitary tumours such as tumours of the lung, prostate, testis, parotid or pancreas. 20% of patients with endogenous Cushing's have adrenocortical tumours, about half of which are benign adenomas⁴.

The first paediatric adrenal tumour was reported in 1865. Since then there have been occasional but regular reports of these cases. With present sophisticated imaging modalities these



tumours are being found incidentally (incidentalomas). However there appear to be few reports of the anaesthetic management of these children unlike pheochromocytoma which has well defined guidelines¹.

Our patient was an 8 month old child who presented with complaints of rapid weight gain (mostly truncal), puffiness of cheeks, development of pubic and facial hair, clitoral hypertrophy and acne. She was diagnosed as a case of Cushing's syndrome due to unilateral adrenocortical tumour and was posted for unilateral laparoscopic adrenal resection.

Paediatric adrenocortical tumours can lead to varying manifestations such as cushingoid features, virilization, premature puberty, hypertension, polyuria and polydypsia. The case examines some of the complex problems arising from the hormonal activity of paediatric adrenocortical tumours which must be addressed and appropriately managed in perioperative period¹.

This patient had severe persistent hypertension which was managed with Nifedipine, Enalapril, Methyldopa and Spironolactone. Though our patient had normal Sr. K⁺ level, Spironolactone was the preferred diuretic as it is K⁺ sparing, because these patients tend to manifest hypokalemia⁵. Hyperglycaemia is also an important consideration in these patients due to high Cortisol levels, and needs to be controlled preoperatively⁵. Our patient had normal blood sugar level preoperatively.

Goals of anaesthetic management should aim at providing optimal surgical conditions and suppress the responses to endotracheal intubation, surgical stimulation, tumour handling and devascularisation. General anaesthesia combined with regional anaesthesia is a preferred technique⁶.

Other challenges faced by anaesthesiologists in these cases are difficult intubation and difficult IV access owing to obesity with excessive pads of fat, indistinct landmarks and restricted extension of neck. This patient was cushingoid with short neck and Mallampati grade III, so we expected intubation to be difficult and had kept difficult intubation cart ready. However the intubation was easy with vocal cords readily visible on direct laryngoscopy.

Perioperative control of blood pressure and steroid cover are the most important perioperative considerations. This patient was controlled on oral antihypertensives in preoperative period, and intraoperative fluctuations in BP were managed with Sodium Nitroprusside drip and Metoprolol boluses. In these patients with unilateral adrenocortical tumours, adrenocorticotrophic hormone (ACTH) secretion by the pituitary is suppressed causing atrophy of contralateral adrenal gland. The inability of the suppressed hypothalamic-pituitary-adrenal (HPA) axis to respond to perioperative stresses can lead to acute adrenal insufficiency and cardiovascular collapse. This necessitates steroid coverage in the perioperative period, the common regime for which is intravenous hydrocortisone 2mg/kg given immediately before surgery followed by 3-5mg/hr infusion perioperatively and continued postoperatively⁷.



Premedication was given in the form of Inj. Midazolam in the ward to reduce anxiety. The anaesthetic agents preferred are Thiopental Sodium, Propofol or inhalational agents such as Sevoflurane⁸. Tracheal intubation is facilitated by Succinylcholine. Anaesthesia is maintained by IV agents like Midazolam, Fentanyl or inhalationals like Sevoflurane and N₂O⁸. Adequate muscle relaxation is necessary for good exposure of surgical field. Any of the nondepolarising muscle relaxants may be employed, however muscle relaxants should be used in titrated doses due to existing myopathy and hypokalemia⁵. In this case we used Thiopental for induction and Succinylcholine and Atracurium for muscle relaxation. Isoflurane was used in our patient because it provides good relaxation and analgesia without sensitizing the heart to catecholamines.

Laparoscopic surgery itself presents a different set of challenges to the anaesthesiologist. Both pneumoperitoneum and adrenal tumour manipulation may induce hemodynamic variations. A laparoscopic approach offers several advantages compared with an open laparotomy; namely, laparoscopic adrenalectomy decreases fluid shifts that may accompany an open procedure, potentially decreases the surgical stress imposed on the patient, decreases the need for postoperative analgesia, shortens postoperative convalescence including an intensive care unit stay, and decreases overall hospital stay⁹.

Postoperative monitoring of serial blood sugars, serum electrolytes is very important. Surgical stress may aggravate the glucose intolerance associated with Cushing's syndrome. Moreover, postadrenalectomy there is a tendency towards hypoglycemia because of impaired hepatic gluconeogenesis especially if steroid replacement is inadequate⁷. Selective hypoaldosteronism often occurs postadrenalectomy, resulting from preoperative treatment with Spironolactone which antagonises Aldosterone receptors and inhibits adrenal Aldosterone production. The mineralocorticoid activity after surgery, though, is often sufficient to prevent hyperkalemia if sodium intake is adequate. Nevertheless, the possibility of such electrolyte imbalance occurring emphasizes the need for frequent electrolyte measurements.

In spite of meticulous and planned anaesthetic management, our patient succumbed to septicaemia and DIC on the 7th postoperative day. This demonstrates the fact that in spite of proper pre, intra and postoperative management, other factors including sepsis, immunity of the patient, postoperative nursing care can influence the morbidity and mortality of the patient.

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