

Anaesthetic Management of Bilateral Adrenal Hyperplasia in a Paediatric Patient

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Abstract

Nearly 25% of the cases of Cushing's syndrome are due to adrenal hyperplasia without an ACTH secreting tumor. PPNAD (Primary Pigmented Adrenal Nodular Disease) is a rare condition with bilateral nodular hyperplasia in adrenal gland. A 4 year old 14 kg child with Cushing features of hyperpigmentation with hypertension was admitted with persistently elevated serum cortisol level but depressed ACTH level. Blood pressure was controlled by ACE inhibitors & diuretics. Diagnosis of PPNAD was done as USG Abdo-pelvis showed no evidence of adrenal tumor and normal brain CT scan ruled out any pituitary tumor. We share our experience in successfully managing a challenging paediatric case of adrenal hyperplasia with childhood hypertension and difficult airway due to cushinoid features.

INTRODUCTION

Cushings syndrome is a clinically entity resulting from adrenocortical hyperfunction. The most common cause is iatrogenic resulting from administration of corticosteroids. 25% of the cases are contributed due to adrenal hyperplasia without an ACTH secreting tumor₁. PPNAD is a condition where there is a bilateral nodular hyperplasia in adrenal gland and the atrophy of adrenal tissue adjacent to the nodules₂. Clinical presentation is typical with features of Cushing's syndrome, but always before 30 yrs of age and before 15 yrs of age in 50% cases. Family history is positive in about 20% of the cases of PPNAD as part of the familial autosomal dominant condition called Corney Complex. This comprises of mesenchymal tumors especially cardiac myxomas, spotty skin pigmentation, peripheral nerve tumors, breast lesions, GH hormone secreting pituitary tumors and testicular tumors_{2,3}. Only about 100 cases of PPNAD have been reported so far. This case report describes the perioperative anaesthetic management in a child with a rare presentation of bilateral hyperplasia due to PPNAD.

CASE REPORT

A four year old male child came to OPD with complaints of rapid weight gain, darkening of complexion, puffiness of face and mild abdominal distension, all within one month duration. Past history revealed that the patient was on steroid therapy (T Wysolone¹ / 2 BD) started by private

dermatologist for white patches on skin and grey hair, 3 months back. General examination showed vellous hypertrichosis, genu valgum and erythematous hue over the cheek. Anthropometrical measurements revealed a low height (95cm, 10-25th percentile) and body weight of 18 kg. He had been on ACE inhibitors & T.Lasilactone which had been started at the referring institution and blood pressure on standing position was 110/80 mmHg.

Haematological investigation were grossly normal but for relative lymphocytosis. X-ray hand revealed the bone age of 2yrs 4mnths and it was lagging behind the chronological age of 4yrs 2mnths. CT of brain and abdomen yielded a normal study. The patient was suspected as case of Cushing's syndrome & hormone level estimation was done, which showed grossly elevated cortisol values -50µ/dl (normal values 9-24µ/dl) with grossly decreased ACTH levels. A repeat ACTH assay too confirmed the same ACTH levels which ruled out ACTH being the cause for hyperpigmentation. Meanwhile MRI too failed to pick up any pituitary or adrenal neoplasm.

The patient was diagnosed as a case of ACTH independent Cushing's syndrome and additional work up to rule out other associated features of Corney complex, namely cardiac myxomas (2D-ECHO), thyroid & testicular tumors (ultrasonography) were done which revealed normal study.

Hence the patient was suspected as a case of PPNAD and bilateral adrenalectomy was planned & preoperative work up was done. Difficult intubation was anticipated in view of limited mouth opening, MPC grade IV, short neck and so adequate precautionary measures were taken.

Antihypertensive medication was changed to T.Amlodipin 2.5mg ½ OD.

Preoperative sedative and premedication was avoided. Inj. Glycopyrrolate 70µg was given I.V. Routine antihypertensive medications were given in the morning. Inj. Hydrocortisone infusion 100mg in 500cc NS was started at 3-5mg/hr. The difficult intubation cart was kept ready. The patient was induced with sevoflurane in 100% oxygen by face mask. Monitoring included heart rate, ECG, NIBP, SpO₂ and EtCO₂. An IV line was secured in the right upper limb. The preinduction HR was 122/min and BP was 110/80 mmHg (mean 90mmHg). Adequate depth of anaesthesia was attained with Inj. Propofol 30mg IV, direct laryngoscopy done after Inj. Scoline 35mg was given & intubation was done with 4.5 No. uncuffed PVC ET Tube. There was no major pressor response to intubation. After ET Tube was secured, left radial artery was cannulated for beat to beat monitoring of arterial BP. The right IJV cannulation was then performed to monitor CVP (initial CVP was 6-8 cmH₂O). Inj. Midazolam 0.5mg and Inj. Fentanyl 30µg IV was given for sedation and analgesia. Anaesthesia was maintained with N₂O:O₂(50:50), intermittent Sevoflurane, Inj. Atracurium for muscle relaxation.

Intraoperatively, due to meticulous surgical dissection and adequate depth of anaesthesia, there were no major Blood pressure fluctuations during adrenal manipulation. Though vasodilators like SNP, NTG were kept ready for BP control, they were not required. Fluid supplementation was given with 530cc of crystalloid & 150cc of Hydroxyethyl starch. Blood loss was 120cc & not replaced with blood. Urine output was maintained throughout the procedure (525cc at the end of procedure).

Postoperatively ABG & HGT were monitored. At the end of surgery patient was reversed with 0.9 mg of Neostigmine & 140µg Inj. Glycopyrrolate and was extubated after checking good air blast & adequate reversal of muscle relaxation with return of good tone.

Postoperatively, the patient was kept in the ICU for 7 days. During immediate post operative period Insulin was started according to sliding scale as HGT was 216-270, but stopped on first day itself as sugar was controlled. Inj.

Hydrocortisone 3-5mg/hr infusion was kept postoperative 24hrs. The child was shifted on oral Dexamethasone 4mg TDS. The patient was discharged on 10th postoperative day on oral prednisolone 5mg OD and fludrocortisone 0.15mg OD.

DISCUSSION

Cushing's syndrome is 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, edema, hypokalemia, glucose intolerance^{1,3}. All these presentations make these patients a challenge to anaesthetists.

The most common cause of Cushing's syndrome is iatrogenic administration of corticosteroids. Approximately 40% of endogenous causes are ACTH producing pituitary tumors and ACTH producing non pituitary tumors such as tumors of the lung, prostate, testis, parotid or pancreas. Nearly 25% of cases are due to adrenal hyperplasia, without ACTH secreting tumors. 20% of patients with endogenous Cushing's have adrenocortical tumors, about half of which are benign adenomas.

This patient was a case of bilateral adrenal hyperplasia, condition called Primary Pigmented Nodular Adrenal Disease. The adrenal nodules are 2-4mm in size & black or brown in cut section with atrophy of adjacent adrenal tissue. The clinical presentation is typical with Cushingoid features, but always seen before 30yrs of age & before 15yrs in 50% cases².

PPNAD^{2,4} forms a part of familial autosomal dominant condition called Corney Complex, presentations has been stated earlier like cardiac myxomas. The genetic loci have been on chromosome 2 & 17.

All these factors make preoperative General & systemic examinations and specific investigations like 2D-ECHO, MRI-CT Scan with hormonal assays important in these patients. In this case, hormonal levels showed gross elevation of cortisol with decreased ACTH levels on repeated assays which ruled out ACTH as the cause of hyperpigmentation. MRI didn't show pituitary or adrenal neoplasm while CT abdomen, brain yielded normal study, ruling out Multiple Endocrine Neoplasia (MEN), which is frequently associated with higher incidence of bilateral pheochromocytoma in children (20%) than in adults^{3, 6,7}.

Hypertension due to adrenal hyperplasia in children tends to be more sustained along with salt retention & increased intravascular & interstitial volume_s. Patients may present with paroxysmal symptoms like palpitation, trembling, sweating due to dopamine & epinephrine secretion. Administration of the aldosterone antagonist spiranolactone 25-100mg BD is the medical treatment indicated for patients with Bilateral hyperplasia. This patient was on T. Lasilactone & T. Amlodipine 2.5mg ¹/₂ BD preoperatively.

Surgical intervention is indicated for failed medical therapy & in cases of adrenal adenoma_s. The key factor to remember when preparing patient for surgery is to get him medically stabilized. Hyperglycemia best controlled with regular insulin if needed. In this patient preoperative sugars were controlled (HGT=72-108). Hypertension was controlled with use of diuretics, which may augment the already existing hypokalemia that should be treated concomitantly.

Hypokalemia also causes muscle weakness which coupled with truncal obesity may compromise respiration in this patients. Therefore hypokalemia should be adequately treated & acid-base status stabilized, The patient posted for adrenalectomy should be treated as Addisonian intraoperatively and postoperatively because normal adrenal tissue is suppressed by high levels of circulating corticosteroids_s. Therefore steroid therapy was instituted by giving Inj. Hydrocortisone 25mg IV, half hour prior to surgery, followed by 3-5mg/hr infusion perioperatively and continued postoperatively.

Goals of anaesthetic management were aimed at suppressing responses to procedures like endotracheal intubation, surgical stimulation, adrenal handling & devascularisation plus providing optimal surgical condition as in case of pheochromocytoma_{7,10}.

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 facilitated by succinylcholine. Anaesthesia is maintained by IV agents like midazolam, fentanyl or inhalationals like Sevoflurane and N₂O. Adequate muscle relaxation is necessary for good exposure of surgical field. Any of nondepolarising muscle relaxants may be employed. Transabdominal resection is recommended in children. SNP, phentolamine can be used for BP fluctuations during adrenal handling or Metoprolol IV bolus to control tachycardia. In this case, meticulous surgical handling didn't cause much fluctuations.

Steroid cover is mandatory for patients undergoing bilateral adrenalectomy. Our patient was discharged on oral prednisolone & fludrocortisone.

CONCLUSION

Diagnosis and management of Cushing's syndrome like clinical disorder called PPNAD in paediatric patient is as challenging as in adult₁₁. 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 & skilled anaesthesia and surgery, all of these have given success to management of bilateral adrenalectomy in a 4yr old child with hypertension & Cushingoid features due to rare cause of adrenal hyperplasia-PPNAD.

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