Case Report

Anaesthetic management of a patient with Allgrove syndrome

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ABSTRACT

Allgrove syndrome is a rare autosomal recessive disorder, which manifests with adrenal insufficiency, achalasia cardia and alacrimia. Autonomic neuropathy can also be associated with it. Adrenal crisis can be precipitated by surgery, infection or trauma. This disorder poses a challenge to anaesthesiologists during anaesthesia for various surgeries. We report the anaesthetic management of a child with Allgrove syndrome.

Key words: Achalasia, adrenal crisis, alacrimia, Allgrove syndrome, anaesthesia

INTRODUCTION

Allgrove (AAA) syndrome is an autosomal recessive disorder characterised by achalasia cardia, alacrimia and adrenocorticotropic hormone (ACTH) resistant adrenal insufficiency, which generally presents in the first decade of life. AAA syndrome was first described by Allgrove *et al*. in 1978.[1] In addition to the above cardinal manifestations, it may be associated with autonomic neuropathy, neurologic and skin manifestations at a later age. We report the successful management of a three year old girl with Allgrove syndrome during Heller's cardiomyotomy and augmentation cystoplasty.

CASE REPORT

A 3 year old girl weighing 13 kg was scheduled to undergo laparoscopic Heller's cardiomyotomy and augmentation cystoplasty in our hospital. She was delivered at full term by caesarean section due to breech presentation. She had history of vomiting after each meal and repeated urinary tract infections. A diagnosis of Allgrove syndrome was made after identification of the triad of achalasia cardia, adrenal insufficiency and alacrimia. She also had right sided vesico-ureteric reflux and high bladder pressures. She had undergone repeated oesophageal balloon dilatations earlier. She was on replacement dose of oral hydrocortisone (10 mg/day) for adrenal insufficiency for the past 2 years. During preoperative assessment, her systemic examination was normal and airway was adequate. The postural tests for autonomic dysfunction like heart rate and blood pressure response to standing for 3 min from the supine position were normal. She had normal haemoglobin (11.6 g%), renal function (serum creatinine - 0.3 mg/dl), serum electrolytes and normoglycaemia (random blood glucose - 130 mg %).

Standard nil per oral instructions were followed and Ringer lactate at 50 ml/h was used as maintenance fluid. Injections ranitidine 25 mg and ondansetron 1.5 mg were given intravenously as premedication 1 h before surgery. After thorough suctioning of nasogastric tube, which was *in situ*, general anaesthesia was induced with intravenous fentanyl 25 μg, propofol 30 mg, atracurium 7.5 mg and cricoid

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pressure was given. The trachea was intubated with 5 mm cuffed endotracheal tube. She was monitored with electrocardiogram, noninvasive blood pressure, pulse oximetry, end tidal carbon dioxide ($EtCO_2$) and nasopharyngeal temperature. Eyes were protected with topical lubricant eye ointment. Anaesthesia was maintained with sevoflurane, oxygen, air and intermittent doses of intravenous morphine and atracurium. Mechanical ventilation was with pressure controlled ventilation and $EtCO_2$ was maintained between 30 and 40 mm Hg. Ringer lactate at 50 ml/h was used as maintenance fluid. Hydrocortisone was given as 50 mg intravenous bolus at induction and then as an intravenous infusion of 2 mg/h. Blood glucose levels was monitored hourly. Laparoscopic Heller's cardiomyotomy for repair of achalasia cardia was performed first followed by augmentation cystoplasty. Total duration of surgery was 7 h. Insulin infusion was started after 3 h as blood glucose rose to 280 mg%. Insulin infusion was successfully stopped 3 h later as normoglycaemia was achieved. 200 ml packed red blood cells was given to replace blood loss and ringer lactate was used to replace fluid loss. Intraoperative haemodynamics were stable and normothermia was maintained. Intraoperative arterial blood gases and electrolytes were normal. Recovery and extubation were uncomplicated. Postoperative analgesia was provided with intermittent intravenous morphine. She received high dose hydrocortisone infusion (50 mg/day) for 2 days and then the steroid was slowly tapered to normal supplemental dose. Serum electrolytes, blood glucose levels were normal in the postoperative period. She was discharged home after 10 days.

DISCUSSION

Allgrove syndrome (triple A or 4A syndrome) is a rare autosomal recessive disorder with features of adrenal insufficiency, achalasia cardia and alacrimia with or without autonomic disturbance.[2] Neurological symptoms such as hyperreflexia, dysarthria, hypernasal speech, ataxia, sensory impairment, muscle weakness, and mental retardation are extremely slow to develop and manifest at a later age. Hyper pigmentation of the skin and osteoporosis may appear in a few patients by adulthood.[3] The pathology is due to mutation in type II keratin gene cluster on chromosome band 12q13 and also to progressive loss of cholinergic function throughout the body.[4] It usually presents in the first decade of life with deficiency of tears, recurrent vomiting and dysphagia due to achalasia, severe hypoglycaemic seizures and shock due to adrenal insufficiency.[5]

Preoperative investigations should include oesophageal motility tests, endoscopy, serum electrolytes, 9 am serum cortisol and ACTH levels, ACTH stimulation test and Schirmer test for semi quantitative measure of tearing.

Preoperative tests for autonomic neuropathy in children are described[6] such as heart rate response to standing (ratio of R-R interval at 30^{th} beat to 15^{th} beat <1 is abnormal), blood pressure response to standing (fall in systolic blood pressure >20 mm Hg and diastolic blood pressure >10 mm Hg after standing for at least 3 min from the supine position is abnormal), heart rate response to Valsalva manoeuvres (ratio of longest to shortest R-R interval <1 is abnormal), Pilocarpine eye test (causes miosis in autonomic dysfunction and no effect in normal children) and thermoregulatory sweat test. The patient described did not have features of autonomic neuropathy, but positional changes should be undertaken carefully in these patients.

Adrenal insufficiency is diagnosed if baseline plasma cortisol is <20 μg/dl and the peak cortisol level is <20 μg/dl following an ACTH stimulation test.[7] These patients need replacement with oral hydrocortisone. During periods of stress (trauma, infection or surgery), the hydrocortisone dose needs to be increased significantly. Non recognition or inadequate treatment of adrenal insufficiency during perioperative period may precipitate adrenal crisis leading to hyponatraemia, hyperkalaemia, hypoglycaemia, hypotension and shock.[7] Thus, they should receive stress dose of steroid in the perioperative period.[8] Hydrocortisone is preferred due to its balanced (1:1) mineralocorticoid and glucocorticoid effects.

The patient described was already on supplemental dose of oral steroids, and we provided augmented dose of steroid by infusion during surgery and in the postoperative period. Continuous infusion of steroids has the advantage of maintaining uniform levels of plasma cortisol. Blood glucose levels should be closely monitored as administered steroid can cause hyperglycaemia. In the present case, hyperglycaemia during the surgery was managed with insulin infusion.

These patients are at high risk of regurgitation and aspiration. Antiemetics, antacids and head end elevation are helpful. Alacrimia, absence of protective corneal reflexes and reduced tear production under general anaesthesia can lead to keratopathy and

corneal ulceration. Hence, topical lubricant eye ointments should be used.[9]

CONCLUSION

Careful preoperative evaluation, stress dose of steroids, perioperative glycaemic control, prevention of aspiration and careful eye protection are imperative for optimal anaesthetic management of a patient with Allgrove (AAA) syndrome.

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