Case Report

Anaesthetic Management of a Child with Pituitary Stalk Interruption Syndrome for Dental Surgery

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Abstract

Pituitary Stalk Interruption Syndrome (PSIS) is a rare congenital anomaly of pituitary gland characterized by triad of thin or interrupted pituitary stalk, aplasia or hypoplasia of the anterior pituitary and an absent or ectopic posterior pituitary (EPP) seen on magnetic resonance imaging (MRI). PSIS is associated with several midline malformations including facial dysmorphism, septal agenesis, partial corpus callosum agenesis, aqueductal stenosis, optic nerve hypoplasia, craniopharyngeal canal, cleft lip and palate. The anaesthetic management of a patient with hypopituitarism presents several problems like precipitating acute adrenal crisis, decreased resting cardiac output which cannot be increased when stressed, relative hypovolemia on induction, low basal metabolic rate and an inability to increase core temperature. The anaesthetic management of a four-year-old boy diagnosed to have growth hormone deficiency with early childhood caries for pulpectomy and root canal treatment under genral anaesthesia is discussed.

Key Words: Pituitary stalk interruption syndrome, Isolated growth hormone deficiency

Introduction

Pituitary stalk interruption syndrome (PSIS) is rare entity with an estimated incidence rate of 0.5/1,000,000 births.[1] This can be associated with mid line defects and various pituitary endocrine deficiencies, ranging from isolated growth hormone deficiency (IGHD) to combined pituitary hormone deficiency (CPHD).

The endocrine outcome seems to be gradual onset of hormone deficiencies leading to pan hypopituitarism, but posterior pituitary function is usually maintained.[2] PSIS is also associated with higher than normal frequency of breech presentation, difficult delivery, or the consequence of adverse perinatal factors such as birth trauma, prolonged labour, forceps delivery. Later in childhood, children may present with short stature, decreased growth rate, seizures, hypotension, intellectual delay and delayed puberty.[3] This case report mainly emphasises the anaesthetic management of a child with isolated growth hormone deficiency (IGHD) posted for pulpectomy and root canal treatment of lower three central incisors.

Case Report

A four-year-old male child weighing 17 kg and height of 70cm with body surface area 0.5m^2, born of non-consanguineous marriage with birth weight of 3.3kg by vaginal delivery, presented to pre-assessment clinic with early childhood caries for pulpectomy and root canal treatment. On routine clinical check up at the age of two, the child was found to have nasal bone hypoplasia and on further evaluation glucagon stimulation test revealed growth hormone deficiency. MRI brain showed partial empty sella with complete absence of pituitary stalk. Child was short statured, less than 3rd percentile according to WHO growth chart and had nasal bone hypoplasia. Child was on regular growth hormone supplements – Injection Growth Hormone 0.7mg subcutaneous once a day and hydrocortisone 5mg per orally as twice a day dose. On general physical examination the child was active, playful, short statured with hypotelorism, nasal bone hypoplasia and bilateral ptosis. (Fig 1)

There was no delay in milestones and intellect was normal. Pulse rate was 90 beats per minute, blood pressure 90/60mmHg and respiratory rate 20 breaths per minute. Systemic examination was normal. Airway examination revealed Mallampatti grade II, adequate mouth opening, no loose tooth. Hb- 12.6gm/dl, random blood sugar - 86mg/dl, Serum Sodium - 136mEq/L, growth hormone stimulation test using glucagon was 3ng/ml (Normal- 5- 20ng/ml) TSH - 2.0 µIU/mL (Normal- 0.5- 7 µIU/mL) and ACTH 14pg/mL (Normal- 7- 28pg/mL).[4] Child was assessed under American Society of Anesthesiologist (ASA)-II physical status, routine fasting guidelines followed, injection growth hormone 0.7mg SC and Tab. Hydrocortisone 5mg orally administered on the day of surgery.

Intraoperative period

Child was pre-medicated with Inj. Glycopyrrolate 0.2mg and Midazolam 1mg Intravenous (IV) prior to

was induced with Inj.Thiopentone sodium100mg IV, Inj.Fentanyl 50mcgIV. After confirmation of adequate ventilation neuromuscular blockade was achieved with Inj.Atracurium 8mg IV and nasotracheal intubation was accomplished with 4 mm ID cuffed RAE tube. Anaesthesia was maintained with oxygen: air, isoflurane at titrated doses (0.6% - 0.8%) with MAC 0.8. Continuous non-invasive monitoring was carried out, which included heart rate (HR), non-invasive blood pressure (NIBP), electrocardiogram, pulse oximetry (SpO_2), end tidal carbon dioxide ($EtCO_2$), and temperature. Forced air warmers and blankets were used to avoid hypothermia.Intraoperative period was uneventful and the duration of surgery was 2 hours. Neuromuscular blockade was reversed using Inj. Glycopyrrolate 0.1mg + Inj. Neostigmine 0.8mg IV and trachea extubated when child was awake and responding to commands, with stable heart rate and blood pressure. He was shifted to post anaesthesia care unit in a stable hemodynamic condition and injection Hydrocortisone 35mg IV was given 6 hours after the surgery. The child was observed for hypoglycemic episodes, hemodynamic stability and discharged on 2nd postoperative day.

Discussion

It is mandatory to have clear knowledge about the clinical manifestation of hypofunction of pituitary gland for the preoperative preparation and successful anesthetic management. Clinical manifestations of hypopituitarism include moderate normocytic and normochromic anaemia reflecting bone marrow hypofunction, decreased bone mineral density.In addition to hypogonadism, there is a decreased resistance to infection and stress, insulin sensitivity and tendency to hypoglycaemia. Hyponatremia occurs in case of deficient cortisol and thyroid hormones secretion.[5]

Patients with pan hypopituitarism are subject to water intoxication and hypoglycaemia, sensitive to sedatives and general anaesthetics, and because of hemodynamic instability they require circulatory support with vasoactive drugs.[6] Hypotension may occur if steroid cover is inadequate, it should be corrected with intravenous hydrocortisone 4mg/kg followed by 2mg/kg after 6 hours.[7]

Continuous monitoring of cardiovascular function, hourly diuresis, fluid balance measurement, the concentration of electrolytes in these patients is of vital importance. Difficult intubation has been described in these patients for several reasons. Midline defects includes cleft lip and palate. Spondiloepiphyseal dysplasia with scoliosis of the neck makes difficult the neck mobility, and there are numerous abnormalities of the cervical spine,such as congenital absence of odontoid protrusion. Abnormalities of lung function as mucopolysaccharidosis may compromise the airway.[8]

Patients with multiple anterior pituitary hormone deficiency presents with secondary adrenocortical insufficiency, dwarfism, and secondary hypothyroidism. These patients are at increased risk of hypothermia and cardiovascular depression refractory to catecholamines. Perioperative management includes administration of supplemental corticosteroids, intravenous infusion of sodium containing fluids. As these patients may be sensitive to drug induced myocardial depression, titrated dose of induction agents should be administered. Plasma concentration of glucose and electrolytes should be measured frequently during perioperative period. In view of skeletal muscle weakness, the initial dose of muscle relaxant may need to be reduced.

Preoperative correction of hyponatremia is necessary in patients with syndrome of inappropriate of antidiuretic hormone secretion(SIADH), as it may contribute to delayed awakening and potentiates the action of neuromuscular blockade. In this case, as the child had isolated growth hormone deficiency he was more prone to hypoglycemia and sensitive to inhalational agents. The child was administered glucose containing crystalloids and continuous monitoring of NIBP, ECG and maintained on isoflurane with the MAC of 0.6 to 0.8.[9]

The child was covered with forced air warmer and warming blankets to prevent hypothermia. Adequate measures were taken while positioning as these patients are prone for fractures due to reduced bone mineral density.[10]

Conclusion

Patients with pituitary gland disorder can have increased perioperative risk because of disturbed homeostasis of the whole endocrine system. Frequency of pituitary gland diseases is not high, but perioperative preparation requires great knowledge of regulative control mechanisms for pituitary gland hormone secretion and also for target glands. Pituitary gland disorders can be in the form of gland hypofunction or hyperfunction with isolated or multiple hormone disturbances. Steady-state hormone status regulation with substitution or suppressive therapy is necessary. So, we can conclude that perioperative anaesthetic care in patients with hypopituitarism requires careful preoperative assessment and meticulous perioperative management to avoid adrenal insufficiency, hypoglycemia, hypotension and hypothermia.

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Answer to : **Interesting ECG**

Rhythm strip showing Aypical Atrial Flutter

- Narrow complex tachycardia
- Regular atrial activity at ~300 bpm(White arrows)
- Variable block - 2:1 to 3:1, with Ashmans phenomenon(Shaded Arrows)
- Atypical Flutter waves ("saw-tooth" pattern) in Leads V1,V2,II. Positive flutter waves in Lead II
- Loss of the isoelectric baseline

Important to distinguish from AVNRT and Atrial tachycardia. Both have an isoelectric baseline, and an atrial rate between 150 – 250bpm. Atrial Flutter has an atrial rate of 250-350bpm.

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