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Case Report

A child with congenital hypothyroidism posted for emergency surgery- anaesthesia challenges

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ABSTRACT

Paediatric cases pose considerable anaesthetic challenges and are more challenging if they are posted for emergency surgeries with associated comorbidities like congenital hypothyroidism. Few reported cases of anaesthesia management of congenital hypothyroid cases are there in the literature. Challenges are due to the difficult airway, associated congenital anomalies and increased sensitivity to anaesthetic drugs. Knowledge of pathophysiology, preoperative optimization and proper selection and planning of anaesthesia techniques are essential for successful anaesthesia management of children with congenital hypothyroidism for emergency surgeries.

Key message: Congenital hypothyroidism in a child poses significant challenges for anaesthesiologists because of altered physiology, airway anatomy and difficult airway and drug metabolism.

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1. Introduction

Congenital hypothyroidism is one of the most common preventable causes of intellectual disability and congenital endocrine disorder in the childhood.[1] It is defined as a deficiency of thyroid hormones at birth. If the condition is not detected and treated early can lead to poor neurodevelopmental outcomes. The worldwide incidence of congenital hypothyroidism is 1 in 3,000-4,000 live births and the associated difficult airway is 11.1%.[2] Because of the newborn screening program, the prevalence is reduced. Children with congenital hypothyroidism especially untreated may have macroglossia, upper airway edema and if goiter is present may lead to a deviated airway. The present case is 3-year-old female child with congenital hypothyroidism posted for emergency abscess drainage of the upper limb.

1.1. Case history

A 3-year-old female child with a known case of congenital hypothyroidism was presented to the pediatric unit with right forearm abscess, cold and cough for 5 days. The child was diagnosed with congenital hypothyroidism at the age of 4 months when she was admitted to the hospital for severe dehydration. And was started on tab. Levothyroxine 50 mcg once a day. She had a previous history of one episode of seizure at the age of 2 years but was not on any medication. She was the fifth child from a lower socioeconomic status, born out of a nonconsanguineous marriage at home. The child is partially immunized and has global developmental delay and stunted growth. The child was posted for emergency incision and drainage for the right forearm abscess. On examination pulse rate was 130 bpm, blood pressure 90/60 mmHg, respiratory rate was 20 per minute and saturation on room air was 98 per cent. She looked pale and anasarca was present. Height is 72 cm, weight is 10 kg and Head circumference is 43 cm. Head-to-foot examination showed thick dry and

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cold mottled skin, opened anterior fontanelle depressed nasal bride. Haemoglobin and platelet were 7.5g/dL and 73,000cells /μL respectively. Thyroid function tests showed a serum T3 value of 0.38ng/ml, T4 value of 4.6 mcg/ml and TSH value of 150 mIU/ml. High risk informed consent was take from parents. Tab. thyroxine 50mcg was given on the day of surgery. Difficult airway equipment was kept ready. In the Operation theatre monitors were attached, 24 G IV was insitu on left forearm and started with Ringer's lactate solution. Inj. Ondansetron 1mg, Inj. Glycopyrolate 40 mcg and Inj. fentanyl 10 mcg, Inj. Hydrocortisone 20 mg IV were given. The case was done by maintaining spontaneous ventilation on the Jackson's Rees circuit with sevoflurane and graded doses of Inj. Propofol. The procedure lasted for 10 minutes. Blood loss was approximately 50 ml and total intravenous fluid given was 100 ml. The child was hemodynamically stable throughout the procedure. When the child was awake, started moving limbs and there were no features of respiratory distress, she was shifted to PICU for further management. In the postoperative period, the child was given 100 ml of packed red cells.

2. Discussion

Thyroid hormones play a major role in growth, development and metabolism. In the nervous system development during prenatal and newborn periods, it helps in neuronal differentiation, synapsis development, and myelination. Congenital hypothyroidism is classified into primary (thyroidal origin) and secondary (central origin). Defects in thyroid gland development (thyroid dysgenesis) or a defect in thyroid hormone biosynthesis (dyshormonogenesis) are the major causes of primary hypothyroidism. 3 The majority of CH is due to thyroid dysgenesis (80%) and 15-20 % is hereditary. In iodine-poor regions, iodine deficiency is the leading cause of congenital hypothyroidism. A newborn with congenital hypothyroidism may have few or no clinical manifestations. The early classical clinical manifestations include lethargy, poor or hoarse cry, large anterior and posterior fontanels, feeding difficulty, pathological jaundice, constipation hypothermia and hypotonia. These manifestations develop gradually over six weeks if untreated. After six weeks classical late manifestation starts appearing which includes coarse facies with depressed nasal bridge, coarse hair, thick, dry and cold mottled skin, puffy eyelids, large tongue, abdominal distension, umbilical hernia anaemia, hypotension, hyporeflexia, bradycardia. Myxoedema may lead to respiratory distress and cardiovascular collapse in the perioperative period. 4 The treatment of choice for congenital hypothyroidism is levothyroxine(T4). Early diagnosis by newborn screening and early treatment improves the outcome in these patients. In infants with congenital hypothyroidism, there is an 8.4% chance of having other congenital abnormalities like cardiac malformations, including septal defects, renal abnormalities, and the risk of neurodevelopmental disorders.[3] It can also be associated with syndromes like Down syndrome, Pendred syndrome, Bamforth-Lazarus syndrome and brain-lung-thyroid syndrome.[4] It is important to rule out all these anomalies and syndromes preoperatively. A preoperative 2D echo can rule out congenital cardiac anomalies, associated myopathies and abnormal cardiac function.

Congenital hypothyroid patients can come for non-thyroid surgeries or thyroid surgeries in case of diffuse goitre. It is better to make the patient euthyroid before surgery, as hypothyroidism patients have reduced cardiac function, abnormal baroreceptor function, ventilatory dysfunction, anaemia, reduced plasma volume, hypoglycaemia, hypothermia, hyponatraemia, and impaired hepatic drug metabolism.[5] These children have macroglossia, upper airway oedema and deviated airway in case of goitre which will lead to Anticipated difficult airway. The choice of anaesthesia is regional if it is permissible as it can avoid airway-related complications and polypharmacy during general anaesthesia. For this child anaesthesia options were sedation with USG-guided supraclavicular block, GA with spontaneous /controlled ventilation using endotracheal tube or LMA, Bag mask ventilation with sevoflurane and IV sedation. Because of the non-availability of a UGS machine in our institute, we could not give a brachial plexus block. The child had a history of upper respiratory tract infection, so we planned for minimal airway instrumentation to prevent laryngospasm. Altered drug metabolism and anaesthetic agent sensitivity may lead to delayed awakening and prolonged and residual neuromuscular blockade in children undergoing general anaesthesia.[6] It is better to avoid premedication drugs in these cases. Hypothermia should be prevented and a stress dose of steroid should be given.[7]

3. Source of Funding

None.

4. Conflict of Interest

None.

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