Anaesthesia and H syndrome: Navigating the complexities

Case Report

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ABSTRACT

H syndrome is a rare autosomal recessive disorder characterized by a spectrum of clinical features, including hyperpigmentation, hypertrichosis and multi-system involvement. Due to its rarity and the potential for systemic manifestations, the anesthetic management of patients with H syndrome presents unique challenges and requires a tailored approach. Facial and neck abnormalities, including sclerodermatous thickening and hypertrichosis makes airway management more challenging. Developing an individualized anesthetic plan based on the patient's specific H syndrome manifestations and comorbidities will help in successful management of these cases. Assessing the patient for comorbidities and potential organ involvement is essential as it might impact anesthesia and postoperative care due to multisystem involvement. This article aims to provide a comprehensive guide to anesthetic considerations and strategies for H syndrome patients. Herewith presenting a case of H syndrome in a 3 year old child with diagnosed H syndrome for Ludwigs angina for anesthesia management and airway management.

Key Words: Anesthetic management, difficult Airway, H syndrome.

Received: 10 November 2023**, Accepted:** 09 February 2024

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ISSN: 2090-925X, 2024, Vol.16, No. 2, July 2024, 956–964 © Ain-Shams Journal of Anesthesiology

INTRODUCTION

H SYNDROME is a rare autosomal recessive genodermatosis, characterized by various comorbidities posing multiple challenges to the managing anesthetist. Here, we report a case of H syndrome in a child with Ludwig's angina.

A 5-year-old male child diagnosed with H syndrome 3 years ago currently presented with complaints of fever and swelling in the floor of the mouth extending bilaterally for 7 days associated with pain during swallowing. A week before the presentation, pus was aspirated from a presumed dental abscess. He had been taking regular medications type 1 diabetes mellitus and hypothyroidism. On examination, tenderness was observed in the submandibular space Figures 1 and 2. Airway examination revealed limited mouth opening to one finger breadth associated with restricted neck extension and reduced compliance of submandibular space. Investigations revealed a hemoglobin of 12 g dl-1 and random blood sugar of 280 mg dl-1 without evidence of ketosis and was controlled with insulin. The thyroid profile was within normal limits. Although the chest radiograph was normal, the neck examination showed reduced upper airway space due to swollen soft tissues Figure 3. As a result, emergency incision and drainage were planned under General anesthesia.

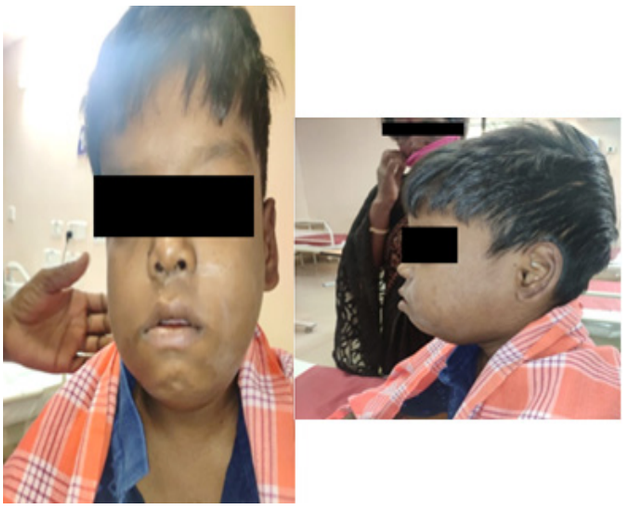

Figure 1 and 2 : Front and lateral view of the face, second figure showing swelling in the left cheek due to Ludwig's angina.

A 24-Gauge peripheral IV cannula was secured in the dorsum of the left hand with difficulty prior to induction of anesthesia. Premedication included intravenous injections of glycopyrrolate (150 mcg) and dexamethasone (3 mg). Standard ASA monitors such as ECG, non-invasive blood pressure, pulse oximetry and capnography were used and

DOI: 10.21608/ASJA.2024.247718.1003

Difficult airway cart was prepared. General anesthesia was induced with incremental doses of sevoflurane of up to 4 % in 100 % O2 and Inj.Fentanyl 30 mcg and Inj. Propofol were administered intravenously in aliquots of up to 20 mg to deepen the plane of anaesthesia and spontaneous ventilation was maintained. Check laryngoscopy was done using a TUOREN video laryngoscope size 2 blade. Cormack Lehane grade 2B with edematous vocal cords allowed a gum elastic bougie aided intubation with cuffed endotracheal tube of size 5. After confirming the endotracheal tube position, Inj.Atracurium 7.5 mg was administered. Ventilation was managed in volume-controlled mode (tidal volume: 140 ml, respiratory rate: 15 breaths/min, PEEP: 4 CmH2O, Fio2: 0.4). Intra-operative blood sugars were maintained within normal range. The surgery lasted approximately 30 minutes. Due to airway edema, extubation was deferred and the child was transferred to the pediatric intensive care unit (PICU) for postoperative mechanical ventilation. Postoperatively, the child received antibiotics, steroids and adrenaline nebulization, in addition to glycemic control. Extubation took place on postoperative day 2 after an air leak test.

H SYNDROME is an autosomal recessive genodermatosis and is a novel form of histiocytosis[1], characterized by cutaneous hyperpigmentation, hypertrichosis, hepatosplenomegaly, hearing loss, cardiac anomalies, hypogonadism, hyperglycemia (Diabetes Mellitus 1), short-stature flexion contractures and hallux valgus. H Syndrome is caused by mutation of nucleoside transporter hNET3, which is included under SLC29A3[3].

The anesthesia concerns in a case of H syndrome are as follows[1 - 6]. Due to facial and neck abnormalities, including sclerodermatous thickening and hypertrichosis, airway management is challenging. We should be prepared for possible difficult intubation or ventilation. Ensure the patient is on appropriate thyroid hormone replacement therapy and monitor thyroid function before planning for anesthesia due to the presence of hypothyroidism. Maintain strict glycemic control with insulin as needed to avoid hyperglycemia. Due to sclerodermatous thickening and potential vascular abnormalities, securing intravenous access can be difficult. Consider using ultrasound guidance or alternative access points (e.g., central venous catheters) if peripheral access is challenging. Patients with H syndrome may have an increased risk of infection due to hyperglycemia and immune system challenges. Assess the patient for comorbidities and potential organ involvement that might impact anesthesia and postoperative care due to multisystem involvement. Develop an individualized anesthetic plan based on the patient's specific H syndrome manifestations and comorbidities. Due to the risk of airway edema and potential complications, closely monitor the patient in the postoperative period, especially in cases where extubation is delayed. Transfer to a pediatric intensive care unit (PICU) or an appropriate critical care setting depending on the case Figure 4.

In our case, airway management becomes more challenging because of the following factors such as pediatric population due to their unique airway anatomy, presence of H syndrome along with Ludwig's angina[4]. Central venous cannulation was not required in our case, manageable with peripheral intravenous access. Early recognition and prompt management is the key to prevent such a life-threatening airway obstruction[5]. Predisposing factors must be looked for and appropriate management such as the administration of appropriate antibiotics, the maintenance of a patent airway and surgical intervention such as incision and drainage must be performed[6]. Conduct a thorough preoperative assessment to identify specific challenges or concerns related to the patient's H syndrome and tailor your anesthesia plan accordingly.

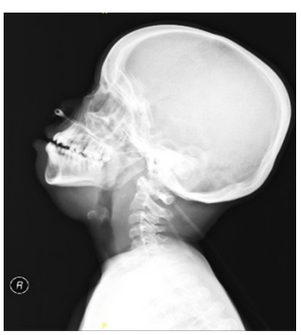

Figure 3: The lateral view reveals reduced upper airway space due to soft tissue swelling.

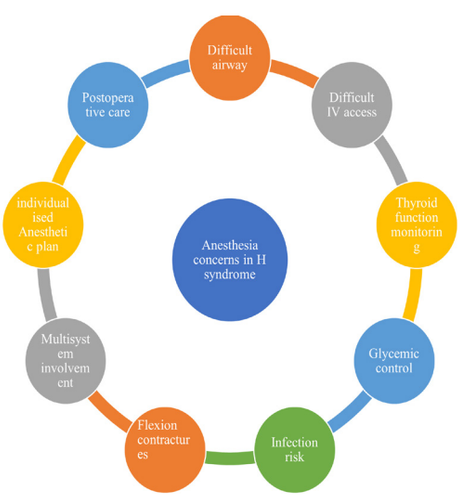

Figure 4: Diagram depicting the Anaesthesia concerns involved in H syndrome.

CONFLICT OF INTEREST

There are no conflicts of interest.

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