ORIGINAL RESEARCH PAPER

Anaesthesiology

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AIRWAY MANAGEMENT OF A CHILD WITH HUNTER'S SYNDROME

Dr Yaram Reddy Susmitha* Postgraduate, Department of Anaesthesia, Saveetha Medical College and Hospital,Thandalam. *Corresponding Author

Dr K.V.L Sanjana Assistant Professor, Department of Anaesthesiology, saveetha Medical College and Hospital,Thandalam.

Dr Vivekanand P Postgraduate,Department of Anaesthesiology, saveetha Medical College and Hospital,Thandalam.

Dr Lakshmi Ramakrishnan Professor and Head of the Department, Department of Anaesthesia, Saveetha Medical College and Hospital,Thandalam.

ABSTRACT

Hunter's syndrome, a very rare genetic disorder caused by a missing or malfunctioning enzyme iduronate-2-sulfatase, resulting in the accumulation of glycosaminoglycans in lysosomes. We report the airway management of an 8-year-old male child diagnosed as Hunters syndrome came with complaints of shortness of breath, lower respiratory tract infection and acute exacerbation of asthma. Admitted in pediatric ICU with severe wheeze and respiratory distress. Started on HFNC. Despite HFNC symptoms got worsened. So, planned for elective intubation. Child has large, protruded tongue, short neck. Difficult airway cart made ready. One attempt of video laryngoscopy and one attempt of normal laryngoscopy done. Unable to intubate so proceeded with tracheostomy as child was desaturating.

INTRODUCTION

Hunter syndrome, also called mucopolysaccharidosis type II, is a rare X-linked metabolic disease caused by a deficiency of iduronate-2-sulfatase, a lysosomal enzyme that cleaves glycosaminoglycans (GAGs) resulting in accumulation of glycosaminoglycans in tissues. It affects various organs, including the airways, heart, and central nerves. In children with MPS type 2, the risk of airway obstruction during anesthesia/sedation is high, and the degree of difficulty increases with aging. Furthermore, this disorder may cause heart valve disease, cardiomyopathy, coronary artery stenosis, and disturbance of the conduction system.

Features of Hunters syndrome include prominent mandible, immobile jaw, macroglossia, irregular shaped teeth, narrowed nasal passages, short neck, hypertrophied tonsils and adenoids, tracheal distortion, tracheomalacia, thickened and copious secretions, kyphoscoliosis, abnormalities of ribs shape and structure, restrictive pulmonary disease, redundant respiratory epithelium, airway edema due to recurrent airway disease, cardiac manifestations, abdominal organ enlargement.

CASE REPORT

Here we report the airway management of an eight-year-old male child of 35kg weight diagnosed with Hunters syndrome. Child got admitted with complaints of acute exacerbation of asthma, lower respiratory tract infection. On second day of admission symptoms worsened with severe wheeze and tachypnoea. So shifted to pediatric ICU and Started on HFNC with FiO2 45% @ 35litres/min. Despite of HFNC symptoms worsened. So planned for emergency intubation with ENT team backup. As necessary equipment's were not available in pediatric ICU planned to intubate in operation theatre. Child had large head, hypertrophied protruded tongue, thick lips, short neck. As child has social and language developmental milestone delay mouth opening could not be assessed. Child shifted to OR with SPO2 of 85% with 15litres/min O2, RR-50/min, HR-130/min. After pre-oxygenation, sedated with inj. Dexmedetomidine - 25mcg slow IV, inj. Fentanyl-25mcg, inj. ketamine-25mg IV, sevo-1%. Video-laryngoscopy was done, able to visualize thick epiglottis but could not pass bougie. Again, preoxygenated and normal laryngoscopy done, epiglottis was seen but unable to intubate. Patient desaturated to SPO2 of 40%. Child can be able to ventilate, so proceeded with emergency tracheostomy for definitive airway.

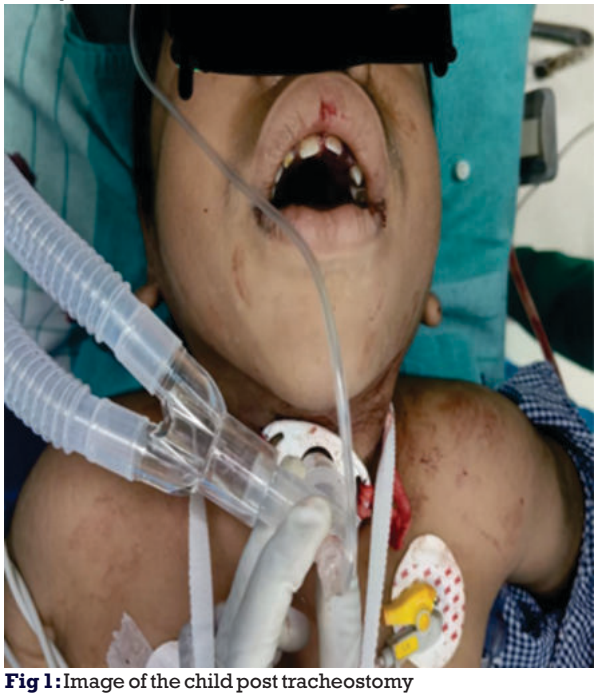

Fig 1: Image of the child post tracheostomy

DISCUSSION

Patients with Hunters syndrome usually present with difficult airway because of mucopolysaccharide tissue infiltration in the upper and middle airways, limited TM joint movement, macroglossia, and frequent upper respiratory tract infections (Chatterjee et al., 2017). We used ketamine and sevoflurane for maintaining spontaneous respiration. Inability to ventilate the lungs after giving muscle relaxant has been reported in these patients, so avoided muscle relaxant (Kaur et al., 2012). Walker et al. found the use of LMA to be extremely useful both to provide a secure airway for short procedures and to clear the obstructed airway in many cases of mucopolysaccharidoses (Walker et al., 2013). Airway in a patient of hunter syndrome can be secured by placing a larger video laryngoscope blade at posterior surface of epiglottis to view

posterior part of larynx to facilitate bougie guided intubation (Punj et al., 2019). In an emergency, tracheostomy may be the choice to secure the airway. Cricothyroidotomy is not recommended for mucopolysaccharidosis patients whose cricothyroid membrane, cricoid and thyroid cartilages are often thickened and deformed by mucopolysaccharide deposits, making rapid dissection difficult and vocal cord damage likely.

CONCLUSION

To conclude prompt recognition of the compromised airway in patients with Hunter's syndrome is important and can guide airway management.

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