OP18 MAROTEAUX – LAMY SYNDROME: AN AIRWAY NIGHTMARE

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Introduction

Maroteaux-Lamy syndrome is a manifestation of mucopolysaccharidosis type VI (MPS VI) disease. It is an autosomal recessive disorder caused by arylsulfatase B deficiency. This report details one such case encountered in our department.

Case description

A 52-year-old gentleman presented to emergency department (ED) with breathlessness and noisy breathing. He was recently diagnosed to have chronic obstructive airway disease (COAD) and had life-threatening exacerbation of COAD on presentation. He did not respond well to medical therapy, including non-invasive ventilation and intubation was attempted. However, he failed intubation and supraglottic airway (SGA) was used. The Ear, Nose and Throat (ENT) team subsequently performed tracheostomy but had difficulty because the strap muscle was adhered to the trachea, and it was hard to separate the pre-tracheal fascia from tracheal cartilage. Palpating the overlying structures was also arduous. Contrast-enhanced computed tomography (CECT) of neck and thorax showed edematous upper airway till oropharynx as well as the trachea inferior to the tracheostomy site was collapsed and both bronchi were narrowed. Bronchoscopy revealed tracheomalacia and similar findings to CECT. Bronchoscopy with tracheal stenting of the stenosis segment was done under general anesthesia. MPS was apprehended as this patient presented with coarse facial features, joint stiffness, decreased pulmonary function and hepatomegaly. Blood and urine samples were performed which indicated he had low levels of serum arylsulfatase B. However, hearing, visual, and cardiac assessment were normal in this gentleman.

Discussion

MPS is a rare metabolic disease which is further classified to seven types based on the deficiency of the lysosomal enzyme. Difficult airway assessment and management of anticipated difficult tracheal intubation should be implemented in this patient. However, in this patient 6 attempts were made using video laryngoscopy engender the upper airway edema.

Conclusion

Tracheomalacia is rare in adults thus we should perpetuate clinical guide of inkling in diagnosing MPS. Intubation in this type of patient should be done in a controlled environment by experts such as the ENT team. SGA could support the oxygenation prior to tracheostomy and definitive management for the patient.

Keyword: Maroteaux-Lamy syndrome, difficult intubation, tracheomalacia