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The Complication of Endotracheal Intubation in a Patient with Mucopolysaccharidosis Type IIIA

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Abstract

Mucopolysaccharidosis type III (MPS III), also known as Sanfilippo syndrome, is an autosomal recessive, neurodegenerative lysosomal storage disorder. Substantial challenges for airway management and endotracheal intubation are predictable due to the MPS patients' specific phenotypic, facial, and airway characteristics. In studies concerning various types of MPS, the incidence of difficult endotracheal intubation ranges between 28%-44%. This study intends to present a case with MPSIIIA, who aspirated her tooth during endotracheal intubation due to acute respiratory failure. This study reports a 12.5 year-old girl who presented with MPSIIIA and aspirated her tooth during endotracheal intubation due to acute respiratory failure resulting in lung lobe segmentectomy. The majority of MPS patients' intubation and airway maintenance are hard and get even more difficult by age because of their mental retardation and oral health deteriorating, which may require tracheostomy.

***Key Words*:** Aspiration, Child, Difficult Airway Maintenance, Intubation, MPS Type IIIA.

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1- INTRODUCTION

Mucopolysaccharidosis type III (MPS III), also known as Sanfilippo syndrome, is an autosomal recessive, neurodegenerative lysosomal storage disorder. MPS III is a progressive disorder specifically affecting the central nervous system and presents itself with delayed speech, hearing loss, and behavioral problems (1, 2). MPS III is divided into four types based on the genetic cause: IIIA, IIIB, IIIC, and IIID. The various MPS III types represent similar signs and symptoms, although MPS IIIA typically represents its features earlier in life and has a rapid progression (1, 2). Substantial challenges for airway management and endotracheal intubation are predictable due to the MPS patients' specific phenotypic, facial, and airway characteristics. In studies concerning various types of MPS, the incidence of difficult endotracheal intubation ranges between 28%-44%. (3). Here, this study reports a 12.5-year-old girl with MPS IIIA, who aspirated her tooth during endotracheal intubation due to acute respiratory failure.

2- CASE REPORTS

The patient was born to consanguineous parents (3rd-degree cousins) after a full-term pregnancy by typical vaginal delivery with typical birth weight. According to her parents, she had a developmental delay from birth. The patient began to hold her head at the 5th month, sat without support at the 10th month, and walked at two years old. She could speak a few words but never spoke fluently. Firstly, she was evaluated at our hospital by the pediatric metabolism and nutrition department at three years. At this time, her examination revealed pectus carinatum, mild coarse face, mild hepatomegaly, mild motor delay, and mild distal spasticity in all extremities. Her metabolic screening showed increased heparin and heparin sulfate in the urine. Low sulphamidase activity in white cells and low beta-galactosidase activity led to the diagnosis of MPS IIIA. There is no family history for the same or similar disorders. When she was five years old, she suffered from recurrent convulsions, anxious and aggressive behavior without electroencephalography abnormality. The brain magnetic resonance imaging showed cerebral atrophy. The convulsions resulted in the rapid loss of neurocognitive functions. The patient has not been able to feed himself since she was five years old, walk since nine years old, sit without support since last year, and has not had eye contact and facial expressions for the last six months. The patient was treated twice under general anesthesia because of dental problems without post-anesthesia complications. The patient had concomitant swallowing dysfunction, vitamin D deficiency, epilepsy, mild mitral valve prolapses, cerebral atrophy, and scoliosis. She has been fed by gastrostomy because of deteriorating swallowing functions that caused more than three pneumonias in the last year.

Just before the patient was referred to our hospital, she was admitted to another center with cyanosis. She was intubated at the center due to acute respiratory failure and then referred to our hospital. On admission, she was intubated and had extensive crackles in both lungs, she had tachycardia and hypotension. Chest X-ray showed bilaterally chronic changes and right lower lobe consolidation. The patient was extubated and followed with non-invasive mechanical ventilation. Due to septic shock and increased carbon dioxide retention, she was reintubated. The patient received appropriate support and antibiotic treatments. Tracheostomy was opened due to failed repeated extubating attempts. Despite appropriate antibiotic treatment, the right lower lobe consolidation enlarged on the chest X-ray, and the patient did not clinically improve. Therefore, thorax computed tomography was performed, which showed a hyperdense foreign body measuring 7 × 7.5 mm at the bifurcation

level of the lower lobe bronchus of the right lung (**Figures. 1a, b**). When the patient's chest X-rays were re-evaluated, the appearance suggested that there was a 1-2 mm radiopaque foreign body at the level of the lower lobe of the right lung that can be noticed on the following chest X-rays, which was not present on the first hospitalization X-ray (**Figures. 2a, b**). The patient underwent rigid bronchoscopy for foreign body removal. Due to distal localization of the foreign body, it was not reachable by bronchoscopy, and henceforth right lower lobe segmentectomy was performed. The foreign body extracted from the parenchyma seems to be a tooth. After all, the patient was discharged without oxygen support with a tracheostomy.

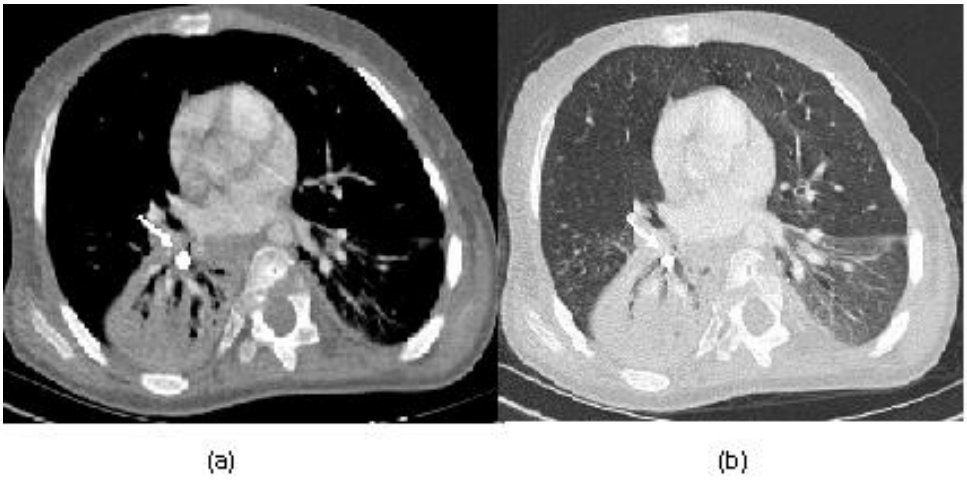

Fig.1: CT-scan demonstrate the presence of radiopaque material at the level of bifurcation of the lower lobe bronchus of the right lung (arrow).

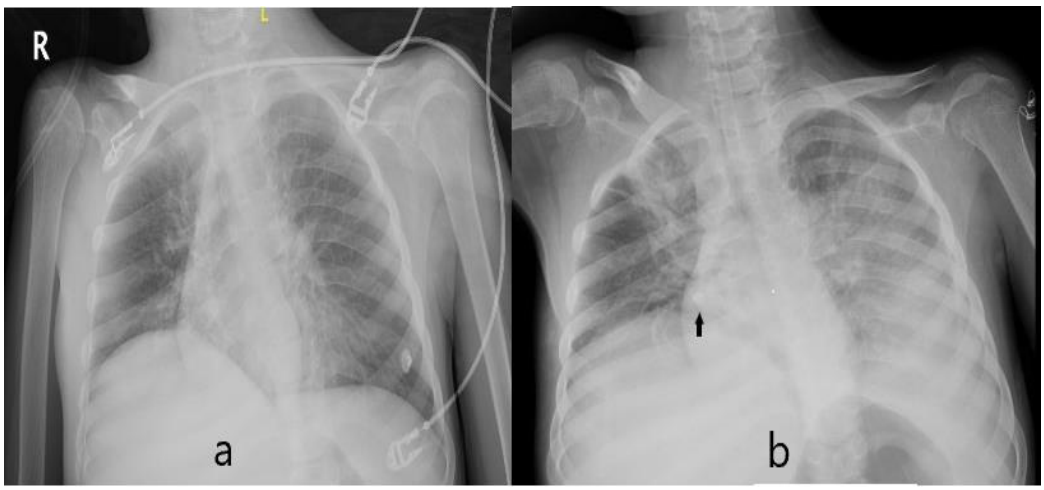

Fig. 2 (a-b): Chest Xray demonstrate the presence of a 1-2 mm hyperdense foreign body at the level of the lower lobe of the right lung (arrow) after the endotracheal intubation (b) which was not present on the first X-ray (a).

3- DISCUSSION

MPS IIIA is the most severe type of MPS III and is the most common subtype in northern Europe. MPS IIIA is characterized by severe central nervous system degeneration with mild somatic symptoms. Neurological symptoms include marked hyperactivity with aggressive behavior, delayed development, mental retardation, hearing loss, seizures, and vision loss. In the early stages of the disease, patients usually represent mild somatic symptoms, such as coarse facial appearance, skeletal involvement, and hepatosplenomegaly. MPS IIIA patients may also have severe diarrhea or constipation, narrow airway passage, enlarged tonsils, adenoids, swallowing dysfunctions, conductive hearing loss due to recurred otitis media, and recurring respiratory infections. There is currently no therapy available for this disease (1, 2). The accumulation of glycosaminoglycan in the upper airway and skeletal involvement makes endotracheal intubation difficult for mucopolysaccharidosis. Restricted mouth opening, Mallampati scores of 3 and 4, and recurrent adenoid hypertrophy make MPS patients difficult to intubate (4). The studies showed that MPS patients have a higher prevalence of dental caries, poor periodontal health, malocclusion, and delayed eruption of teeth, cystic lesions or enlargement of the dental follicle because of their oral features; gingival hyperplasia, diastemas, macroglossia, high palate and condylar defects (5, 6). Therefore, MPS patients can easily lose their teeth even with minor trauma and can aspirate them during the intubation. Several studies have documented a progressive increase in the incidence of a difficult airway with aging in MPS patients. Madof et al. (7) showed a clear progression toward difficult intubation as the MPS patients got older. Similarly, Scaravilli et al. (8) reported that 29% of the intubations were difficult for MPS patients, and older age was associated with a higher risk for difficult intubation. The patient in this study received general anesthesia twice in her early life without any complications but needed tracheostomy later. While patients with MPS have a high incidence of difficult ventilation and endotracheal intubation in general, MPS III patients have a lower incidence compared with other types of MPS. Many studies support that the intubation and airway problems of MPS III patients can be resolved with simple maneuvers (9-11). A prospective review of MPS IIIA patients by Cingi et al. (12) revealed that airway management in 25 patients who underwent a total of 94 anesthetics was all easy. The study conducted by Cohen et al. (13) on 44 MPS III patients demonstrated that a difficult airway is unlikely when anesthetizing an MPS III patient, although the risk may remain. Eventually, many of the MPS patients require tracheostomy. Tracheotomy is also a difficult operation in MPS patients. One study documented that patients with MPS II develop progressive upper airway obstruction and tracheostomy remains an important intervention to safeguard the airway (14). In that study, the mean age of tracheostomy requirement was found as 11 years and 2 months. Another MPS II patient was reported in a study who developed sudden respiratory distress in the emergency department and required tracheostomy as endotracheal intubation (15). Our patient also needed a tracheostomy as she could not tolerate extubation. In conclusion, intubation and airway maintenance in the majority of MPS patients are difficult. This difficulty tends to get aggravated by age due to their mental retardation and oral health deterioration. As our case demonstrates, clinicians should take care during intubation and keep in mind that complications, such as tooth aspiration due to difficult intubation may occur more frequently in MPS patients, resulting in segmentectomy.

4- CONCLUSION

In conclusion, the majority of MPS patients' intubation and airway maintenance are difficult and gets even more difficult by age because of their mental retardation and oral health gets worse. As our case demonstrates, clinicians should take care during intubation and keep in mind that complications due to difficult intubation may occur more frequently such as tooth aspiration in MPS patients which resulted with segmentectomy.

5- CONFLICT OF INTEREST: None.

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