CASE REPORT

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A Complete Tracheal Cartilage: An Unexpected Difficult Airway in an Adult with Down Syndrome

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ABSTRACT A congenital complete tracheal ring is an infrequent cause of tracheal stenosis. It is usually diagnosed in infants and young children, and adults diagnosed with complete tracheal rings are even rarer entities. There is limited literature regarding the adult complete tracheal ring. Down syndrome patients have a wide array of documented anatomic variations especially airway abnormalities such as laryngomalacia, tracheomalacia, tracheal bronchus and bronchomalacia and rare to have complete tracheal ring. Hence, we described a Down syndrome adult who remained asymptomatic until the recent onset of severe respiratory illness, which requires endotracheal intubation, leading to an unexpectedly difficult airway. A differential diagnosis of a complete tracheal ring should be considered when attending such cases of difficult intubation.

Keywords: Down syndrome; chromosome 21; congenital trachea stenosis; intubation; bronchus

Complete or near complete tracheal ring formation is a rare occurrence first described in 1897.[1] The patients usually present with respiratory distress in the first year of life.[2] The severity of symptoms is related to the length and degree of the narrowing, cardiovascular abnormalities, and the age of presentation.[3] There is a paucity of literature on the complete tracheal ring in the adult population. Most of the literature regarding the adult complete tracheal ring is incidental findings during elective intubation for operation. Down syndrome is the most common chromosomal disorder and airway abnormalities are responsible for significant mortality and morbidity in Down syndrome patients.[4] We describe a Down syndrome patient with an unexpectedly difficult airway secondary to a complete tracheal ring found during recent bouts of severe respiratory infection requiring endotracheal intubation.

CASE REPORT

Informed consent was obtained from the patient in this case report.

A 22-year-old patient with Down syndrome and morbid obesity presented to the emergency department with a complaint of shortness of breath, fever, and chesty cough for one month. She was tachypneic with an oxygen saturation under room air of 78% and lung auscultation had generalized reduced air entry. The total white cells and the C-reactive protein were elevated, while the arterial blood gas showed a type two respiratory failure with carbon dioxide narcosis. The chest X-ray showed pneumonia features. The patient was treated for sepsis secondary to community-acquired pneumonia with type two respiratory failure and carbon dioxide narcosis. Due to the impend-

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ing respiratory failure despite non-invasive ventilation, a decision for intubation was made. She was given ketamine, fentanyl, and rocuronium for rapid sequence induction. The patient was successfully intubated with an endotracheal tube (ETT) size 5.5 mm after multiple intubations attempt due to difficulty pushing the ETT beyond the subglottic region.

Tracheostomy was performed. During the procedure, there were difficulties inserting the tracheostomy tube size 7.0 mm, 6.5 mm, and armored ETT size 6.0 mm as the tubes were unable to pass through the distal part. A tracheostomy tube size 5.0 mm was inserted eventually. The airway study was done in the same setting with findings of a circumferential stenotic segment seen 23 mm beyond the vocal cord. A high-resolution computed tomography (CT) thorax was performed, showing no trachea extraluminal compression. Alas, the scan could not describe the presence of tracheal stenosis as there was an ETT in situ (Figure 1). A repeated airway study was performed subsequently after reviewing the scan. Intraoperatively, there was presence of a circumferential tracheal ring which was 23 mm below the vocal cord until just above the carina. Airway sizing was performed and equal to ETT 5.0 mm. The right tracheal bronchus was also present (Figure 2, Figure 3). The trachea mucosa was healthy. ETT was removed from the tracheostoma and proceeded with endotracheal intubation with ETT 5.0 mm. She was referred to cardiothoracic surgery for surgical intervention, however the family refused, and she was kept intubated for about one-month duration. After the respiratory infection settled, she was extubated and managed to be on room air with normal breathing.

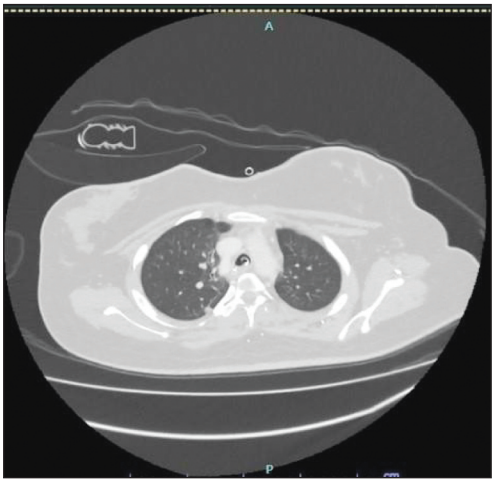

FIGURE 1: A computed tomography scan in axial view showed an in-situ endotracheal tube that occupies the whole tracheal lumen, which makes it difficult to assess presence of the complete tracheal ring.

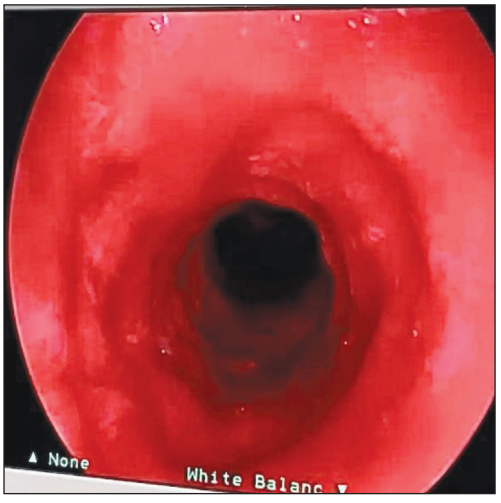

FIGURE 2: Rigid tracheobronchoscopy showed a concentric tracheal ring, that extends from the first tracheal ring until just above the carina.

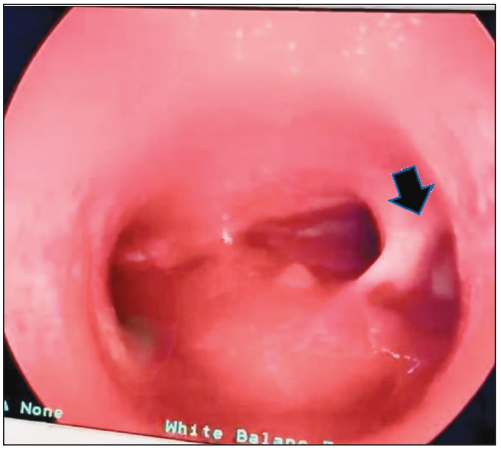

FIGURE 3: A view from rigid tracheobronchoscopy showed a right tracheal bronchus, as depicted by arrow.

DISCUSSION

Congenital tracheal stenosis is one of the common diagnoses when an infant presents with biphasic stridor, which is often exacerbated following an upper respiratory infection.[5] It is often associated with pulmonary slings, tracheoesophageal fistula, esophageal

atresia, VACTERYL syndromes, cardiac abnormalities, Pfeiffer's syndrome, and scoliosis.[5]

Down syndrome is the commonest human chromosomal syndrome, with an incidence rate of one in every 700 babies born.[6] The average life expectancy in Down syndrome patients is 47 years old.[6] Airway abnormalities are responsible for significant morbidity and mortality in children with Down syndrome and the commonest are laryngomalacia, tracheomalacia, tracheal bronchus, and bronchomalacia.[4,7] There is no clear embryological etiology for the association between Down syndrome and tracheal anomalies.[4]

The complete tracheal ring consists of an O-shaped ring. It is seen in the middle or distal part of the trachea, ranging from a few centimeters to the whole tracheal length.[5] It can present directly after birth with symptoms such as stridor, wheezing, exertional dyspnea, cyanosis, and respiratory distress.[8] Patient may also become asymptomatic, as seen in this patient and usually been discovered incidentally during elective endotracheal intubation, due to difficulty in advancing the ETT.[4] Undoubtedly, it is a rare cause of tracheal stenosis in adults as the patients often present in the first year of life.[2] There are only 14 reported literatures on the complete tracheal ring discovered in adults, with eight of them found during failure of the endotracheal intubation. There is a paucity of literature about life expectancy in complete tracheal ring patients. However, the oldest patient with a complete tracheal ring is 70 years old.[9]

The tracheal narrowing caused by the complete ring may increase the risk of recurrent respiratory infection and respiratory compromise in severe cases.[2] Down syndrome patients are prone to respiratory tract infections and lung injury due to lung parenchymal abnormalities, as compared to other patients.[4] Moreover, a complete tracheal ring is usually associated with an abnormal bronchial branching pattern, particularly the trachea bronchus, seen in this patient.[10]

Endoscopic examination is the gold standard for evaluating anomalies of the trachea, as seen in this patient.[1] It allows for the differentiation between the stenosis and the complete tracheal ring with the letters having no pars membranacea and enables visualization between intraluminal and extraluminal tracheal compressions.[11]

The radiological investigation, which is CT with 3- dimensional reconstructions, can aid in diagnosing and monitoring patients with the completetracheal ring. It can differentiate a complete tracheal ring from stenosis as the complete tracheal ring will have an O-shaped configuration and absence of the airway wall thickening, which are not seen in trachea stenosis.[12] Unfortunately for this patient, we could not visualize the characteristic findings of the complete tracheal ring as the ETT is in-situ.

Patients can either be managed conservatively or through surgery, depending on the symptom severity. Conservative management is reserved for asymptomatic or mildly symptomatic patients, as the observation period will often help in determining whether the patient requires tracheoplasty.[13] Slide tracheoplasty is the treatment of choice for severe cases of complete tracheal rings.[5] It is a versatile approach to long-segment tracheal stenosis, and previous studies have demonstrated a low mortality rate between 5 to 20%.[14] Endoscopic approaches such as laser, balloon dilatation, and stenting may be considered when an open approach is not advisable.[5]

In conclusion, this is a peculiar case as this Down syndrome patient remains asymptomatic for 22 years and is only diagnosed following a severe episode of respiratoryinfection. An attending physician should be aware of this possible upper airway abnormality when attending to an adult patient with Down syndrome who presented with respiratory symptoms.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Siti Farhana Abdul Razak, Marina Mat Baki, Siti Hajar Sanudin;* ***Design:*** *Siti Farhana Abdul Razak, Yazid Pol Ong, Marina Mat Baki, Siti Hajar Sanudin;* ***Control/Supervision:*** *Siti Farhana Abdul Razak, Yazid Pol Ong, Marina Mat Baki, Siti Hajar Sanudin;* ***Data Collection and/or Processing:*** *Siti Farhana Abdul Razak, Yazid Pol Ong, Siti Hajar Sanudin;* ***Analysis and/or Interpretation:*** *Siti Farhana Abdul Razak, Yazid Pol Ong, Marina Mat Baki, Siti Hajar Sanudin;* ***Literature Review:*** *Siti Farhana Abdul Razak, Siti Hajar Sanudin;* ***Writing the Article:*** *Siti Farhana Abdul Razak, Yazid Pol Ong, Marina Mat Baki, Siti Hajar Sanudin;* ***Critical Review:*** *Siti Farhana Abdul Razak, Yazid Pol Ong, Marina Mat Baki, Siti Hajar Sanudin;* ***References and Fundings:*** *Siti Farhana Abdul Razak, Marina Mat Baki, Siti Hajar Sanudin;* ***Materials:*** *Siti Farhana Abdul Razak, Marina Mat Baki, Siti Hajar Sanudin.*

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