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Severe Tracheomalacia Secondary to Double Aortic Arch. Any Role in Posterior Aortopexy? A Case report

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ABSTRACT

Tracheomalacia is the abnormal collapse of the tracheal lumen and is common after cardiac compression. In the severe types, tracheomalacia can lead to significant morbidity, especially if encountered in a critical area, such as the distal part of the trachea, immediately above the tracheal bifurcation. We report a case that did not improve after cardiac surgery with persistent airway narrowing of more than 90%, requiring ventilatory support for a year. The case was difficult to manage, but the patient eventually showed a significant improvement after a **posterior aortopexy** that helped a lot in weaning him off the ventilator with a residual persistent audible wheeze and recurrent chest exacerbation. The diagnostic and therapeutic options for the case will be discussed.

Keywords: malacia, tracheomalacia, double aortic arch, stridor, wheeze and aortopexy.

INTRODUCTION

Malacia is an abnormal collapse of the airway walls. There are three main types of malacia: **laryngomalacia, tracheomalacia and bronchomalacia**. The distal third of the trachea is most commonly affected in tracheomalacia. Tracheomalacia appears to be related to a developmental defect in the cartilage of the tracheal wall; the lesion typically occurs in infants and young children. In most children, the tracheal cartilage normalizes as the airway enlarges, and symptoms may resolve in late childhood. Clinical symptoms can range from minor expiratory stridor with typical barking cough, recurrent/persistent wheezy chest, and severe respiratory distress episodes to apnea [1].

Case report:

Patient information.

An 18-month-old boy, born at term through a spontaneous vertex delivery, presented respiratory distress, which was diagnosed as transient tachypnea of the newborn soon after delivery.

Clinical finding.

On examination, two days after birth, he had marked respiratory distress and inspiratory stridor. Additionally, the patient required CPAP followed by intubation and mechanical ventilation due to a high PCO_2 level.

Diagnostic assessment

The patient screen was positive for RSV bronchiolitis during the peak of the winter season. Nonetheless, since the stridor is not a common manifestation of bronchiolitis, a flexible bronchoscope of 2.8 mm in size was introduced under light anesthesia through ETT. In the first time that the procedure was performed in PICU, the finding was a marked narrowing of the airway immediately above the carina with the recommendation to roll out vascular compression [Fig. 1]. An upper GI study [Fig. 2] showed indentation of the esophagus supporting the diagnosis of a vascular ring. Another bronchoscopy showed severe tracheomalacia with more than a 90% narrowing of the tracheal lumen with pulsatile airway approximately 1 cm above the carina (length 1.5 cm). The finding of a complete vascular ring was confirmed by CT chest and CT angiogram, which showed a dominant posterior right-sided aortic arch and a smaller anterior left-sided aortic arch [Fig.3 and Fig. 4].

Therapeutic intervention. The patient was managed in the PICU, where he was ventilated for 1 year with an unusual treatment, during which he had multiple failures that prevented extubation or even weaning of the ventilatory support. He underwent a vascular ring repair 'resection of the anterior aortic arch". Although he showed good signs of recovery, he unfortunately failed extubation, despite being on a ventilator at a low setting. This case was discussed during the Saudi Pediatric Pulmonology Cross Country Round, a multiple case conference with an airway expert in the international medical center "Grand Ormond Street Hospital." The recommended options were stenting, tracheostomy, and posterior aortopexy. Each option has its pros and cons, but the majority advised for posterior aortopexy. Repeated CT angiogram and bronchoscopy showed severe narrowing of the airway, approximately 80%, with a marginal improvement in the narrowing of the airway lumen. **Posterior aortopexy** occurred, and the patient showed significant improvement and was

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able to be extubated to CPAP until he weaned off gradually to room air in several weeks. He had a persistent audible wheeze after any excitation, but he was thriving well, achieving normal milestones. Currently, the patient is at home doing fine, suffering only from recurrent admissions due to a persistent wheezy chest with a regular follow-in the clinic for the expected residual moderate tracheomalacia. A third bronchoscopy was done (at 18 month of age) to further evaluate a persistent wheeze. A 70% narrowing of the tracheal lumen was documented with no strong response to an increasing PEEP, as expected in malacia, luckily the child condition continued improving, and he is thriving well, managed at home with inhaled corticosteroid, anti reflux therapy and occasional Ventolin as needed, with aggressive chest physiotherapy especially with flu illness with lower threshold for antibiotic in treating chest infection.

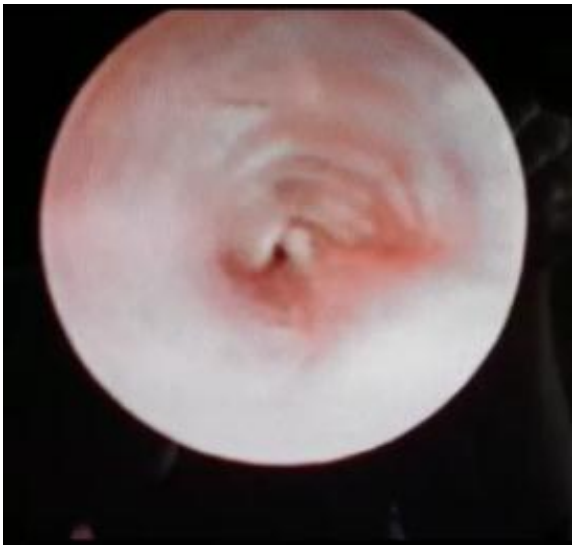

Figure 1: Bronchoscopy picture: 2.8 mm Olympus showing marked narrowing of the airway lumen >90% just above the carina, pulsatile compression through the video

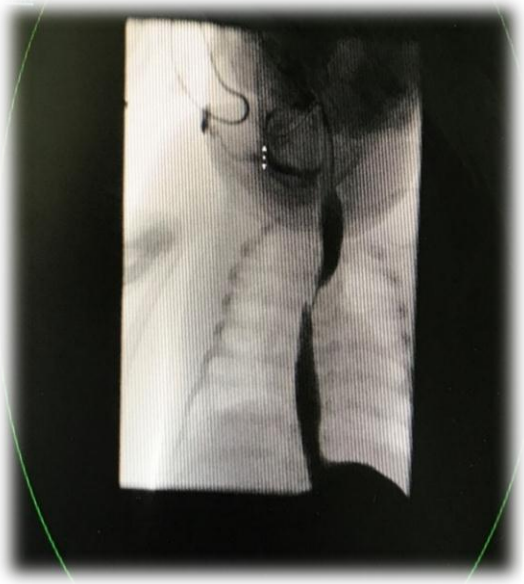

Figure 2: Barium meal showing bilateral indentation of the esophagus at mid portion. Consistent with double aortic arch with high grade reflux.

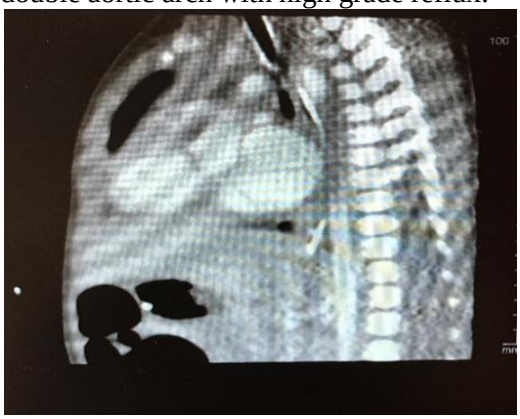

Figure 3: Cat Scan- sagittal view of the lung showing deep ETT to the carina and aortic compression with (NG tube in situ).

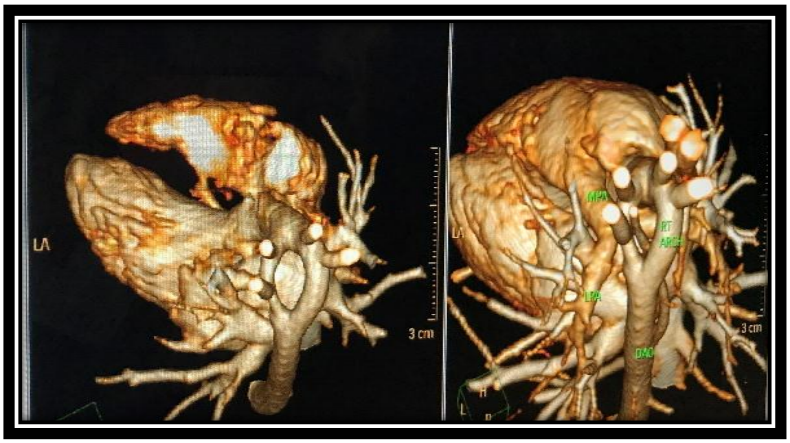

Figure 4: CT Angio confirming the complete vascular ring by the double aortic arch with posterior predominant arch.

Time Table

Medical history and final resolution of the case

A newborn male born at full term through a spontaneous vertex delivery developed respiratory distress soon after delivery. Later, it was confirmed to have tracheomalacia due to a vascular ring.

Current illness, physical evaluation, diagnosis

The patient admitted to NICU to treat his respiratory distress required mechanical ventilation to support his respiratory status, with marked respiratory stridor.

A flexible bronchoscope of 2.8 mm in size [Fig.1] was introduced under light anesthesia through ETT. In the first time that the procedure was performed in PICU, the finding was a marked narrowing of the airway just above the carina.

An upper GI study [Fig.2] showed an indentation of the esophagus supporting a vascular ring diagnosis.

A chest CT angiogram [Fig.4] showed severe tracheomalacia with more than 90% narrowing of the tracheal lumen with pulsatile airway approximately 1 cm above the carina (length 1.5 cm).

Intervention and follow-up

The Patient under regular follow up, and a further evaluation of the airway by flexible bronchoscopy (at age of 18 months) Still showing significant narrowing of the tracheal lumen about 70%, with a marginal improvement compared to the earlier bronchoscopy finding further, there was no strong response to an increasing PEEP, making possibility of fixed narrowing of the airway lumen at malacia site. Currently, the patient is improving but, suffering from intermittent exacerbation wheezy chest and chest infection with frequent visit to the pulmonary clinic.

DISCUSSION

Tracheomalacia has been well-described in the literature for many years. There are two primary types of tracheomalacia: A **primary** lesion, in which the cartilage of the trachea develops abnormally with no apparent causes (idiopathic), and a **secondary** lesion, which is more common, often due to anatomical problems, such as a tracheoesophageal fistula, esophageal atresia or lesions that compress the airway, such as vascular slings, chest mass, or prolonged mechanical ventilation [2].

A tracheoesophageal fistula is usually repaired early in life. The associated tracheomalacia also appears in early infancy or shortly after surgery. In the majority of cases, the lesion presented as variable Lengths of malacia at the fistula site, usually at the middle or low third part of the trachea. The other common causes of malacia are cardiac-related.

Vascular rings, as in our patient, are often symptomatic at birth or seen early in the neonatal period. Vascular rings are a congenital anomaly of the aortic arch that results in a compression of the tracheobronchial tree and/or esophagus, leading to respiratory and gastrointestinal symptoms. Due to anomalies at the 4th week of embryogenesis, the aortic arch develops from six symmetrical paired aortic arch vessels and the paired dorsal aortae.

During the next several weeks of embryogenesis, remodeling and rearrangement of these structures result in the formation of the normal left aortic arch. Abnormal development of this complex vascular remodeling process results in malformations, which lead to the different types of vascular rings. The double aortic arch is a common cause of vascular rings, usually posterior is predominant size [3].An initial assessment depends on the frequency and severity of the symptoms. Our patient developed a symptom, stridor, from the first several days after being born. However, in winter time, he caught the flu, which required ventilatory support. His plain chest radiograph was normal apart of mild hyperinflation. An X-ray of the lateral neck did not show a narrowing of the intrathoracic trachea. Changes in the airway caliber can often be evaluated by fluoroscopy. A double aortic arch often causes

indentations on both sides of the esophagus on the AP projection of the **barium swallow,** as in our case**.** Computed tomography/magnetic resonance imaging are both valuable to detect extrinsic airway compression [4]. **Bronchoscopy** is the procedure of choice to confirm the diagnosis (airway malacia) and assess its severity. It can also differentiate between the tracheal stenosis and malacia in the operation room by applying positive pressure. It is usually performed under light general anesthesia with spontaneous respiration and can be performed with either a rigid or a fiber-optic bronchoscope [5] .A malacic airway assumes a comma-like configuration under direct visualization with bronchoscopy.

Management. In **mild** cases, a malacic airway is often a self-limited condition. Nonetheless, **moderate** cases, in which the patient is symptomatic under current respiratory infections, should be treated promptly with antibiotics, and early admission to the hospital may be necessary, particularly in young children. Inhaled bronchodilators may aggravate the condition, rather than improving the airway by reducing the tracheal muscle tone [6]. Prescribing anti-reflux medication to avoid feeding problems is particularly important in infants with tracheomalacia secondary to OA/TOF. Parents should be taught basic life-support skills on the **severe** types of malacia**,** as in our case. More complex tracheomalacia improves with growth and continuous positive airway pressure, and ventilation may even be required to maintain respiratory hemostasis.

Surgery may be an option when the patient has one or all the following: difficulty gaining weight and developing, recurrent pneumonia or apnea, and/or sufficient airway obstruction to require long-term airway support. There are different surgical options that were discussed regarding our patient:

-Tracheostomy is often used to provide internal stenting of the trachea for long-term airway support but may exacerbate tracheomalacia because it bypasses the physiological function of the glottis to maintain positive airway pressure. Furthermore, with time and growth, the airway obstruction resolves, and the patient can be decannulated. We were hesitated because of the malacia is in a critical position, immediately above the carina. Options for attempting a lower tracheal position vs. a longer ETT were weighted because of the concern on touching the carina and increasing vagal stimulation and possible traumatizing the airway into the adjunct blood vessels, which made us eliminate this choice.

-Tracheoplasty has been reported in similar cases, but because of a shortage of experts on this procedure, this option was canceled [7].

-Tracheal resection procedure is more beneficial for a localized, shorter segment. This method was eliminated due to a shortage of experts on this technique.

-Stenting of the airway has been an area of interest in recent years**.** Endotracheal stenting to stabilize the malacic airway was an option, but with the patient being an infant and the concern of tissue granulation and increased risk of metaplasia, we passed on this option [8].

- A second opinion was taken from the Great Ormond Hospital, where experts in the airway suggested a trial of **posterior aortopexy.** Hence, the anterior AA was ligated, and we decided to go with the previous approach. The results of the aortopexy are encouraging and show significant improvement or cessation of symptoms reported in over 95% of the cases of such TA, where an anterior aortopexy is easy and beneficial [9]. Our patient was extubated 3 weeks after the surgery, changed to CPAP for 2 weeks and later weaned off and was discharged in good condition with an audible wheeze but thriving well. The aortopexy and stent placement have been compared over a 10-year follow-up. Both are equally effective in improving the symptoms and allowing for normal growth and development. Aortopexy is associated with more perioperative complications, whereas stents are associated with long-term complications and the need for removal [10]. Most patients outgrow this malacia over time, but the severe type may present with asthma-like recurrent chest infections, or a barky cough might be misdiagnosed as croup. Aggressive chest physiotherapy, lower threshold for antibiotics, judicious use of Ventolin, as it may aggravate the bronchospasm, early use of CPAP during an exacerbation, and reflux therapy are often added to the treatment regime. Reassurance during acute respiratory illnesses may be necessary. Patients may need to be frequently seen during these illnesses. If the patient can achieve adequate oral intake and is acting normally, intervention is usually not necessary.

CONCLUSION

Severe tracheomalacia (post complete vascular ring surgery) is a life-threatening condition in the infancy period. There is no standardized therapy, but **posterior aortopexy** exhibits promise for severe residual types in DAA. Severe tracheomalacia required a persistent use of positive airway pressure. The presence of ‘cyanotic/apneic spells’ indicates the need for surgical intervention. Persistent wheeze is a common complication in which asthma therapy is needed to be used with great caution.

Abbreviations

CT: Cat scan, CPAP: Continuous positive airway pressure
AA: Aortic arch, DAA: Double aortic arch
ICU: Intensive care unit
PCO2: Partial pressure of carbon dioxide
SIMV: Synchronized Intermittent Mechanical Ventilation
OA: Esophageal atresia, TOF: Tracheoesophageal fistula

Consent:

Written informed consent was obtained from the patient's legal guardian for the publication of this case report. All accompanying images and the manuscript was approved by our local ethics committee (IRB) of the PSMMC.

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