Fellow's Column: Persistent Respiratory Distress in A Preterm Newborn

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"Congenital tracheal web is a rare upper airway malformation. It is characterized by a layer of tissue, which can vary in its thickness, draped across the tracheal lumen causing incomplete obstruction. (1) This condition usually goes undiagnosed or misdiagnosed from infancy to as late as adulthood. The estimated incidence is 1:10000 live births. (4)"

Introduction:

Congenital tracheal web is a rare upper airway malformation. It is characterized by a layer of tissue, which can vary in its thickness, draped across the tracheal lumen causing incomplete obstruction. (1) This condition usually goes undiagnosed or misdiagnosed from infancy to as late as adulthood. The estimated incidence is 1:10000 live births. (4)

We report a case of a preterm newborn with persistent respiratory distress and repeated episodes of apneas, bradycardias, and desaturations, diagnosed with congenital tracheal web.

Case Report:

A 27-week-old male newborn was delivered to a 26-year-old gravida 6 mother, with a significant past medical history of pregestational diabetes mellitus on metformin, and anxiety and depression on Lexapro. Her obstetric history is significant for fetal loss of a 20-week-old pregnancy and short cervix. During this pregnancy, she had ten antenatal visits. She took only prenatal vitamins, iron, Lexapro and metformin. She denied alcohol or illicit drug use. Prenatal labs were normal, and the quad screen was low risk.

Prenatal ultrasounds showed normal fetus and a shortened cervix. She was followed closely during the pregnancy with serial cervical length measurements. Cervical cerclage was placed at 19 weeks gestation age (GA), and vaginal progesterone was also started. Mother received two doses of Betamethasone at 25 weeks GA.

Mother went into labor at 27 weeks GA and delivered a male infant vaginally. There was thick meconium with a nuchal cord. The newborn received positive pressure ventilation for poor respiratory effort and bradycardia. He was then intubated, stabilized, and transferred to NICU. APGARs were 5 and 7 at 1 and 5 minutes. Birth weight was 1060g.

In the NICU, he was placed on SIMV, surfactant was administered, and blood was sent for laboratory investigations. He was started on fluids and antibiotics via central lines. Chest x-ray done showed signs of respiratory distress syndrome. He was also started on prophylactic phototherapy. Initial resuscitation and NICU management were uneventful.

He was extubated on day of life (DOL) 4. However, there was difficulty weaning him off non-invasive ventilation (NIV) support. Over the next three weeks, he was alternated between SiPAP and nCPAP/HFNC with little improvement in respiratory distress during weaning intervals. On DOL 27 he had to be re-intubated due to frequent episodes of apneas, bradycardias, and desaturations. He was extubated again on DOL 32 with similar difficulty weaning off NIV due to persistent respiratory distress during multiple attempted weans.

Evaluations done for possible etiologies were normal. Blood cultures done were negative, there were no electrolyte abnormalities, echocardiography showed a PDA with no significant shunt and no PPHN. Head ultrasound showed grade 1 Intraventricular hemorrhage, which resolved on follow-up imaging. Newborn screening was normal.

Due to an inability to wean off respiratory support by DOL 43, a

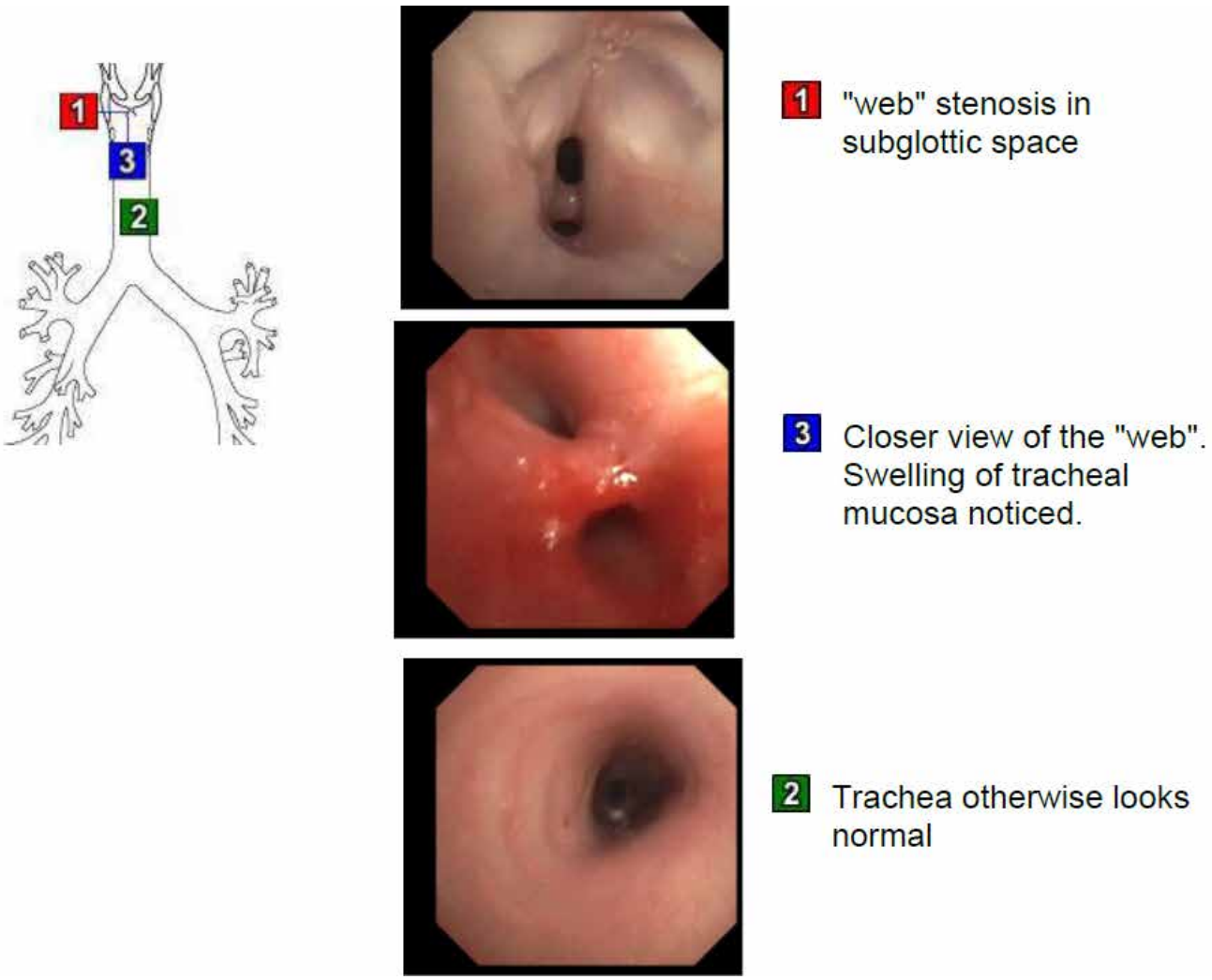

Figure 1 . Bronchoscopy

diagnosis of chronic lung disease was made, and he was commenced on diuretics with little improvement in his respiratory status. By DOL 73, he developed new onset wheezing. He was transferred to a tertiary center for further evaluation.

At the tertiary facility, chest computed tomography was negative for vascular anomalies of the great vessels (vascular ring or pulmonary sling), echocardiography showed moderate PDA with a left to right shunt and no PPHN. Bronchoscopy done revealed subglottic stenosis with tracheal webbing (Figure 1).

Following the diagnosis, he underwent rigid bronchoscopy with dilation of the tracheal web and stenosis. There were no acute complications following the procedure. He, however, developed stridor and respiratory distress days following the procedure, due to tracheal stenosis. He was intubated for 1 week and was able to be weaned to NIV and then room air after extubation without any issues. He developed some residual tracheal stenosis following full recovery from the subglottic stenosis and web dilation. He was discharged home 4 weeks post-op on therapy to manage his chronic lung disease and hypertension, which he developed during his NICU course.

Over the next year, he was hospitalized several times for respiratory failure secondary to acute bronchiolitis. He was intubated once and needed nCPAP, HFNC or BiPAP on multiple occasions during his hospitalizations.

Discussion:

Congenital tracheal web is an intrinsic luminal malformation usually with normal surrounding cartilage. It is characterized by a layer of tissue, which can vary in its thickness, draped across the tracheal lumen causing incomplete obstruction. (1)

This malformation may arise following unequal separation of the primordium when it differentiates into the esophagus and trachea during fetal development. (1,2) It is usually an isolated condition.

As a rare condition with a prevalence of 1 in 10000 live births and having varying degrees of symptomatology and severity in its presentation, it may go undiagnosed or misdiagnosed from infancy to as late as adulthood. (1,4)

In symptomatic newborns, the presentation may vary depending on the location of the lesion, degree of obstruction and/or other associated comorbidities in the newborn period. It may present as stridor, dyspnea, wheezing, persistent respiratory distress, recurrent respiratory infections, and respiratory failure. (1,2) While evaluating infants with this varying presentation, it is important to rule out other possible etiologies when the usual suspects are eliminated. This includes tracheal stenosis, laryngeal stenosis, pulmonary artery sling, or vascular ring.

Although a rare anomaly, multiple cases have been reported across all age groups either diagnosed incidentally or following the management of respiratory symptoms, which were refractory to all therapies administered.

Yin & Zhang in 20102, described a case of a 47-day-old female infant who after birth had a poor appetite, choking episodes with drinking milk and slow weight gain. Following workup for extended and severe cough, she was eventually diagnosed as a congenital tracheal web using coronal CT reconstruction and bronchoscopy.

Legasto et al. in 20041 described a 9-year-old female who had a history of wheezing, chronic cough, shortness of breath with exertion, recurrent pneumonia, and multiple hospitalizations since 18 months of age. She was treated aggressively with inhaled corticosteroids, bronchodilators, and leukotriene inhibitors with no improvement. Family history was significant for parental asthma and allergies. Pulmonary function test showed flow obstruction, which was suggestive of the tracheal lesion. Spiral CT scan with sagittal and coronal reconstruction showed a web-like structure obstructing approximately 60% of the airway lumen proximal to the carina.

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Another report by Al Badaai & Nguyen in 20083 described a case of a 16-year-old female with a past medical history of resolved laryngomalacia diagnosed at age 3.5 month and asthma diagnosed soon after managed with bronchodilators and steroids. She had multiple admissions for asthma exacerbations and pneumonia. A congenital tracheal web was identified using bronchoscopy following multiple unsuccessful attempts at endotracheal intubation in the operating room in preparation for hemithyroidectomy for a hyperfunctioning thyroid nodule.

Computed tomography is the appropriate first step in investigating patients with symptoms suggestive of tracheal web. CT reconstruction can help identify the presence, location, and severity of the obstruction. (1,2) It could also help rule out some other possible etiologies of tracheal stenosis. Bronchoscopy is the gold standard for confirmation of diagnosis, and may also be used therapeutically. Tracheal web may be easily torn by applying pressure using the bronchoscope. However, with increasing age and thickness of the web, balloon expansion, laser or surgical dissection will be indicated. (1)

Conclusion:

In conclusion, we have discussed the case of a preterm newborn with persistent respiratory distress and episodes of apneas, bradycardia, and desaturations, diagnosed with a very rare congenital tracheal anomaly. Congenital tracheal web, although rare, should be considered in newborns with persistent respiratory symptoms with difficulty weaning off respiratory support and development of new respiratory symptoms.

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