Late Presentation of Long Segment Tracheal Stenosis with Complete Tracheal Rings

A 3-year-old boy presented to our aerodigestive clinic with biphasic stridor, which had been present since infancy, and poor weight gain. Medical history was significant for birth at 37 weeks after a prenatal course complicated by mild intrauterine growth restriction, maternal preeclampsia, and a single pelvic kidney. He was referred to otolaryngology at 2 months of age for tachypnea and wet biphasic stridor and was diagnosed with laryngomalacia. His tachypnea resolved and his stridor improved over the first year of life. However, he developed recurrent prolonged, severe respiratory infections every 4-6 weeks between 2 and 3 years of age, necessitating flexible bronchoscopy computed tomography scan of the chest; he also had poor weight gain. During intubation the anesthesiologist had difficulty passing age-appropriate endotracheal tubes, but was able to place a 3.5 endotracheal tube. A computed tomography scan of the chest showed bilateral bronchiectasis and possible midtracheal complete cartilaginous rings (**Figure 1**). Flexible bronchoscopy was aborted owing to severe bronchospasm. Several weeks later, he underwent a direct laryngoscopy and flexible bronchoscopy and he was found to have congenital tracheal stenosis due to long-segment complete tracheal rings (**Video** [available at a www.jpeds.com] and **Figure 2**, A). At the level of the carina, tracheal origin of the right upper lobe bronchus (**Figure 2**, B), displacement of the right middle lobe bronchus, and severe distal tracheobronchomalacia were evident. The patient was referred to a quaternary pediatric airway center for consideration for slide tracheoplasty.

Stridor, a high-pitched respiratory sound typically produced by structural narrowing of the larynx and/or the extrathoracic trachea, can be inspiratory, expiratory, or biphasic. Inspiratory stridor suggests a supraglottic etiology, whereas biphasic stridor suggests obstruction at the level of the glottis, subglottis, or thoracic trachea. Biphasic stridor may be due to glottic webs, subglottic stenosis, tracheomalacia, or tracheal stenosis. Patients with persistent stridor may have poor weight gain, difficulty feeding, syndromic features, frequent and severe respiratory illnesses, and cyanotic or apneic episodes.[1]

Congenital tracheal stenosis is a rare cause of biphasic stridor, with the most common cause being complete tracheal rings.[2] Congenital tracheal stenosis can be classified as short segment or long segment and is often associated with congenital heart defects, the most common being a left pulmonary artery sling. In up to 50% of cases, tracheal origin of the right upper lobe bronchus can be seen. Symptoms typically begin a few days after birth, but may present later in childhood depending on the severity of the stenosis. Some children will present after intubation for an unrelated sedation when an age-appropriate endotracheal tube cannot be introduced into the trachea.[3]

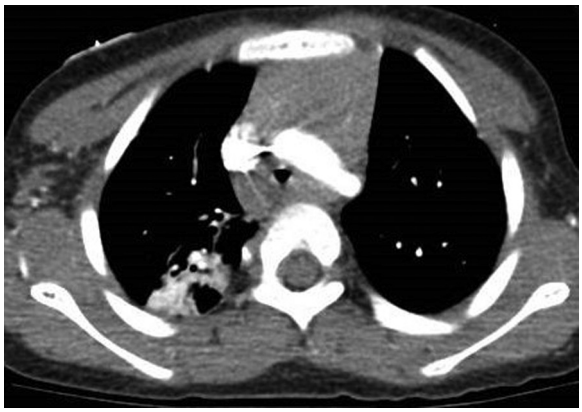

Figure 1. Axial computed tomography scan of the chest showing circumferential narrowing of the midtrachea consistent with complete tracheal rings.

The gold standard for the diagnosis of congenital tracheal stenosis is airway endoscopy. Computed tomography scans and magnetic resonance imaging are used to assess the relationship between the airway and cardiovascular anomalies.[3] In patients with less severe symptoms and no cardiac issues, observation as treatment may be appropriate.[4] The standard of care for surgical management of long-segment congenital tracheal stenosis is open reconstruction with slide tracheoplasty.[5] ■

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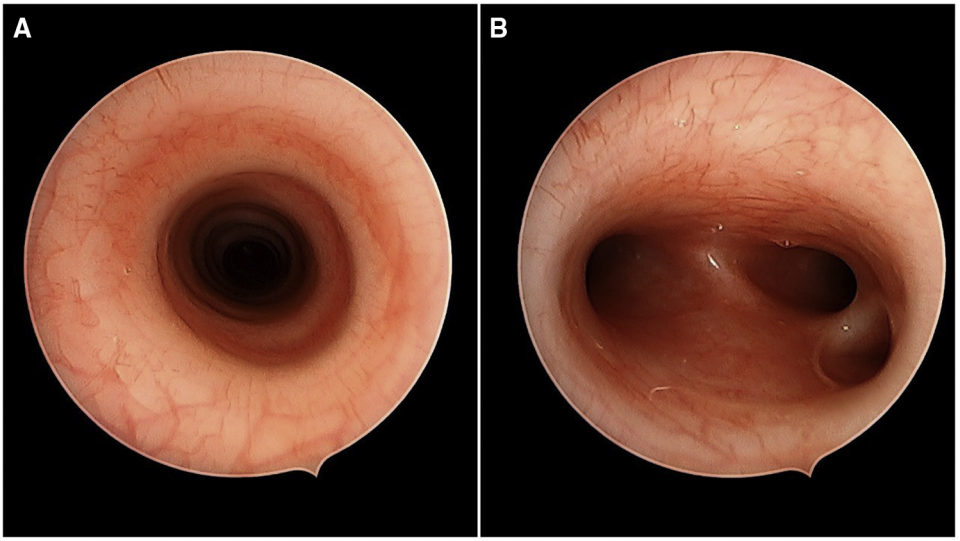

Figure 2. **A,** Midtracheal endoscopic view of complete tracheal stenosis. **B,** Tracheal origin of the right upper lobe bronchus.

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Salmon Patch Conjunctival Tumor

A 10-month-old boy presented with a salmon patch like lesion on the right superomedial conjunctiva that had been increasing in size for 3 months. Physical examination revealed an elevated pink-orange smooth surface, salmon patch-like conjunctival mass, measuring 14 mm × 8 mm, encroaching the limbus from 11 o'clock to 3 o'clock (**Figure**, A). The remainder of the ocular and systemic examination was normal.

The patient was treated with full surgical excision of the conjunctival mass. Histopathological examination (**Figure**, B-D) revealed tissue morphology and immunochemistry consistent with the diagnosis of juvenile xanthogranuloma. At a 1-year follow up, the lesion had regressed, without recurrence.

Juvenile xanthogranuloma is a non-Langerhans cell histiocytotic inflammatory disease accounting for 0.5% of pediatric tumors.[1] Ocular juvenile xanthogranuloma is a rare disease, occurring in 10% of patients with systemic juvenile xanthogranuloma.[2] The most commonly involved sites in ocular juvenile xanthogranuloma are the iris and corneoscleral limbus; conjunctival involvement is rare. Conjunctiva juvenile xanthogranuloma is frequently described as a yellowish mass on the conjunctiva.[1,3,4] In the present case, the salmon patch conjunctiva lesion hindered an accurate diagnosis, because it mimicked conjunctiva lymphoma. Consequently, histological and immunohistochemical examinations were used to differentiate juvenile xanthogranuloma from a malignant lesion (lymphoma) or other causes of histiocytosis (eg, Langerhan cell histiocytosis).

Currently, there is no consensus regarding the treatment of ocular juvenile xanthogranuloma. Successful treatment of conjunctiva juvenile xanthogranuloma has been reported with surgical excision, cryotherapy, and topical and systemic steroids.[3–5] Surgical resection can be useful in managing conjunctiva juvenile xanthogranuloma, with both diagnostic and therapeutic roles. ■

Data Statement

Data sharing statement available at www.jpeds.com.

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