Tracheobronchomalacia A Rare Cause of Respiratory Distress in Children

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SUMMARY: Tracheobronchomalacia is a rare cause of respiratory distress in children. An infant with tracheobronchomalacia is presented. The aetiology, pathophysiology and management are discussed.

Case Report

A six month old male infant was admitted to the Children's Ward at the Cambridge Military Hospital with symptoms of increasing wheeze. He had been symptomatic for nearly two months prior to admission with cough and wheeze for which he had been prescribed antibiotics by the General Practitioner. There was no

history suggestive of a foreign body or choking and he did not have any other systemic symptoms.

Birth history, past history and family history were non-contributory. On examination, at the time of admission, he was well grown, afebrile, acyanotic but with respiratory distress. There was no evidence suggestive of

Fig 1. X-ray of the chest (expiratory film) shows evidence of underinflation of the right lung with hyperinflation of the left lung with mediastinal shift.

Fig 2. Follow up X-ray done during re-admission (inspiratory film) shows hyperinflation of the left lung with mediastinal shift.

Fig 3. Krypton/Technetium 99 ventilation/perfusion scan.

a. Impaired ventilation of the left lung (41%). Right lung (59%) shows patchy abnormalities of the upper lobe and apical segment of the lower lobe.

b. Decreased perfusion of the left lung (41%) and right lung (59%).

(A. Krypton: 1 anterior 2 & 3 posterior. B. Technetium: 1 anterior 2 & 3 posterior).

chronic respiratory disease such as finger clubbing, Harrison's sulcus or pigeon chest. Systemic examination was unremarkable, except for diffuse rhonchi and crepitations.

Full blood count and biochemical screen were normal. Initial chest X-ray showed an under-inflated right lung with hyperinflation of the left lung (Fig 1), which was thought to be due to mucous plugging. He was started on oral erythromycin, nebulised salbutamol and regular chest physiotherapy. He improved over the next 72 hours and follow-up X-ray of the chest was normal. He was discharged home on oral salbutamol and erythromycin with advice to continue regular chest physiotherapy. He was re-admitted 17 days later, with symptoms of increasing respiratory distress and wheeze. Repeat chest X-

ray revealed hyperinflation of the left lung with mediastinal shift to the right (Fig 2). A diagnosis of foreign body in the left main bronchus was considered, and he was transferred to the Hospital for Sick Children, Great Ormond Street, London, for bronchoscopy and further respiratory investigations. On examination of the respiratory system at the Hospital for Sick Children, he had tachypnoea (respiratory rate 50/minute) intercostal recession, bilateral wheeze and decreased air entry in the base of the left lung. Bronchoscopy showed minimal subglottic narrowing, a tracheomalacic segment between middle and distal thirds with about 20% narrowing, and a 2cm long bronchomalacic segment of the left main bronchus leading on to 80–90% obstruction. Barium swallow and 2 D echocardiogram were normal,

ruling out vascular ring, double aortic arch or pulmonary artery sling. Magnetic Resonance Imaging (MRI) of the chest did not reveal any abnormality to cause the tracheobronchomalacia. A ventilation perfusion scan showed decreased ventilation and perfusion on both sides, the left side showing 33% of overall ventilation and 41% of overall perfusion (Fig 3).

The child is now nearly three years old and has remained symptomatically well, without further respiratory distress, requiring hospital admission, the use of regular bronchodilators or antibiotics. Chest X-ray has remained normal. He is growing and developing normally. It is probable that with advancing age, the tracheobronchomalacic segments have become more competent, preventing further respiratory complications.

Discussion

Respiratory distress is a common reason for hospital admission in childhood. Chest infection, either bacterial or viral, and bronchial asthma are the two most common causes. Foreign body inhalation and congenital abnormalities of the respiratory tract, such as tracheobronchomalacia, are rarer causes and require flexible bronchoscopy to confirm the diagnosis and for removal of the foreign body.

Tracheobronchomalacia can be either congenital or acquired (1). Congenital tracheobronchomalacia may be an isolated abnormality, or be associated with other congenital abnormalities such as laryngomalacia, choanal atresia or cleft palate (2). Recently Larsen's syndrome has been associated with tracheobronchomalacia (3). Congenital heart disease with enlarged pulmonary arteries can present as bronchomalacia due to external compression (4).

Acquired tracheobronchomalacia, an adult disease, usually follows chronic bronchitis, bronchial carcinoma or pulmonary tuberculosis and often progresses with increasing respiratory morbidity and mortality (1). Chronic inflammatory diseases affecting the respiratory tract break the protein-polysaccharide bond, which leads to the loss of resilience of hyaline cartilage, causing tracheobronchomalacia (5). The area of tracheobronchomalacia can be either isolated or multiple (6) and rarely bronchomalacia can be familial (7). More recently, with increasing survival of extremely low birth weight babies and the complication of bronchopulmonary dysplasia, bronchomalacic segments have been observed on flexible bronchoscopic examination for worsening respiratory functions (8). Whether these areas of bronchomalacia are congenital or acquired due to bronchopulmonary dysplasia is debatable. Children with non-productive intractable cough, usually considered to be psychogenic, should have flexible bronchoscopic examination under local anaesthesia and sedation to rule out an underlying abnormality of the tracheobronchial walls, as an increased incidence of

tracheobronchomalacia in these patients has recently been reported (9). Tracheobronchomalacia produces a flaccid airway, which has an adequate lumen on inspiration but develops obstruction on expiration. This can lead to collapse of the airway, stridor and respiratory failure, infrequently needing ventilatory support (10,11,12,13).

As compared to adults the tracheobronchial tree in infants is extremely soft and the mucosa is redundant especially around the carina (14). During forced expiration, crying and coughing, the lumen may become completely obliterated due to increased intrathoracic pressure exceeding the transbronchial pressure, leading to herniation of the posterior membrane and antero-posterior narrowing (15).

Usually children with primary tracheobronchomalacia recover spontaneously and become asymptomatic by the second year of life (2,7). Children with congenital heart disease, enlarged pulmonary arteries and secondary bronchomalacia will improve with surgical correction of the heart defect and arterioplasty (4,16). Recently in children with bronchomalacia and respiratory failure, endoscopic placement of expandable stents has been found to be useful, thus avoiding complex high risk surgical procedures such as arterioplasty, tracheopexy, external stenting, tracheobronchial reconstruction and pneumonectomy (17,18).

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