A Dual Diagnosis: Follicular Bronchiolitis & Williams Syndrome

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OBJECTIVES

- Present a case of a pediatric patient with persistent respiratory distress.
- Describe the disease process, diagnosis, and treatment of follicular bronchiolitis.
- Emphasize the importance of a complete workup to identify all possible diagnoses.

INTRODUCTION

Respiratory distress in infants and children is common, and the differential diagnosis is broad. Two of the most common causes include infectious (most often viral) and inflammatory (such as reactive airway disease or asthma). Interstitial lung disease should be highly considered when peribronchial ground glass opacities are seen on chest CT.

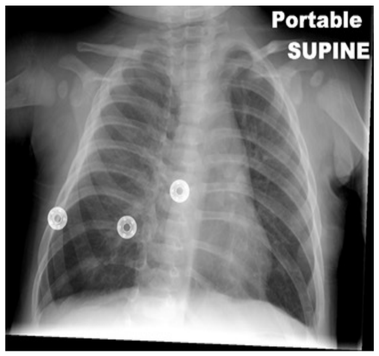

Figure 1. Chest radiograph showing hyperinflated lungs, mild interstitial and bronchial wall thickening throughout the lungs, central peribronchial opacities bilaterally, and atelectasis.

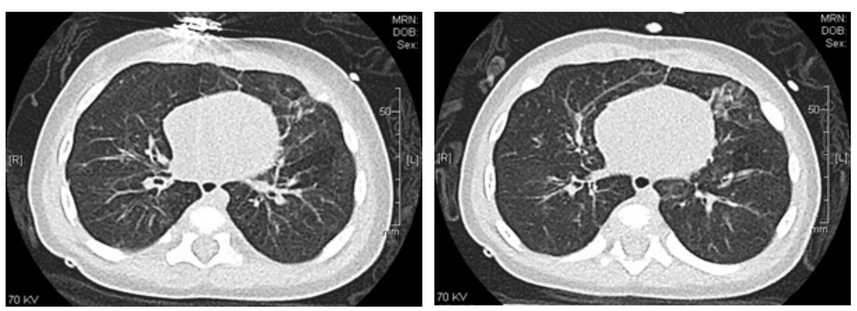

Figure 2. Chest CT showing bronchial wall thickening most severe in the upper lobes, subtle heterogenous attenuation of the lungs likely secondary to air trapping, and areas of bandlike, patchy, and nodular ground glass opacities.

CASE

GA is a full term 7-month-old female who presented to the ED with cough, tachypnea, and worsening respiratory distress for 2 months.

Physical exam: Significant for tachypnea with mild-moderate subcostal retractions and crackles on auscultation of the bilateral lower lobes of the lungs, 2/6 holosystolic murmur, and generalized low tone with delayed motor milestones.

Imaging: A chest x-ray was nonspecific (figure 1). A CT scan showed bronchial wall thickening, air trapping, and bandlike, patchy, and nodular ground glass opacities (figure 2).

Lung biopsy: Demonstrated the presence of lymphoid follicles in the walls of bronchioles and narrowing of the bronchiolar lumen, consistent with follicular bronchiolitis (figure 3).

Further workup: An echocardiogram was obtained due to the development of hypertension in addition to the murmur present on admission, which showed aortic stenosis and left ventricular hypertrophy (LVH). In addition to genetic testing sent for causes of interstitial lung disease, the patient underwent genetic testing for hypotonia, which revealed a diagnosis of Williams Syndrome.

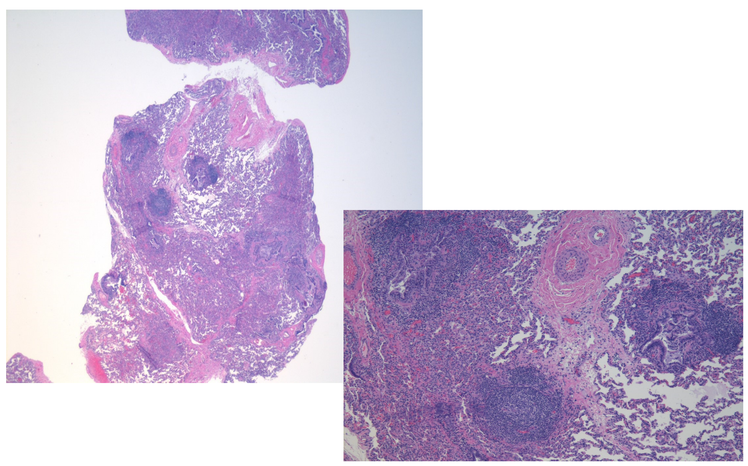

Figure 3. Lung biopsy demonstrating well-formed lymphoid follicles in the walls of bronchioles and narrowing or obliteration of the bronchiolar lumen.

DISCUSSION

Follicular bronchiolitis is a rare interstitial lung disease. It is a non-neoplastic lymphoproliferative pulmonary disease thought to be caused by antigenic stimulation of bronchial associated lymphoid tissue (BALT) and subsequent hyperplasia of lymphoid follicles within the walls of bronchioles. Patients with follicular bronchiolitis typically present with chronic, nonspecific respiratory signs and symptoms as early as the neonatal period. Common signs and symptoms include cough, tachypnea, increased work of breathing or respiratory distress, adventitious lung sounds on auscultation, and hypoxemia. Diagnosis is made by chest CT and lung biopsy. Treatment is largely supportive and can include the use of supplemental oxygen, systemic steroids, and macrolide antibiotics.

The diagnosis of follicular bronchiolitis was necessary in order to initiate proper treatment and achieve improvement and ultimate resolution of this patient's respiratory symptoms. While addressing her primary complaint, further evaluation of the history and physical exam findings ultimately led to an additional diagnosis. It became evident that her aortic stenosis may have been multifactorial, and possibly associated with her new diagnosis of Williams Syndrome. This is the first reported case of a patient with both follicular bronchiolitis and Williams Syndrome.

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