



Something is missing in the bronchus— Williams-Campbell syndrome

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A 60-year-old female who was a former smoker was admitted with a five-day history of progressive dyspnea, cough, sputum production, and fever. She had a history of recurrent pneumonia since her childhood. Physical examination revealed expiratory wheezes and an SpO₂ of 91% on room air. A chest CT scan was performed (Figure 1). On the basis of the radiological findings and the exclusion of other causes, the patient was diagnosed with Williams-Campbell syndrome (WCS). After 10 days of antibiotic therapy, she was discharged with improved symptoms.

WCS is a rare congenital disorder characterized by the absence of cartilage in subsegmental bronchi, leading to bronchiectasis.^(1,2) The pathophysiology of WCS involves airway collapse caused by deficiency of cartilage, resulting in chronic respiratory symptoms such as dyspnea and recurrent pulmonary infections.⁽¹⁾ Diagnosis is typically based on clinical manifestations and characteristic

radiological findings on chest HRCT scans, as well as on exclusion of other causes of bronchiectasis.⁽³⁾ Management of WCS remains challenging because of its rarity, being based on the use of antimicrobials. Treatments such as noninvasive positive pressure ventilation have shown promise in managing respiratory failure, and lung transplantation may be considered in severe cases.^(1,2)

AUTHOR CONTRIBUTIONS

FMC and ACPG: study conception, planning, and design; data collection; and drafting of the manuscript. FMC, ACPG, and AKM: revision of the manuscript. All authors read and approved the final version of the manuscript.

CONFLICTS OF INTEREST

None declared.

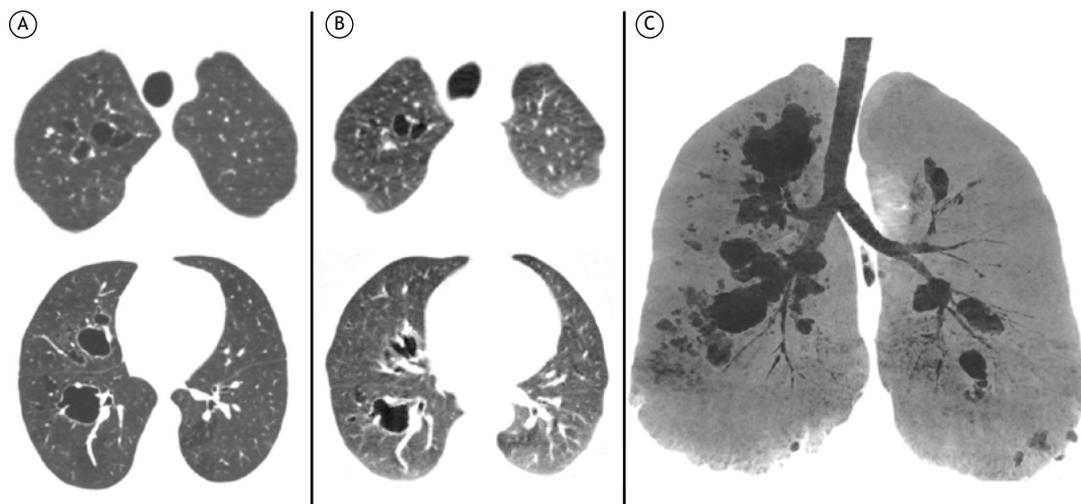


Figure 1. Axial CT scans of the chest taken during inhalation (in A) and exhalation (in B). Note complete or partial airway collapse during exhalation. In C, coronal CT scan of the chest with minimum intensity projection to improve visualization of the airways. Note bronchiectasis in both lungs, involving fourth- to sixth-order bronchi. The peripheral and central airways, including the trachea, main bronchi, and lobar bronchi, remain unaffected by bronchiectasis.

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