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Case Report

William Campbell Syndrome as a Cause of Asymmetrical Bronchiectasis In a 10-Year-Old Child: A Case Report

Maliha Baseer[1], Saima Shan[2], Asim Rehman[3], Jovaria Ehsan[4], Aisha Hameed[5], Farkhanda Jabeen[6]

[1,2,4,5,6]Department of Radiology, Federal Government Polyclinic Hospital, (PGMI), Islamabad; [3]Department of Radiology, CITI lab and Diagnostic Center, Rawalpindi

ABSTRACT

Background: William Campbell Syndrome is a rare congenital disease of the bronchial cartilages involving the 4th to 6th generation of bronchial divisions with involvement of bilateral lungs. There is deficiency of bronchial cartilages resulting in dilated bronchi, with collapse of the distal lung. Diagnosis is by clinical and radiological findings.

Case Presentation: A 10-year-old female child presented in OPD with fever, wheezing and productive cough. The patient has been taking treatment for past 1 year for asthma. Chest x-ray showed reduced left lung volume, with multiple infiltrates in the left mid and lower zones. Patient was referred for a CT scan which demonstrated segmental and subsegmental bronchiectasis in the bilateral lungs, asymmetrically. Expiratory / Inspiratory CT demonstrated a significant diminution in size of the involved bronchi. William Campbell Syndrome was proposed as the underlying etiology, after exclusion of common causes of bronchiectasis.

Conclusion: Bronchiectasis in children is often underdiagnosed and undertreated. When young patients present with recurrent infections, the underlying cause is important to elucidate for the management of the patient and for improving life quality and expectancy. William Campbell syndrome is a rare cause of congenital bronchiectasis due to deficient bronchial cartilage. Diagnosis in our patient was made by history, imaging and exclusion of other causes of bronchiectasis.

Keywords: Bronchiectasis, High Resolution Computed Tomography Chest (HRCT Chest), William Campbell Syndrome (WCS),

Authors' Contribution:
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Correspondence:
Maliha Baseer
Email: malihabaseer@hotmail.com

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Introduction

William Campbell syndrome (WCS), is a rare congenital disease of the bronchial cartilages principally affecting the bronchi in the 4th to 6th generation of bronchial divisions. It is characterized by defective or absent cartilage in the involved segments, leading to abnormally dilated bronchi and collapse of the airway distal to the involved segment[1]. WCS is most often associated with diffuse and bilateral bronchiectasis, usually manifesting in early childhood with recurrent lower respiratory tract infections, chronic cough, and wheezing[2].

Diagnosis of this rare disease is based on clinical and radiological findings and after exclusion of other causes of bronchiectasis[2]. We present a case of asymmetrical bronchiectasis in a 10-year-old child with a history of asthma and repeated infections. The final diagnosis was proposed to be William-

Campbell Syndrome, highlighting the importance of considering congenital airway malacia in the differential diagnosis of bronchiectasis in paediatric patients.

Case Presentation

A 10-year-old female child presented in OPD with fever, wheezing and productive cough. Patient was on inhaled steroids and leukotriene modifier (montelukast) for the past 1 year for the treatment of clinically diagnosed asthma. On examination, there was an asymmetrical appearance of chest, with reduced expansion of left hemithorax. There were bilateral crackles on auscultation, in the mid and lower chest, more on the left side. No signs of cyanosis or clubbing was appreciated. Chest x-ray was performed which showed reduced left lung volume and multiple infiltrates in left mid and lower zones; situs solitus was observed with heart and liver in their normal orthotopic positions (Figure 1). X-ray Paranasal Sinuses was also performed which revealed normal pneumatization of paranasal sinuses. Patient was referred for Computed Tomography (CT) Chest. Considering atypical symptomatology Contrast Enhanced Computed Tomography (CT) Scan of Chest was performed to evaluate for mediastinal structures and pulmonary vasculature. X-ray findings were confirmed with situs solitus and partial loss of left lung volume; there was an ipsilateral shift of mediastinum and trachea (Figure 2 and 3). Cystic and tubular dilatation of segmental and subsegmental bronchi was observed in bilateral lungs, with significant involvement of the left upper and lower lobes and compensatory hyperinflation of the right lung. Evidence of active infection was demonstrated with few tree in bud nodules in the right middle lobe and with enlarged mediastinal lymph nodes, significant of which were in subcarinal location (6 x 11mm) (Figure 4). Few subpleural bullae were also observed in apical segment of right upper lobe (Figure 3); there was no evidence of bronchial atresia. Pulmonary trunk, right and left pulmonary arteries were normal in caliber, without any evidence of pulmonary hypoplasia (Figure 4). Based on the pattern of involvement of lung and bronchi, a provisional diagnosis of congenital airway malacia was made, and the patient was called for High Resolution Computed Tomography (HRCT) Chest in both expiratory and inspiratory phases. Careful consideration was awarded to the young age of the patient and radiation dose for additional CT; low dose paediatric CT was planned for both inspiratory and expiratory phases. This revealed a detectable collapse of airways involving subsegmental and distal bronchi in expiratory CT as compared to inspiratory CT; mild reduction in luminal diameter of dilated segmental bronchi was noted (Figure 2 and 3). This collapse / diminution of bronchial lumen was observed in both right and left lungs.

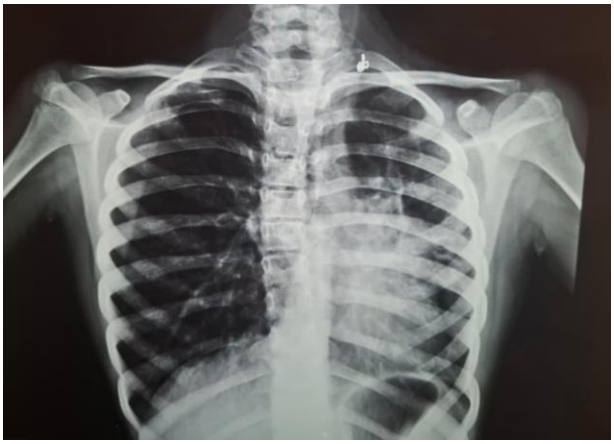

Figure 1: Chest X-ray Frontal projection showed partial loss of left lung volume, with infiltrates in left mid and lower zones

Based on the clinical and radiological findings, provisional diagnosis of William Campbel Syndrome, a disease with congenital airway malacia was made. Differential diagnosis included cystic fibrosis, Kartagener's syndrome and Mounier-Kuhn syndrome which were ruled out

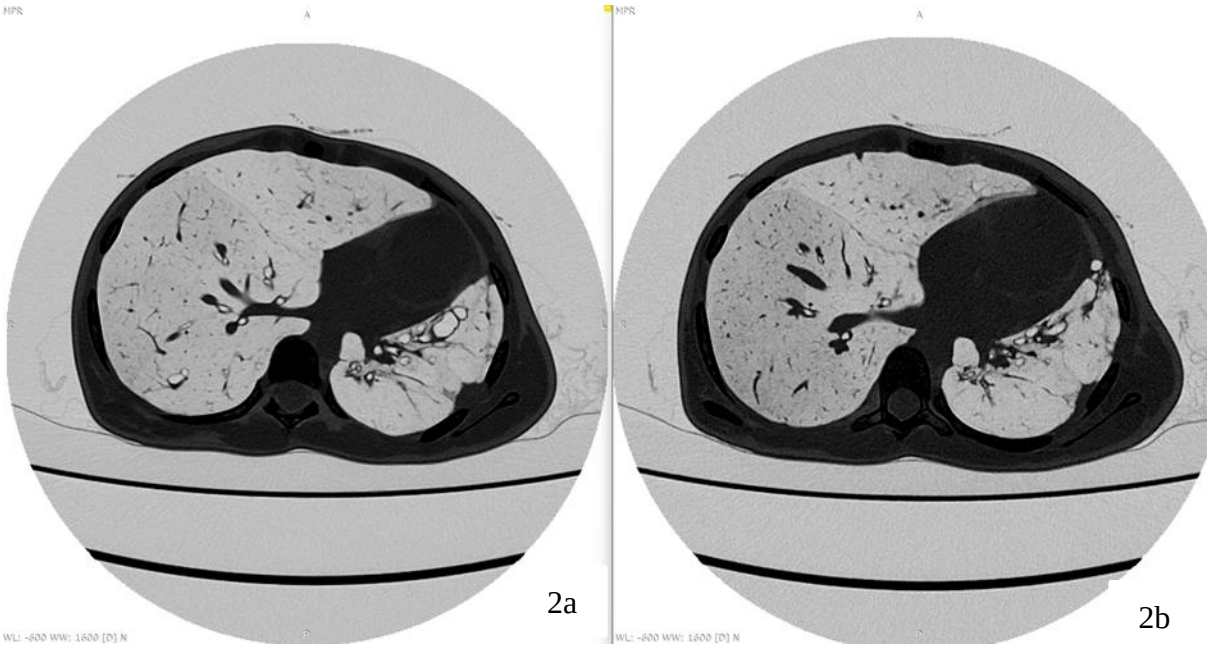

Figure 2. Figure 2a: Axial images of expiratory chest CT, lung window showing bronchiectasis involving bilateral lungs, more in left lower lobe with principal involvement of segmental and subsegmental bronchi. Volume loss of left lung and ipsilateral shift of mediastinum

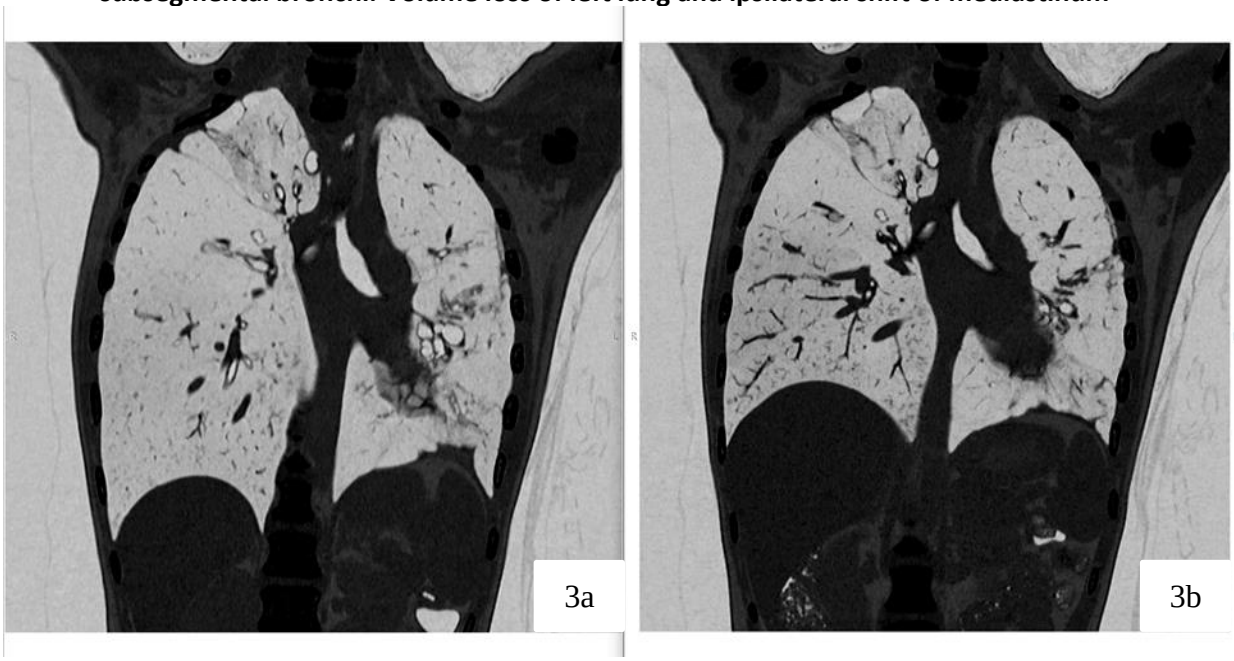

Figure 3. Figure 3a: Coronal images of expiratory chest CT, lung window showing bronchiectasis in bilateral lungs. Few subpleural bullae are evidence in apical segment of right upper lobe.

based on the absence of situs inversus, sinusitis, sweat chloride test and lack of tracheomalacia on imaging. The patient and her caregivers were counselled about the condition and importance of adhering to treatment regimens and were counselled about exacerbating symptoms.

Genetic counselling was recommended, due to the familial nature of the condition, but it was not possible due to the unavailability of a genetic testing facility in Pakistan. Patient was stable at discharge and advised regular follow-up.

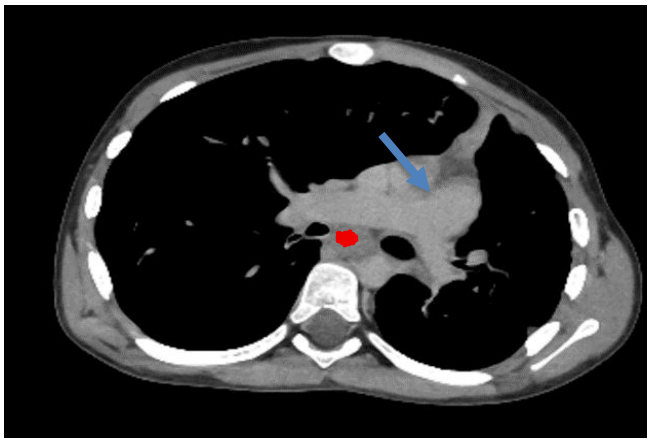

Figure 4: Axial Sections of Contrast Enhanced CT Chest, mediastinum window, showing normal pulmonary trunk and right and left pulmonary arteries, blue arrow. Enlarged subcarinal lymph node is also seen, labelled as red asterisk.

Discussion

Bronchiectasis is a complex pulmonary condition characterized by repeated infections and bronchial inflammation. Histopathologically, it is characterized by destruction of elastic and muscular components of the bronchial wall, resulting in abnormal and often irreversible dilatation of bronchi[3]. This results in a progressive decline in lung function. Despite widespread use of High-Resolution Computed Tomography Chest (HRCT) scans in clinical practice, bronchiectasis in children remains an under-recognized and undertreated pulmonary disorder[4]. Clinically, the young patient presents with persistent productive cough, most of the time, which may be non-purulent. Other clinical features may include, wheezing, hemoptysis and growth retardation.[5] Causes of bronchiectasis in children are wide and varied and include recurrent infections, immune deficiencies, mucociliary diseases and congenital structural defects[6]. Congenital structural defects include Mounier-Kuhn Syndrome, Tracheoesophageal fistula, bronchomalacia and William Campbell Syndrome[7]. William-Campbell Syndrome (WCS) is a rare condition first proposed in 1959 by William and Campbell as maldevelopment of bronchial tree cartilage[8]. Familial/genetic etiology was proposed in 1976 when two siblings presented with respiratory discomfort immediately after birth. There may be associated congenital abnormalities of cardiovascular system[8]. Rare sporadic cases haves also been reported, one of which was proposed to be secondary to adenovirus infection.[7] Pathophysiologically, there is a deficiency of cartilage in the walls of bronchi, at 4th and 6th order divisions. This deficiency occurs early in life although rare instances of adult presentation have also been reported[1,2]. There is no cartilage deficiency elsewhere in the body in reported cases.

Diagnosis of this rare syndrome is made by clinical presentation, imaging and by excluding other congenial and acquired causes of bronchiectasis in children. Typically, the patients may present with shortness of breath, productive cough and repeated infections[1,9]. Computed tomography plays an important role in diagnosis of WCS, with inspiratory and expiratory scans having a pivotal role.

In our child patient, bronchiectasis was documented on initial CT, and definitive diagnosis was made on HRCT Chest in inspiratory/ expiratory phases. There was a significant reduction in the size of involved bronchi in expiratory CT. Evidence of hyperinflation was also seen with subpleural blebs. Bilateral involvement and relative sparing of trachea, main and lobar bronchi excluded other differentials like allergic aspergillosis, cystic fibrosis and Mounier Kuhn syndrome. Definitive diagnosis of this rare syndrome is by taking a biopsy specimen of bronchi and submitting it for histopathology, demonstrating deficiency of bronchial cartilage. However, due to significant morbidity involved with this procedure, it is not recommended in routine clinical practice and living subjects[2]. Although the pathogenesis of both WCS and Mounier Kuhn syndrome involves a deficiency of cartilage, radiologically, these can be differentiated by following two important points. First, William Campbel Syndrome involves the 4th to 6th generation of bronchial cartilages, the level where the dilated airways in Mounier Kohn syndrome should return to normal. Secondly the

dilated bronchi in William Campbell Syndrome can collapse partially or completely on expiratory CT[10]. The distribution of bronchiectasis also helped in differentiating from other disorders i.e. cystic fibrosis and primary ciliary dyskinesia. Predominant upper lobe involvement suggests cystic fibrosis, while primary ciliary dyskinesia typically involves the middle and lower lobes. Involvement of the right middle lobe and lingular segments of the left upper lobe typically signify non-tuberculous mycobacterial infection[1].

Our case represents importance of keeping a wide differential in radiological diagnosis while also reflecting significance of tailoring radiological procedures according to the diagnostic question.

Management of WCS is the prevention of acute exacerbations, which can be done by a prophylactic antibiotic course for 7-10 days. Treatment of acute attack involves bronchodilators, oxygen therapy, and chest physiotherapy. Non-invasive positive pressure ventilation is indicated in respiratory failure. Severe refractory cases may be offered a surgical procedure, i.e. lobectomy or lung transplant, but the research is limited in these areas, with unfavourable outcomes[9].

Conclusion

Bronchiectasis in children remains an under-recognized and undertreated condition. Determining its etiology is essential to direct therapy and enhance outcomes. Our case established William-Campbell Syndrome (WCS) as the cause of bronchiectasis, which is a congenital deficiency of bronchial cartilage. HRCT Chest proved decisive, paired inspiratory/expiratory scans demonstrated expiratory collapse of dilated bronchi after exclusion of alternative etiologies. We strongly recommend considering WCS in children with recurrent lower respiratory symptoms and bronchiectasis. Adopting tailored HRCT protocols that include expiratory imaging helps prevent missed diagnoses and enables timely, targeted management.

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