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William Campbell syndrome in an adult male – a rare entity

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Abstract:
Introduction
William Campbell syndrome is a rare congenitaldisorder which is characterised by the deficiency of cartilage in the sub-segmental bronchi leading to collapse of distal airways and bronchiectasis.It is also known as bronchomalacia. There have been few reports in recent years of adult patients being affected by diminished cartilage, saccular bronchiectasis and para-cicatricial emphysema, all of which are characteristic features of Williams–Campbell syndrome.
Case presentation
Here we present a case of a 45-year old male having chronic cough, dyspnoea, and sputum production since 30 years. The clinical and laboratory examination suggested that he had recurrent respiratory infections due to bronchiectasis caused by Williams–Campbell syndrome, which was undiagnosed in the patient till his presentation.
Conclusion
Although it is a rare syndrome, but Williams–Campbell syndrome should be considered as a differential diagnosis in patients presenting with signs and symptoms of recurrent respiratory infections, bronchiectasis, productive cough and dyspnoea.

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I. Introduction

Williams–Campbell syndrome is a rare congenital disorder of the airways characterized by the deficiency of cartilage in the sub-segmental bronchi. The cartilage rings beyond the second and sometimes even first division of bronchi are absent which results in bronchiectasis in the fourth- to sixth-order bronchi. The central bronchi and trachea are unaffected. Williams–Campbell syndrome thus is characterized bycentral saccular or cystic bronchiectasis and collapse of distal airways.

The prognosis and severity of symptoms depend on the extent of cartilage mal-development of the bronchi.[1] Though the syndrome is best described in pediatric population with history of recurrent pneumonia orbroncho-obstructive symptoms such as cough, wheeze, there have been a few cases diagnosed in adults as well.[1,2]

II. Case Report

A 45 year old farmer presented to our out-patient clinic with complains of productive cough, intermittent fever without chills and breathlessness on exertion for past 7 days. He had a history of repeated visits to the hospital since 15 years of age with similar complaints. On these hospital visits, he was treated with antibiotics, antipyretics and inhaled bronchodilators. There was no previous history suggestive of Tuberculosis. Rest of the systemic review and past medical history was unremarkable. Patient smoked occasionally and was a social drinker. The patient had a younger brother, a chronic smoker, with similar features who expired 5 years back at the age of 34 years. On examination at the time of presentation, the patient was afebrile, tachypnoiec, normotensive. Clubbing was present but rest of the physical examination was normal. Respiratory system examination revealed wide spread bilateral crepitationsover the lower part of both lungs. The Arterial blood analysis and electrocardiogram were normal.

Lab data demonstrated slightly elevated leucocyte count(12,400/mm3) without peripheral eosinophilia, Hb 14.3 gm/dl, ESR 10 mm/hr. Biochemistry profile wasnormal. Serum was negative for HIV antibodies. Induced sputum for acid-fast bacilli, Aspergillus and other fungal organisms was negative on multiple occasions.Sputum for culture grew Pseudomonas.

Chest radiograph revealed multiple ring like shadows in both the lungs, almost symmetrical in distribution, more apparent in the mid and lower zones. (Figure 1)

Serum α-Antitrypsin levels and total IgE (33.2/ml) was normal.Immediate cutaneous reaction and Serum precipitins against Aspergillus fumigates were negative. A. fumigatus-specific serum IgE and IgG levels were within normal range (to rule outallergic broncho-pulmonary aspergillosis). Total serum IgG, IgA and IgM levels were normal (to rule out hypogammaglobulinemia). Hematological profile like anti-nuclearantibody and anti-neutrophiliccytoplasmic antibody tests (vasculitic profile to rule out Churg-Strauss Syndrome) were negative.

Spirometry revealed a mixed obstructive-restrictive pattern with reduced DLCO. The patientdid not consent for bronchoscopy.

HRCT Thorax revealed central cystic and varicose bronchiectasis bilaterally predominantly in mid/lower zones.Collapse of bronchi with distal air trapping due to presence of excessively compliant bronchi was also present. (Figure 2) Based on history, clinico-radiological findings and laboratory testing a diagnosis of William Campbell Syndrome was made.

III. Discussion

Williams and Campbell were the first to report an unusual pattern of bronchiectasis in 1960. They reported a series of 5 cases with disease presentation in infancy with similar clinical features. They noted that the central bronchi were markedly soft and compliant, ballooning on inspiration and collapsing on expiration. In this syndrome, bronchiectasis is secondary to the deficiency in cartilage of the third- to sixth-order of bronchi generation.[2]

This syndrome usually presents inpediatric population with a history of recurrent pneumonia and broncho-obstructive symptoms like cough and wheeze. However, several adult cases have been reported in the recent years without any pathologic confirmation.[2,4–6]

CT imaging demonstrates bilateral central cystic/cylindrical bronchiectasis distal to the third-generation of bronchi i.e. the segmental and sub-segmental level of bronchi along with hyperinflation of the affected lungs.[4,7]On expiratory film, collapse of the bronchi with distal air trapping may be observed as a result of excessively compliant bronchial walls. The trachea and central bronchi are normal in calibre which is a distinguishing feature of this syndrome. The mechanism behind the deficiency in bronchial cartilages is not understood till date; however the cartilage deficiency in this syndrome is found exclusively in the bronchi.[8,9]Diagnosticbronchoscopy is often unrevealing.[10]

A genetic background with familial occurrence has also been reported.[10–12] Themarkedly compliant bronchi collapse during coughing thus leading to poor drainage of the airways which subsequently leads toa progressive obstructive disease causing hyperinflation of the affected lungs and segmental or lobar collapse. Recurrent destruction of the bronchial tree and inadequate clearance of mucus leads to further damage to the lung parenchyma.[8,9]

The long-term prognosis is variable, with rapid clinical deterioration and death in some children and prolonged survival in others.[13] The prognosis and severity of the symptoms depend on the extent of the cartilage mal-development.

There is no specific treatment for this syndrome. Prophylaxis from exacerbations forms the mainstay of the treatment.[14,15]Prophylaxis can be achieved by administration of an oral or intravenous antibiotic for 7–10 days or until the sputum production is reduced. For severe cases, several different antibiotics may be used sequentially in a continuous regimen to minimize the risk of bacterial resistance. Transplantation has been reported in a patient with severe respiratory symptoms, but the patient died in a year and post-mortem examination revealed that the main bronchi had bronchomalacia, which had attributed to a respiratory infection during the post-surgical period.[1]

Depending on the patient's symptoms like bronchospasm, thick, tenacious sputum, etc. a bronchodilator, combined with postural drainage and chest percussion/physiotherapy may be done when required, to help remove secretions. Respiratory exercisesand bronchoscopy may be helpful in some cases to help mobilize secretions. In cases presenting with hypoxia, oxygen therapy should be considered.[14] Non-invasive ventilation can be used in cases presenting with respiratory acidosis.[16] Other acquired and congenital conditions associated with bronchiectasis must always be excluded before establishing a diagnosis of William-Campbell Syndrome. We were able to exclude cystic fibrosis, immune-deficiencies(Ig, α-1 antitrypsin), and allergic broncho-pulmonary aspergillosis.

IV. Conclusion

Although it is a rare syndrome, Williams–Campbell syndrome should always be included in the differential diagnosis of bronchiectasis with recurrent chest infections, repeated episodes of productive cough and breathlessness and bronchiectasis. Characteristic HRCT features along with clinical presentation gives a clue in diagnosing but a definite diagnosis can be made only after detailed workup and excluding the other common causes of cystic bronchiectasis viz. Cystic Fibrosis, Immunodeficiencies (Ig, α-1 antitrypsin),allergic

broncho-pulmonary aspergillosis(ABPA), Mounier-Kuhn syndrome, Radiation Fibrosis, Tuberculosis, immotile cilia syndrome etc.

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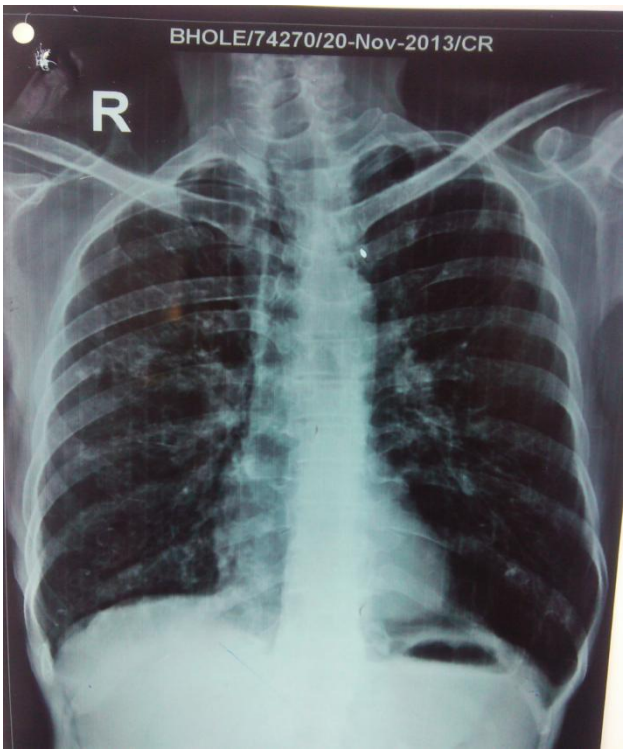
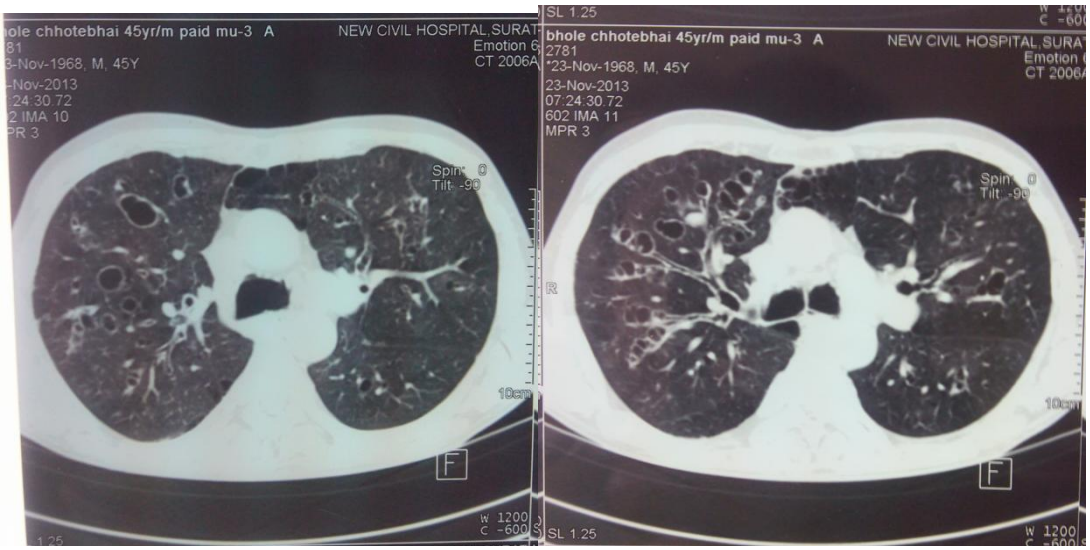


(Figure 1)



(**Figure 2)**

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