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Images in COPD

Images in COPD: Alpha-1 Antitrypsin Deficiency and Bronchiectasis

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Abbreviations: forced expiratory volume in 1 second, **FEV₁**; forced vital capacity, **FVC**; alpha-1 antitrypsin deficiency, **AATD**; computed tomography, **CT**

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Case: Two brothers aged 70 (RS) and 72 (WS) each with a history of alpha-1 antitrypsin deficiency are presented

RS presented with a 10-year history of cough together with multiple episodes of bacteria-laden purulent sputum production. A diagnosis of bronchiectasis was confirmed by chest radiographs and CT (Figure 1). Pulmonary function tests showed no evidence of airway obstruction with a forced expiratory volume in 1 second (FEV₁) of 3.2L (79% of predicted) and a FEV₁ to forced vital capacity (FVC) ratio of 0.77. Diffusing capacity for carbon monoxide was mildly decreased. The diagnosis of alpha-1 antitrypsin deficiency (AATD) (25mg/dl PI*ZZ) was made at age 66 and augmentation therapy was begun. Currently, his symptoms show improvement but intermittent exacerbations of purulent bronchitis still occur requiring antibiotic therapy.

WS has had a clinical profile of COPD for the last 10 years. Pulmonary function tests show moderately severe airway obstruction with an FEV₁ between 47% and 59% of predicted. Chest radiographs and computed

tomography (CT) showed evidence of panlobular emphysema (Figure 2). The most recent results show an FEV₁ of 1.35L (37% of predicted) and a FEV₁/FVC ratio of 0.57. The diagnosis of AATD was made at age 67 (17mg/dl PI*ZZ). Augmentation therapy was begun following the diagnosis of his disease.

Discussion

In 1963, association between emphysema and AATD was described by Laurell and Eriksson.¹ Since then, panlobular emphysema most frequently found in the lower lobes and to a lesser extent in the upper lobes has been most closely associated with AATD.² Although airway disease including chronic bronchitis and bronchiectasis have been reported in patients with AATD, the relationship of airway disease has not been clearly defined nor has the incidence of bronchiectasis been clearly established.³⁻⁶ King et al in 1996 found CT evidence of bronchiectasis in 6 of 14 patients and 2 patients in this cohort demonstrated diffuse cystic bronchiectasis without predisposing conditions other than the history of AATD.⁴ The authors stressed the importance of including AATD in the differential diagnosis of bronchiectasis and suggested that the presence of bronchiectasis may be important if a single lung transplant is planned since bronchiectasis in the remaining lung may predispose to infection during the post-transplantation period.⁴ Parr et al in 2007 concluded that the prevalence of clinically significant bronchiectasis was greater than previously recognized in a series of 74 patients of whom 20 (27%) had

Figure 1.

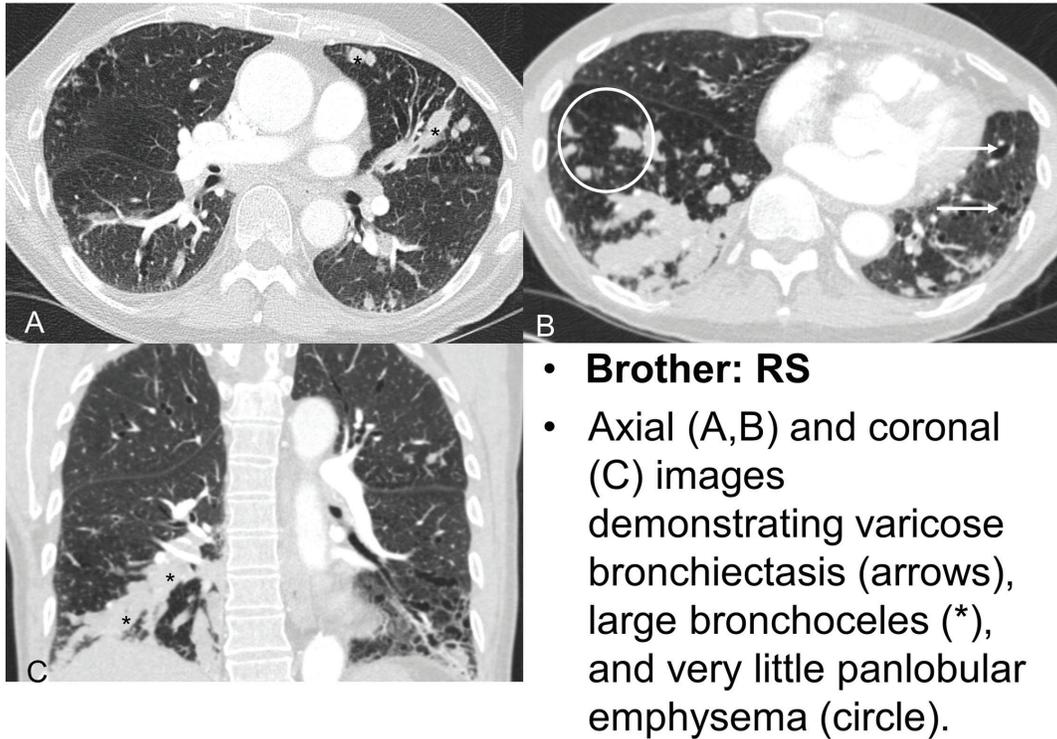
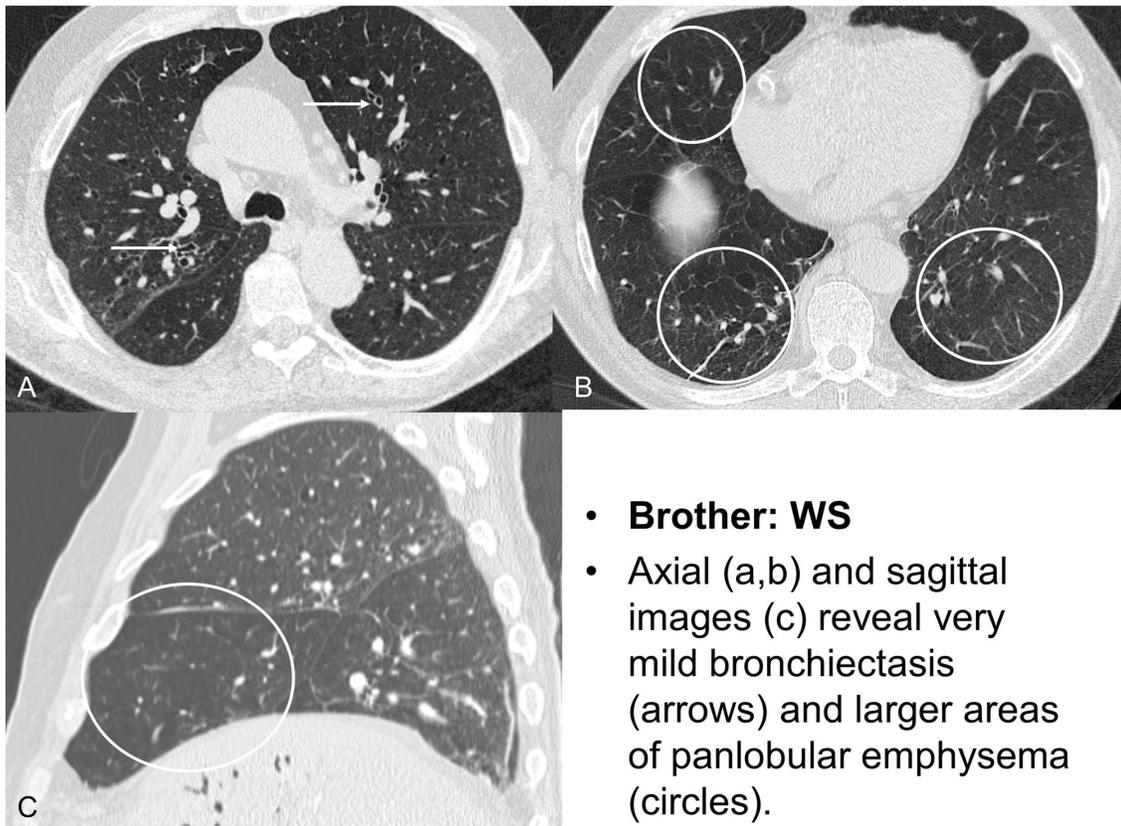


Figure 2.



radiologic evidence of bronchiectasis in 4 or more bronchopulmonary segments together with symptoms of regular sputum production.³

Two brothers are presented, one with predominately airway disease and evidence of bronchiectasis and the second with panlobular emphysema, little to no airway disease and pulmonary function test findings of

obstructive disease. These cases illustrate the spectrum of CT findings in AATD. The presence of bronchiectasis with or without emphysema should alert the clinician to include AATD in the differential diagnosis.

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