Recurrent Pneumonia in Kartagener’S Syndrome with Rheumatoid Arthritis

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Introduction: Kartagener’s Syndrome (KS) is a rare, autosomal recessive, ciliary disorder characterized by situs inversus, chronic sinusitis and bronchiectasis. Nontuberculous mycobacteria (NTM) have been described in bronchiectasis secondary to rheumatoid arthritis (RA), specifically in patients on DMARD therapy. Together, these two syndromes require a complex management strategy to reduce progression of joint and pulmonary disease. There have been ten reported cases of RA and KS together, while, this is the first reported case of treating NTM in rheumatoid arthritis and Kartagener’s syndrome. Case Report: 42-year-old Caucasian male presented with chronic cough and recurrent nasal drainage. He also had seropositive rheumatoid arthritis that has been resistant to many combinations of disease modifying agents (DMARDs) leading to reliance on chronic steroids. He reported a history of sinus infections since childhood with recurrent pneumonias as an adult. Chest radiograph revealed dextrocardia along with right lobe airspace disease. Initial sputum cultures showed multidrug resistant pseudomonas and patient was treated with appropriate antibiotics. After initial recovery, patient frequented the ED with complaints of persistent cough and joint pains. Computerized tomography of the thorax revealed right lower lobe bronchiectasis along with situs inversus. Given his constellation of symptoms, and negative CF genetic testing, he was diagnosed with primary ciliary dyskinesia. He was started on an aggressive airway clearance and bronchodilator regimen with nebulized albuterol, hypertonic saline, percussion vest, and postural drainage. Due to his Pseudomonas colonization, he was prescribed inhaled tobramycin on alternating months. During the subsequent months, there was persistence of sputum and recurrent exacerbations requiring intravenous antibiotics. Sputum cultures grew mycobacterium avium intracellulare (MAC) on multiple occasions. This was considered the cause of his recurrent exacerbations and he was started on azithromycin, ethambutol, and rifampin. Sputum cultures were negative for MAC two months after initiation of triple therapy. Through his course, his arthropathy did not specifically worsen with MAC medications. Discussion: Primary ciliary dyskinesia, pseudomonas colonization, MAC infection and rheumatoid arthritis all complicate this patient’s course and present therapeutic challenges. These disease states are all known to independently cause progression of bronchiectasis. Treatment difficulties arise when deciding if symptomatic management of RA, with immunosuppression, or infection eradication is the therapeutic goal. The choice of immunosuppression in the presence of chronic infections presents a dilemma as the use of DMARDs and steroids in RA have been associated with increased NTM infection specifically MAC.
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