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Repeated Hemoptysis With Progressive Bronchiectasis: A Case Report of Lady Windermere Syndrome

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1. Abstract

Bronchiectasis is a type of incurable structural lung disease with clinical manifestations of chronic cough, expectoration or recurrent hemoptysis, which is often given anti-infection and symptomatic treatment. In this study, a patient suffering from bronchiectasis with repeated hemoptysis caused by nontuberculous mycobacterium (NTM) was discussed. A 54-year-old female immunocompetent patient was admitted to our hospital due to repeated hemoptysis for 5 years. Computed tomography (CT) scan revealed progressive bronchiectasis in the upper and middle lobes of her right lung. She subsequently underwent thoracoscopic lobectomy of the right middle lobe plus segmentectomy of the anterior segment of the right upper lobe. Postoperative pathological diagnosis was confirmed to be intracellular mycobacterium. In view of her results, the patient was concluded to have "Lady Windermere syndrome" and was clinically cured following 15 months of anti-NTM treatment.

2. Introduction

Nontuberculous mycobacterial lung disease (NTM-LD) is a group of pulmonary infectious disease which is confirmed via respiratory secretions culture. Its symptoms are often non-specific. Bronchiectasis is one of the most common radiographic appearances of NTM-LD, which is a common respiratory disorder in clinical practice. Patients suffering from bronchiectasis are prone to infections, such as NTM. Due to their frequent coexistence, it is difficult to determine causality [1] and may even be misdiagnosed. Here, a case of intracellular mycobacterium lung disease is reported in an immunocompetent middle-aged female, presenting with repeated hemoptysis and progressive bronchiectasis for 5 years that meets the diagnostic criteria of Lady Windermere syndrome.

3. Case Report

A 54-year-old non-smoker female with no family or personal history of nontuberculous, was admitted to our hospital presenting with a 5-year history of recurrent episodes of cough, yellow phlegm and hemoptysis. She had been healthy without chronic pulmonary disease or connective tissue disease until her first bout of hemoptysis occurred in 2013, with 3-5 ml of bloody sputum. An initial thoracic computed tomography scan demonstrated mild localized bronchiectasis with a tree-in-bud sign at the middle lobe of her right lung (Figure 1A). Since then, the patient suffered from repeated attacks of hemoptysis once or twice a year, which gradually increased to 100 ml of blood on each bout of hemoptysis. She had already been six admissions by April 2018 for hemoptysis and pulmonary infection in which she was treated with antibiotics and hemostatic for presumed bronchiectasis with infection all the time. Meanwhile, the scope of bronchiectasis gradually expanded, involving the right middle lobe and the anterior segment of the right upper lobe (Figure 1B and 1C). During the course of her disease, she occasionally had fever but without dyspnea or weight loss. In terms of personal history, she had lived in Australia and Singapore since 2001 for five years. She decided to undergo surgery on account of the disease's serious impact on her quality of life. A physical examination demonstrated that her weight was 53 kg, height was 165cm, body mass index (BMI) was 19.4 Kg/

m2 and she had fine moist rales in her right lung. The laboratory data, including routine blood test, C-reactive protein, erythrocyte sedimentation rate (ESR), biochemical indexes, procalcitonin, and anti-HIV antibody were normal. T-SPOT test was negative. No abnormality was found in echocardiography. She underwent thoracoscopic lobectomy of the right middle lobe plus segmentectomy for anterior segment of her right upper lobe. Histological examination of the resected specimens revealed multifocal and chronic granulomatous inflammation with partial necrosis (Figure 2A) and positive acid-fast bacilli (Figure 2B), indicating mycobacterial infection. Identification of mycobacterium species by fluorescence quantitative polymerase chain reaction (PCR)-probe melting curve technique revealed Mycobacterium intracellular. She then received treatment for nontuberculous mycobacterium pulmonary infection with azithromycin, amikacin, rifampicin, ethambutol and moxifloxacin for 15 months. No bronchiectasis or other abnormal lesions were found in both lungs of her thoracic CT reexamination following drug withdrawal (Figure 1D). So far, the patient has been followed up for nearly one year and a half without hemoptysis after drug withdrawal.

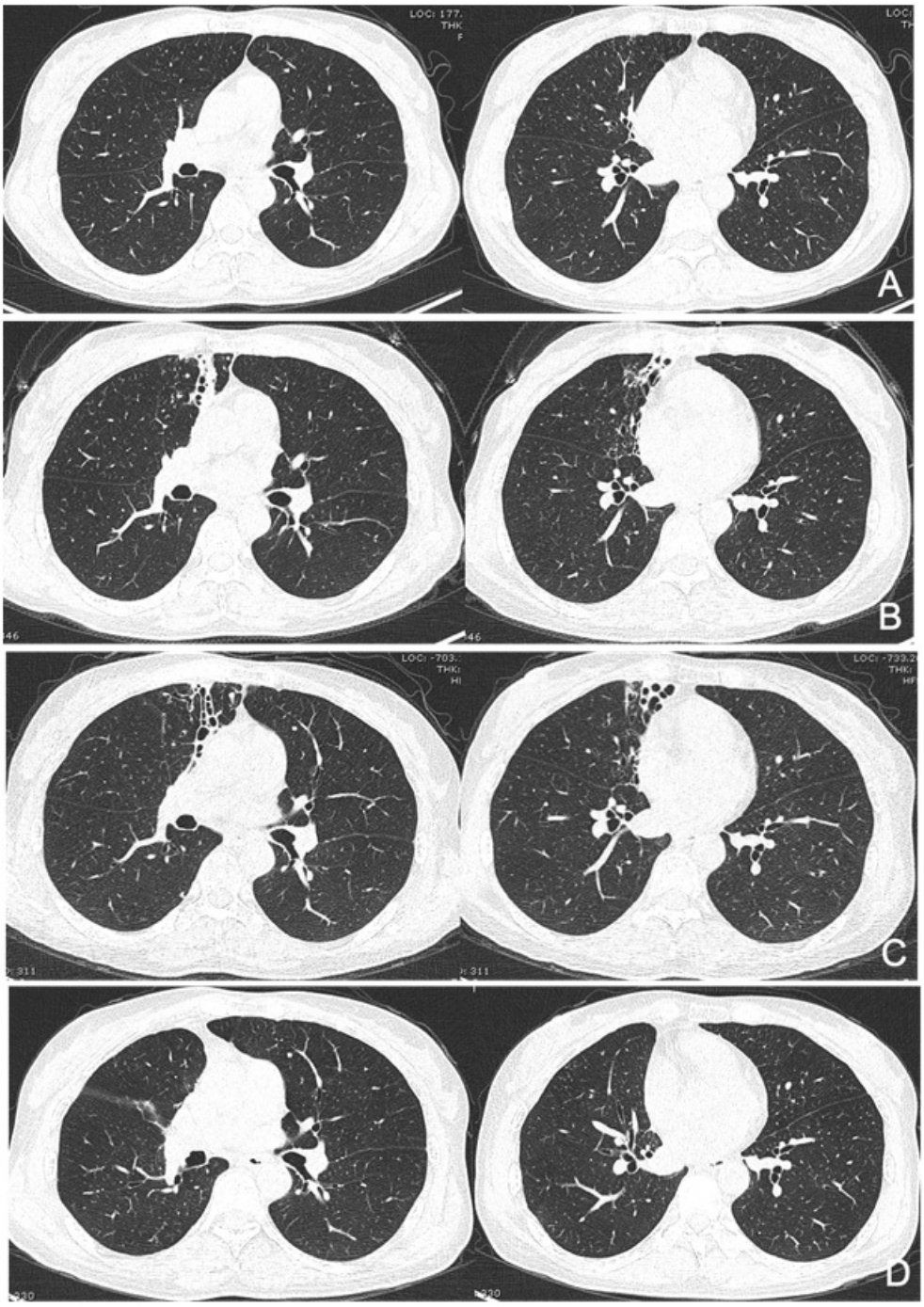

Figure 1: An initial thoracic computed tomography scan demonstrated mild localized bronchiectasis with a tree-in-bud sign at the middle lobe of her right lung.

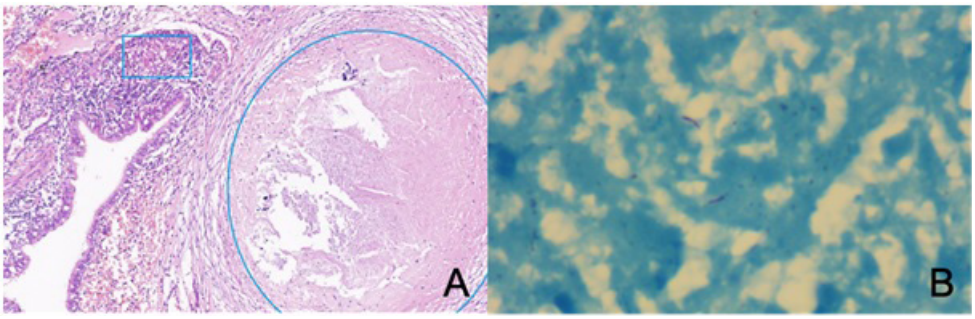

Figure 2: Histological examination of the resected specimens revealed multifocal and chronic granulomatous inflammation with partial necrosis and positive acid-fast bacilli indicating mycobacterial infection.

4. Discussion

Due to the increased presence of advanced diagnostic techniques and increased awareness among clinicians, the incidence of NTM infection has dramatically increased in the past two decades globally, which is also true in China [2-4]. Mycobacterium avium complex (MAC) is ubiquitous and has become a common finding worldwide in respiratory secretions and tissues [5]. The majority of humans infected with NTM develop a chronic illness that slowly progresses. Middle-aged patients are more likely to be infected with NTM, especially in female, which is referred to as Lady Windermere syndrome. Reich and other researchers [6] first described the syndrome in 1992, which was named after a fussy character in Oscar Wild's plays, "Lady Windermere's Fan". It is characterized that female patient without apparent predisposing pulmonary disease has habit of voluntary suppression of cough which induces to unable to clear the secretions from the right middle and left lingual lobe of lung. Since then, dozens of cases have been reported, primarily from the West (more likely to be the Caucasian) [7], though a few were reported from Southeast Asia [8, 9] and Taiwan [10]. However, no cases have been reported from China.

Postmenopausal women who are tall and have a thin morphotype and low BMI, scoliosis, pectus excavatum and mitral valve prolapse are prone to this disease, which is a unique phenotype for this disease. Host factors of NTM PD also include rheumatoid arthritis and use of immunomodulatory drugs. Moreover, warm, humid environments with high atmospheric vapor pressure contribute to population risk [2]. Although the pathophysiology for susceptibility to NTM lung disease in these patients remains unclear, multiple factors interacting endocrine systems have the potential to contribute to Lady Windermere syndrome by modulating immune responses to NTM [11]. Another study identified that rare variants in MST1R contribute to Lady Windermere syndrome by decreasing airway ciliary function and reducing IFN-γ production in response to NTM [12].

The diagnosis comprises three components of microbiologic, radiographic, and clinical criteria. The clinical criteria include cough, sputum, occasional hemoptysis, and febrile episodes. Radiologically, chest X-ray or CT scans show a spectrum of reticulonodular opacities, a tree-in-bud nodularity, and cavitary lesions superimposed on cylindrical bronchiectasis of the right middle lobe and left lingular segments. Furthermore, sputum culture or bronchoalveolar lavage culture demonstrates the infection caused by MAC. In the present case, the patient had a low BMI, was perimenopausal and had repeated hemoptysis as the main clinical manifestation. The patient's imaging demonstrated progressed bronchiectasis, and MAC lung disease was confirmed by postoperative pathology, which was found to be in accordance with Lady Windermere syndrome.

Regarding the treatment of NTM lung disease, it is important to assess the pathogenicity of the organism, risks and benefits of therapy, the patient's wishes and ability to receive treatment. The American Thoracic Society （ATS） guidelines suggest initiation of treatment in patients who meet the diagnostic criteria for NTM pulmonary disease, especially in the context of positive acid-fast bacilli sputum smears or cavitary lung disease. In this regard, the ATS strongly recommends that the initial therapy for patients with macrolide-susceptible MAC lung disease consist of a minimum three-drug regimen that includes azithromycin rather than clarithromycin [13]. Although medical therapy remains the primary treatment modality for patients with pulmonary NTM disease, pulmonary resection may reduce the incidence of symptomatic disease recurrence [14]. Surgical resection is appropriate in cases of localized disease or resistant to medical therapy [15].

5. Conclusion

Whereas bronchiectasis is a common respiratory disorder encountered in clinical practice, NTM infections should be considered in atypical scenarios. When patients with repeated hemoptysis and bronchiectasis seek health care, especially in middle-aged women with bronchiectasis in the right middle lobe and lingular segments, clinicians should suspect the possibility of NTM diagnosis.

6. Acknowledgement

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