

LETTERS TO THE EDITOR

Lady Windermere Syndrome

To the Editor: Respiratory infection by environmental mycobacteria in immunocompetent patients is an increasingly important epidemiologic concern.¹ Of all lung diseases caused by environmental mycobacteria, infection by *Mycobacterium avium* complex has the second-highest incidence in immunocompetent patients in Spain; in other parts of the world, such as North America, it is the most prevalent.² Less common clinical patterns of infection by this environmental mycobacterium have been defined in recent years. One such pattern relates specifically to older nonsmoking Caucasian and Asiatic women with no remarkable history of respiratory disease.

We describe the case of a 61-year-old woman from South Korea, where she lived until the age of 30 years. At that age, she took up residence in the south-central region of the United States of America. With a history of cirrhosis of the liver and hepatocellular carcinoma related to a hepatitis B virus infection under treatment with lamivudine, she was on the wait list for a liver transplant. In addition, she had undergone splenectomy after detection of an aneurysm in the splenic artery and had had breast augmentation surgery. She was an ex-smoker (5 pack-years). Noteworthy was the fact that 20 years earlier she had had a positive Mantoux test but no subsequent chemoprophylactic treatment and no symptoms indicative of tuberculosis during that period. The patient reported a history of chronic cough, which she voluntarily suppressed and which had been accompanied by expectoration for 6 months. She presented no additional general systemic syndromes or other symptoms. The results of physical examination were normal. DNA probe assays detected *M avium* complex in 3 serial sputum samples, prompting us to perform contrast enhanced chest computed tomography (Figure). The scans showed bronchiectasis, adjacent areas of consolidation, and a tree-in-bud pattern in the medial segment of the middle lobe and the lingula. Also visible were slightly enlarged mediastinal lymph nodes, some of which were calcified. In lung function tests, spirometry showed a forced vital capacity (FVC) of 2850 mL (107%), a forced expiratory volume in the first second (FEV₁) of 2000 mL (92%), and a FEV₁/FVC ratio of 70%. Carbon monoxide diffusing capacity was 112% and lung capacity by the helium dilution method was 4.46 L. We decided to perform bronchoscopy; macroscopic inspection

revealed normal mucosa, but the bronchoalveolar lavage fluid from the lingula tested positive for nontuberculous mycobacteria. The patient met the American Thoracic Society criteria for environmental mycobacterial infection and was diagnosed with Lady Windermere syndrome. The decision was therefore to initiate treatment with azithromycin, rifampicin, and ethambutol and to refrain from ruling out transplantation in spite of the postoperative immunodepressor treatment. The patient's subsequent progress was satisfactory.

In 1992 Reich and Johnson³ first described a specific syndrome based on 6 cases of immunocompetent elderly women with no significant history of smoking or respiratory disease who developed *M avium* complex infection in the middle lobe and lingula. The authors hypothesized that voluntary suppression of cough and expectoration, thus hindering drainage of secretions, gave rise to a local inflammatory response that predisposed the middle lobe and lingula to infection by *M avium* complex. They gave it the name of Lady Windermere syndrome after the character of a Victorian lady in Oscar Wilde's play.³ The hypothesis was subsequently confirmed by reports of other cases.^{4,5} This syndrome, which is not described in other case series published in Spain,² seems to affect only women, due to their aforementioned social habit of suppressing cough, and only involves the middle lobe and lingula because these segments' long, narrow bronchi are predisposed to accumulate respiratory secretions in such a way that the cough effort required to maintain bronchial hygiene is greater there than for other parts of the lung. Menopause and the consequent decline in estrogens and progesterone may also play an important role in the onset of infection by environmental mycobacteria by decreasing immune defenses in previously colonized menopausal patients.⁶ After reviewing the literature, we believe that our patient's splenectomy—an intervention that predisposes mainly to infections by capsulated microorganisms—had no causal relation to the infection by *M avium* complex.

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Chest computed tomography scan of the lung window showing bronchiectasis in the middle lobe and lingula.