Atypical mycobacterium pulmonary infection: Lady Windermere Syndrome

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Resumen

Introducción: La infección respiratoria por micobacterias atípicas en pacientes inmunocompetentes es cada vez más relevante desde el punto de vista epidemiológico. En los últimos años se han definido patrones clínicos característicos. Uno de ellos es el que ocurre en mujeres de edad avanzada, no fumadoras y sin antecedentes de enfermedad respiratoria previa, conocido como síndrome de Lady Windermere. Objetivo: Presentar los hallazgos imagenológicos característicos del síndrome de Lady Windermere y otras infecciones pulmonares causadas por micobacterias atípicas. Discusión: En 1992 Reich y Johnson describieron por primera vez un síndrome específico en mujeres inmunocompetentes de edad avanzada y sin antecedentes significativos de tabaquismo o enfermedades respiratorias, quienes desarrollaron...

Abstract

Introduction: Respiratory tract infections caused by atypical mycobacterium in immunocompetent patients are becoming increasingly important from the epidemiological point of view. In recent years, characteristic clinical patterns have been defined. One of them is seen in elderly women who do not smoke and who do not have a history of the respiratory disease called Lady Windermere Syndrome. Purpose: To present characteristic imaging findings of Lady Windermere Syndrome and other pulmonary infections caused by atypical Mycobacterium. Discussion: In 1992, Reich and Johnson described for the first time a specific syndrome in immunocompetent elderly women who did not have a relevant history of smoking habits or respiratory diseases and who developed an infection caused by Mycobacterium Avium. It is characterized by plurifocal...
Introduction

The term “Atypical Mycobacteria” has been used to group a collection of the genus Mycobacterium (M), which includes M. Tuberculosis and M. Leprae. These species include: Intra-cellular M. Avium, M. Kansaii, M. Xenopi, M. Malmoense and rapidly growing Mycobacterium. This group of Mycobacteria produces pulmonary disease, ganglionar or disseminated. Respiratory tract infections caused by atypical mycobacterium in immunocompetent patients are becoming increasingly important from the epidemiological point of view. In recent years, two clinical and radiological patterns have been defined in immunocompetent patients. Lady Windermere Syndrome is seen in elderly women who do not smoke and who do not have a history of a significant respiratory disease, except for bronchiectasis. The second pattern defined is the cavitary disease of the upper lobe (Tuberculosis-like) present in male patients with a history of smoking habits and pulmonary fibrosis (2).

Discussion

Bacteria of the genus Mycobacterium belong to the family of Mycobacteriaceae. They have a wall with a thick, lipid-rich layer with special properties of staining, mainly acid-alcohol-resistant. There are two groups of Mycobacteria, those pathogenic specific of men or animals, which transmit Tuberculosis (Mycobacterium Tuberculosis, Mycobacterium Bovis) and Lepra (Mycobacterium Leprae). The second group includes the species, which are usually not pathogenic, specific of men. In 1954, Timpe and Runyon proved that Atypical M. has a pathogenic power. In 1959, Runyon establishes a classification based on pigmentation of the bacterial colony and the growth speed of Atypical Mycobacterium (3). They are aerobic, non-motile, nonsporulating and characteristically acid-alcohol-resistant. They are widely distributed in the atmosphere: water, soil, dust, food, birds and other animals (1-3). They can inhabit bodily surfaces and secretions without causing diseases.

Atypical Mycobacteria include: M. Avium Complex (avium- intracellulare), M. Kansaii, M. Xenopi, M.
Lady Windermere Syndrome

Malmoense, among others. Only some of Atypical M. have a proved pulmonary pathogenesis, such as M. Avium Complex, M. Kansassi, M. Xenopi and M. Malmoense. From this group, those which frequently cause respiratory tract infections in human beings are M. Avium and M. Kansassi (1-4).

The importance of these agents has increased in relation to the advances of diagnostic procedures and the description of clinical features they produce, as well as the predisposition to their development in immunocompromised patients infected with HIV. Nowadays, respiratory tract infections in immunocompetent patients have an increasing importance.

The two main presentation forms are fibrocavitary disease, predominantly in young or elderly men who smoke, clinically and radiologically similar to TBC (Tuberculosis Like) and Lady Windermere Syndrome in elderly women who do not smoke (2-5). Another form of presentation is the syndrome known as Hot Tub Lung. It is a hypersensitivity reaction due to inhalation of Mycobacterium Avium by users of hydro massage tubs (3).

The fibrocavitary form is radiologically presented as cavities with thin walls in upper lobes in up to 90% of the cases, accompanied by perilesional ground glass density pulmonary areas (Figure 1). They are also accompanied by reactive pleural thickening. Germs that produce Tuberculosis-like with a greater frequency are Mycobacterium Avium and Mycobacterium Kansasii (2).

Lady Windermere Syndrome was described for the first time in 1992 by Reich and Johnson who registered a specific syndrome in women who were over 50 years of age, immunocompetent and without a relevant history of smoking habit or respiratory disease developing infection caused by Mycobacterium Avium Complex. It is characterized by the presence of bilateral bronchiectasis in the middle lobe and lingula, associated with signs of multifocal bronchiolitis. It can also be associated with scoliosis, pectus excavatum and mitral valve prolapse. Regarding pathologic mechanisms, the germs of Mycobacterium Avium Complex are capable of compromising the airway tract, especially bronchi in the middle lobe and lingula, and due to anatomical characteristics, it is difficult to eliminate secretions. If we add suppression of expectoration to these factors, as it is the case in women, there is an accumulation and thickening of secretions in favor of the development of mycobacteria (4-5).

There is controversy regarding bronchiectasis, since these germs themselves can produce them due to the intense inflammatory reactions, but they can also compromise preexistent bronchiectasis (3). The main clinical manifestations are coughing, dyspnea, fever, weight loss, and occasionally hemoptysis. CT findings consists of bronchiectasias and multiple centrilobular nodules associated with the tree-in-bud pattern affecting predominantly the middle lobe and lingula, without compromising upper lobes (Figures 2 and 3).

Finally, there is a clinical presentation called «hot tub lung». It is a disease caused by Mycobacterium Avium Complex especially in people who frequently go to spas or saunas (Mycobacterium Avium develops in warm water and is resistant to most germicides). Antigens of Mycobacterium Avium would be responsible of an immune response causing hypersensitivity pneumonia (3).

Conclusion

The frequency of respiratory tract infections caused by atypical Mycobacterium is increasing in everyday practice. Recognizing the clinical and radiological signs allows the suspicion of this pathology and thus biochemical diagnostic methods can be applied to reach an early diagnosis.
Figure 1. “Tuberculosis Like” fibrocavitary form.
65-year-old patient, with smoking habits who presented symptoms of cough, dyspnea and weight loss during 11 months. Bacilloscopy results were negative. Bronchoalveolar lavage was performed and Mycobacterium Avium was identified at bacterial culture.

A-B) High-resolution CT shows the presence of an apical thin-walled cavity (thick white arrow), surrounded by consolidated areas, ground glass density areas (thin white arrow) and pleural thickening (black arrow).

Figure 2. Lady Windermere Syndrome.
Female 63-year-old patient who presented symptoms of cough, dyspnea and weight loss during 8 months. Bacilloscopy results were negative and treatments with antibiotics did not work. Bronchoalveolar lavage was performed and Mycobacterium Avium was identified at bacterial culture.

A-B) Axial views in CT. Note the presence of multiple centrilobular micronodules (thin white arrow), bronchiectasis (thick white arrow) and isolated ground-glass density areas (black arrow). Those findings compromised the middle lobe and lingula. There were centrilobular micronodules disseminated through the inferior lobes.
Figure 3. Lady Windermere Syndrome.
65-year-old patient with symptoms of cough, dyspnea, night fever episodes during 4 months. After finding Mycobacterium Avium in bronchoalveolar lavage, a diagnosis was made.
A) Axial CT at pulmonary parenchyma window showing centrilobular micronodules in the middle lobe and lingula (thin white arrow) and bronchiectasis (thick white arrow). B) MIP reconstruction of CT. Note the tree-in-bud pattern (white arrow).

Bibliography