

# Lady Windermere syndrome: an inappropriate eponym for an increasingly important condition

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## ABSTRACT

**Non-tuberculous mycobacterial infection (NMI) occurs in elderly women with no pre-existing lung disease, and this has been termed the Lady Windermere syndrome. NMIs are increasing in prevalence and an increasing number of pulmonary mycobacterial infections is due to non-tuberculous mycobacteria. The diagnosis is often difficult because the organism is not readily isolated or cultured, and the condition may not be considered by the radiologist. We report NMI in a 64-year-old woman, based on clinical and radiological findings. Although termed the Lady Windermere syndrome, the name does not correspond to the character in Oscar Wilde's play; hence the eponym is not widely used.**

**Keywords: high-resolution computed tomography, Lady Windermere syndrome, non-tuberculous mycobacterium infection, tree-in-bud appearance**

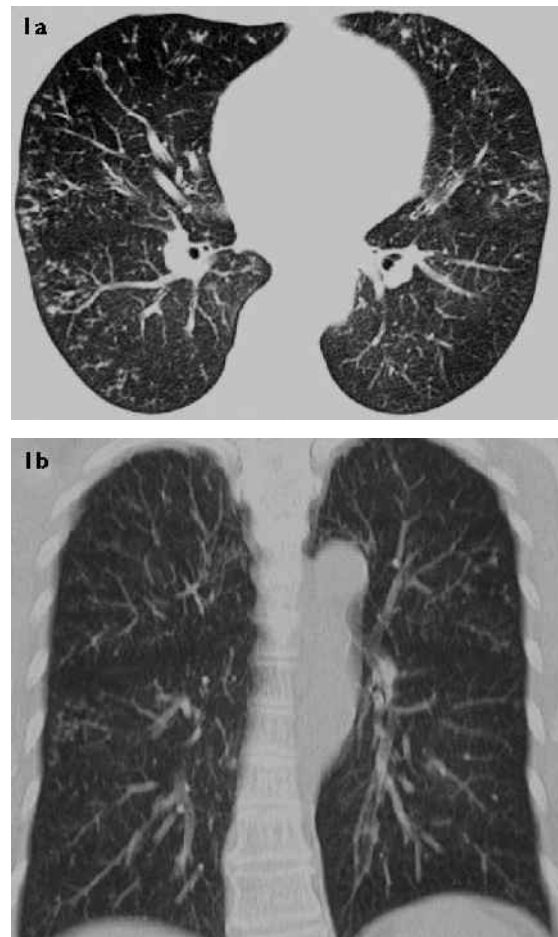
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## INTRODUCTION

Non-tuberculous mycobacteria (NTM) represent a significant proportion of mycobacterial infections and may prove difficult to diagnose due to their non-specific clinical and radiographical presentations. Timpe and Runyon reported the first case of pulmonary non-tuberculous mycobacterial infection (NMI) in 1954.<sup>(1)</sup> The clinical presentations of this infection vary. The three most common forms of presentations in the immunocompetent host are: pre-existing lung disease with fibrocavitary and/or reticulonodular appearance; no pre-existing lung disease with focal bronchiectasis and nodular lesions in right middle lobe and lingula—the “Lady Windermere syndrome”; and atypical presentations like focal masses and solitary nodules. Reich and Johnson first used the term, Lady Windermere syndrome, in 1992.<sup>(2)</sup> We observed a case of Lady Windermere syndrome in an elderly woman with no pre-existing lung disease which fit the description given in previous literature.

## CASE REPORT

The patient was a 64-year-old woman, who had no



**Fig. 1** (a) Axial and (b) coronal HRCT images taken at presentation show a nodular and tree-in-bud appearance peripherally in both lungs, more marked on the right side with underlying ground glass opacities.

previous respiratory illness and who complained of progressive shortness of breath for the past six months, especially when climbing stairs, and found it difficult to partake in her hobby of mountain climbing. She had no fever or other respiratory symptoms, but she had lost 2 kg in weight. She smoked one pack of cigarettes a day for two years, 17 years ago. She went to a health screening centre where a comprehensive range of investigations, including high-resolution computed tomography (HRCT) of the chest, was performed. Physical examination did not reveal any abnormality and blood biochemical investigations were normal. The HRCT showed small centrilobular nodules and a “tree-in-bud” appearance with

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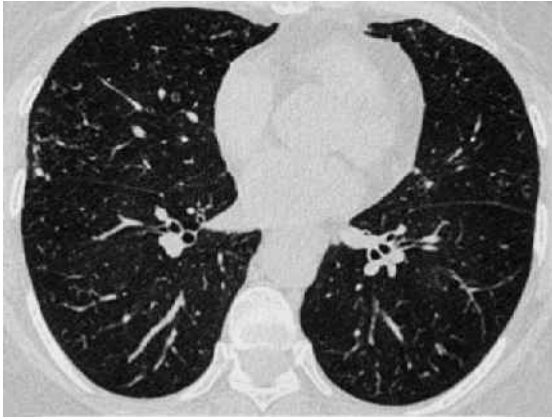
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**Fig. 2** Axial HRCT image shows a marked improvement after nine weeks of treatment.

a background of ground glass opacities (Fig. 1). Findings on flexible bronchoscopy were normal, bronchioalveolar lavage specimens were negative for acid-fast bacilli, and pyogenic organisms and cultures yielded no growth. Her Mantoux tuberculin skin test reaction measured 11 mm in diameter.

In view of the likely diagnosis of NMI, the patient was started on isoniazid (300 mg once daily), ethambutol (800 mg once daily) and clarithromycin (500 mg twice daily). Her breathlessness on exertion improved and she gained 2.5 kg in weight. A repeat HRCT nine weeks later showed a considerable improvement (Fig 2). In spite of the inability to isolate an organism, the diagnosis of an NMI was presumed to be correct. Antimycobacterial chemotherapy with the same drugs was planned for at least another ten months, depending on the clinical and HRCT response.

## DISCUSSION

There are up to 20 species of NTM, also known as atypical mycobacteria, associated with infection in humans. Unlike *Mycobacterium tuberculosis*, the NTM organisms are ubiquitous, and infection arises from inhalation of organisms from reservoirs in the environment, such as soil and dust. The commonest NMIs in humans are *Mycobacterium avium intracellulare* and *Mycobacterium kansasii*.<sup>(3,4)</sup>

In the non-immunocompromised patient, NMI occurs broadly in two groups of patients. One group consists usually of males of over 50 years of age with a pre-existing lung disease, commonly, chronic obstructive airways disease. The other group comprises thin, elderly females with no pre-existing lung disease and often no history of smoking. In the latter group, NMI has been named the Lady Windermere syndrome. The most common radiological finding in NMI is a fibronodular bronchiectasis with a predilection for the right middle lobe and lingula. This pattern is most commonly seen in *Mycobacterium avium* complex, but can be caused by other NTMs including

*Mycobacterium abscessus* and *Mycobacterium kansasii*.

The clinical presentation is variable, with complaints of fever, weight loss, dyspnoea, malaise, cough and haemoptysis. The disease may have an insidious onset and symptoms may be present for months or years before the diagnosis is made. Early cases may be asymptomatic and only discovered by routine screening chest radiographs.<sup>(4)</sup> Diagnosis can be made by polymerase chain reaction, but identification of the organism from the sputum is difficult. A multiple simultaneous skin test, which is the NTM equivalent of the Mantoux test, is available. Treatment consists of a combination of anti-mycobacterial drugs to prevent emergence of resistant organisms, and should be continued for a further 12 months once culture findings are negative, or 24 months for those who are initially culture positive.<sup>(5)</sup>

Radiologically, there is an overlap between the appearances of NTM and *Mycobacterium tuberculosis*. The 'classical' appearance, often indistinguishable from post-primary tuberculosis, tends to occur in men with pre-existing lung disease. The 'non-classical' appearance of NMI is usually seen in elderly women with no pre-existing lung disease, where the radiological findings are mild bronchiectasis, centilobular nodules and a tree-in-bud appearance on HRCT, indicating small airways disease.<sup>(6-8)</sup> This tree-in-bud appearance is due to dilated centrilobular bronchioles with their lumen, filled with mucus, fluid or pus. This appearance was previously thought to be due to a *Mycobacterium tuberculosis* infection, but it is now recognised to occur in many conditions, such as other infections, inhalation of toxic fumes, aspiration, immunological and connective tissue disorders and pulmonary intravascular tumour embolism.<sup>(9)</sup> In the present case, the only diagnosis that really fit with the clinical symptoms was infection. The long history and weight loss made mycobacteriosis the most likely diagnosis, but the Mantoux reaction of 4 mm and the fact that no organism could be cultured, supported the diagnosis of NMI.

The diagnosis of NMI is often difficult as the organism is not easily isolated or cultured. Moreover, as a relevant skin test is not readily available, the clinical history, physical examination and radiological findings are of paramount importance. The condition has been named Lady Windermere syndrome,<sup>(2)</sup> as the name is based on the aphorism, "ladies don't spit". A lady, as described in literature, is a woman of good social standing and has a polite and refined disposition. Ladies are assumed to be fastidious, and as spitting is socially unacceptable, they voluntarily suppress their cough and infected secretions are retained in the lungs. The name, Lady Windermere, comes from Oscar Wilde's play, *Lady Windermere's Fan*, written in 1892, and is a witty dramatisation of manners and morals in Victorian upper class society. The fastidious

behaviour is illustrated by Lady Windermere's remark, "How do you do, Lord Darlington? No, I can't shake hands with you. My hands are all wet with these roses." However, Lady Windermere was a vivacious 21-year-old girl, who was married for two years and had never coughed or showed any other illness. Although the name Lady Windermere is rather inappropriate and hence has not come into general use, NMIs must be considered in elderly women with unexplained respiratory symptoms and a tree-in-bud appearance on HRCT.

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