中文題目：先前無明確肺部疾病的 70 歲女性有大量咳血

英文題目：A 70-year-old woman without previously documented lung disease presented with life-threatening hemoptysis.

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Introduction: Bronchiectasis, typically involving the right middle lobe or lingula induced by non-tuberculous mycobacterium (NTM)infection, was referred to as Lady Windermere syndrome. Most patients without underlying pulmonary disease are female predominant, especially those who tend to suppress their cough ability. We described a case that first presented as fresh blood vomitus and was diagnosed as Lady Windermere syndrome with right middle lobe bronchiectasis followed by Mycobacteria abscesses complex infection

Case presentation: A 70-year-old Taiwanese woman without any underlying disease, suffering from intermittent fresh blood vomitus for one day. Associated symptoms included productive cough for one month, but denied fever, dyspnea, and chest tightness. She also denied previous anticoagulant medication use. She visited Far Eastern Memorial Hospital emergency department and was treated as upper gastrointestinal bleeding first. Upper gastrointestinal endoscopy was arranged, and the gastric submucosal (subepithelial) tumor was impressed, but no active bleeding was found. The blood test revealed no coagulopathy or thrombocytopenia detected.

Chest X-ray revealed small patches of infiltrate in bilateral lungs . Chest computed tomography showed ill-defined ground glass opacities and tree-in-bud lesions in the left lung and right lower lung, in favor of infectious process and bronchiectasis, atelectasis in right middle lung, and lingular segment of left upper lung. The patient was discharged with oral medication but returned to the emergency department on the same day due to persistent symptoms. The patient was admitted and intubated later due to massive hemoptysis for airway protection, suspected related to bronchiectasis. Due to refractory hemoptysis, although no active contrast extravasation was revealed by computed tomography angiography, empirical transarterial embolization of left bronchial artery was performed. Further bronchoscopy showed no active bleeder, but bronchial washing showed a positive acid-fast stain with a negative result of GenExpert tuberculosis polymerase chain reaction test. The final culture report showed Mycobacteria abscesses complex. The patient had no under-specific anti-NTM therapy for Mycobacteria abscesses complex. The patient was extubated smoothly later and now has regular outpatient clinic follow without O2 support.

Discussion: Lady Windermere syndrome was firstly described by Reich and Johnson in 1992, named after a fastidious character in “Lady Windermere’s Fan.” The pathogenic

theory assumed that people, especially old women, had long-term suppressed voluntary coughing habits, in order to avoid embarrassment in public.

There are many types of image features of NTM infection, including fibrocavitary form predominantly in chronic obstructive pulmonary disease patients, upper lobe bronchiectasis in cystic fibrosis of youngsters, but Lady Windermere syndrome mostly presented as right middle or left lingular lobe nodular bronchiectasis, consequently also known as nodular-bronchiectatic phenotype of pulmonary NTM disease.

Lady Windermere syndrome is caused by NTM infection related right middle or left lingular lobe bronchiectasis, mostly by Mycobacterium avium complex, a slow-growing NTM. It usually manifests as chronic cough and other nonspecific symptoms, such as fatigue, weakness, weight loss, and hemoptysis, which is similar to tuberculosis infection.

Our patient was infected by Mycobacteria abscessus complex. Unlike Mycobacterium avium complex, is a rapid-growing NTM and notorious for its poor prognosis because of its multidrug-resistant. It presented as bonchiectasis-induced massive hemoptysis with acute respiratory failure, but resolved uneventfully after empirical trans-arterial embolism, without specific anti-NTM antibiotic use. From reviewed literature and case reports so far, the patient is the first documented case of Lady Windermere syndrome caused by Mycobacteria abscessus complex to our best knowledge.

Our patient denied habitual voluntary cough suppression. Except for this proposed mechanism, there are still other potential pathogenic factors, such as endocrine changes and connective tissue defects. In addition, the vulnerability of females was challenged since this disease is an infectious disease rather than an X-linked inheritance disorder. The pathogenesis of Lady Windermere syndrome still left much to be investigated

Conclusion: We introduced a case of Lady Windermere syndrome, who had no typical habit of cough suppression, and was infected by a rare pathogen such as Mycobacteria abscessus complex. Despite initially presenting as life-threatening hemoptysis, the patient was discharged without significant sequelae after empirical left bronchial artery embolism.