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RESEARCH ARTICLE

A CASE OF BRONCHIECTASIS PRESENTING TO EMERGENCY DEPARMENT WITH SYMPTOMS OF RARE ENTITY "LADY WINDERMERE SYNDROME"

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ABSTRACT

Introduction Reich and Johnson (1) first used the term "Lady Windermere syndrome"in 1992. They described 6 elderly women who were immunocompetent, had no significant smoking history or underlying pulmonary disease, and developed Mycobacterium avium complex (MAC) pulmonary infection limited to the right middle lobe or lingula. They hypothesized that these women could have had the habit of voluntary suppression of cough, responsible for the inability to clear the secretions from the right middle lobe and lingula. This habit results in a focus of inflammation in these areas, which in turn predisposes to MAC infection. They named this condition Lady Windermere syndrome after Oscar Wilde's Victorian-era play Lady Windermere's Fan to suggest the fastidious behavior. Because it was a retrospective study, no history of voluntary cough suppression was obtained from these patients. We report a similar case in a 51-year-old woman who had bronchiectasis and associated right lower lobe infection due to MAC. Our patient had a significant history of lifelong habitual voluntary cough suppression.

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INTRODUCTION

Reich and Johnson (Samjot Singh Dhillon, 2000) first used the term "Lady Windermere syndrome"in 1992. They described 6 elderly women who were immunocompetent, had no significant smoking history or underlying pulmonary disease, and developed Mycobacterium avium complex (MAC) pulmonary infection limited to the right middle lobe or lingula. They hypothesized that these women could have had the habit of voluntary suppression of cough, responsible for the inability to clear the secretions from the right middle lobe and lingula. This habit results in a focus of inflammation in these areas, which in turn predisposes to MAC infection. They named this condition Lady Windermere syndrome after Oscar Wilde's Victorian-era play Lady Windermere's Fan to suggest the fastidious behavior. Because it was a retrospective study, no history of voluntary cough suppression was obtained from these patients. We report a similar case in a 51-year-old woman who had bronchiectasis and associated right lower lobe infection due to MAC. Our patient had a significant history of lifelong habitual voluntary cough suppression.

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CASE REPORT

CASE: A 51 year old female presented to emergency department at 2:05 pm. With history of intermittent episodes of persistent cough, occasionally productive of sputum and mild excertional dyspnea. She has be treated for community acquired pneumonia and her symptoms had initially improved, but she experienced discrete, recurrent episodes of shortness of breath with productive cough. Testing for tuberculosis had been negative. Her medical history included dilated cardiomyopathy, severe MR, acute LVF (LVEF 10-15%). On arrival in emergency, her airway was patent, breathing was unlabored, symmetrical, bilaterally wheeze, crepts, respiratory rate was 22/min, spo2 -85% on room air, blood pressure 130/80, pulse 112. On examination, the patient was moderate build. She appeared calm. When she coughed, she tried to suppress her cough. Expectorated sputum sample were obtained. Stains for acid fast bacilli were negative. Serology studies were negative for fungal infection Based on her symptom and clinical finding ,imaging studies (HRCT CHEST) and sputum culture ,we arrived at diagnosis of nontuberculous mycobacterium lung infection, specially ,Lady windermere syndrome.

PRIMARY SURVEY

Airway- patent,
Breathing -rr-20 spo2-85%on roomair
Circulation-bp 130/70mmhg pulse – 112beat/min
Dissability- gcs-15/15 with no focal neurological deficiet seen
Exposure - no abdominal mass or ecchymosis seen

SAMPLE HISTORY

S- shortness of breath assocaited with cough, dyspnea on exertion
A – no known drug allergies
M- tab ecosprin, tab clopitab, tab atorva,tab dytor plus,tab cardivas, tab ivabeat,tab doxibid,tab montair fx
P- acute lvf(recovered),lrti,dilated cardiomyopathy,severe lv dysfunction (15%),severe mr
L- 3 hrs pta
E- there was no significant event prior to arrival

SECONDARY SURVEY

HEENT: no pallor Icterus cyanosis seen.jvp not raised, neck gland not palpable, no neck mass, tongue dry.
CHEST –
On inspection- there was no deformity or scar mark seen and no swelling or lump on the chest seen, and equal chest rise is present
On palpation-- equal chest rise, no tenderness seen
On percussion—chest is bilateral resonant on percussion
On auscultation – bilateral, wheezes and crepitus
CVS-first and second heart sound audible, no murmur or friction rub sound heard
CNS- patient is conscious, and oriented to time. Place and person .higher mental function within normal limit .motor and sensory function bilateral limb normal, no flapping tremor seen

ABDOMEN

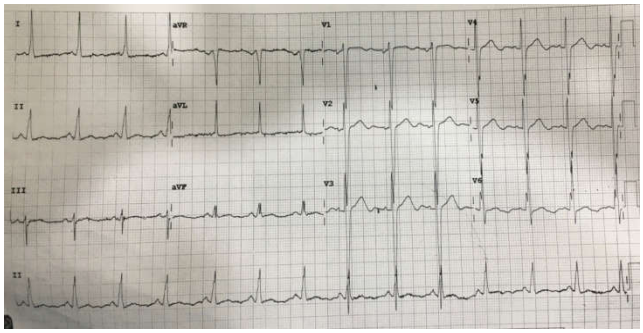
Inspection: No scar seen, no ecchymosis, no swelling, umbilicus normal and in position, no engorged vein seen
Palpation: Soft. There is no rebound tenderness and no guarding or rigidity seen. No palpable mass felt and normal bowel sound.
Percussion: Dull on percussion, no fluid thrill or shifting dulness observed.
Auscultation: Bowel sound heard, no bruit heard, no inguinal hernia seen, normal external genitalia ,per rectal examination was within normal limit

Extremities—within normal limit

TREATMENT IN ED

In view of the finding, point of care (POC) Test were sent which includes ,electrocardiogram which was suggestive of Normal Sinus Rhythm with Left Bundle Branch Block(LBBB).Patient's Blood Gas Analysis, revealed Metabolic Alkalosis pH 7.522,Pco2 48.1,HCO3 39.2,NA+ 122,K+ 2.4,LAC 1.0. Patient was canulated with 18G canula over the left anti-cubital vein.

- Nebulization with levosalbutamol (0.6) and budesonide (0.5)
- Injection pantoprazole 40mg iv stat
- Injection ondensetron 4mg iv stat
- Inj furosemide 40 mg iv stat
- Inj hydrocortisone 200 mg iv stat
- Cardiology and pulmonology opinion taken



The patient was re-evaluated after 15 min, presenting symptoms didn't subsides was advice bipap support (6:12)

Re-evaluation done in ED.

Airway- maintained and talking. Breathing- Bilateral mild to moderate wheezes and crepts heard and Spo2 on bipap 98%, Circulation- The pulse of 98beats/minute, Blood pressure was found to be 130/80 Disability- none, GCS 15/15. Extremity—WNL

Airway – patent and talking. Breathing – RR 15/m, Spo2-100%, bilateral normal vesicular breath sound. Circulation—BP 130/70, Pulse- 88/m, Patient responded well to bronchodilators, corticosteroid and on bipap. Cardiology Consultation was taken in view of patient's past history acute Lvf severe MR and dilated cardiomyopathy .Patient was admitted in CCU. She was investigated relevantly. Patient was started on iv diuretics infusion, iv antibiotics and other supportive treatment. Pulmonology consultation was taken for cough with expectorant, dyspnea and advice followed .Patient was advice HRCT chest ,sputum smear AFB stain, fungal smear/ KOH specimen, sputum for gram stain and other supportive care

Lab findings:

Sputum AFB: AFB negative

Sputum for gram stain: several gram positive cocci in pairs and short chain seen

Fungal/ KOH: no fungal elements seen. Anti – glycopeptidolipis IGA antibodies for Mycobacterium avium complex (unavailable at center)

Radiology finding

HRCT chest: evidence of bronchiectasis in lingular segment of left lung extending to involve in left lower lobe s/o variant lady Windermere syndrome. CT scan of a woman with Lady Windermere syndrome that shows localized bronchiectasis in the Lingular segment of left lung, involve left lower lobe.

DISCUSSION

MAC pulmonary infection presenting as an interstitial and/or nodular pattern instead of a cavitary pattern on chest radiograms is being increasingly described in elderly women who have no underlying lung disease and no smoking history

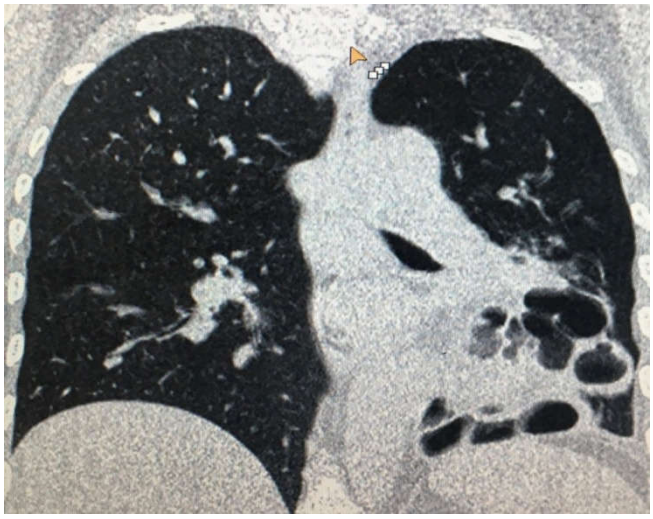


Figure 1.

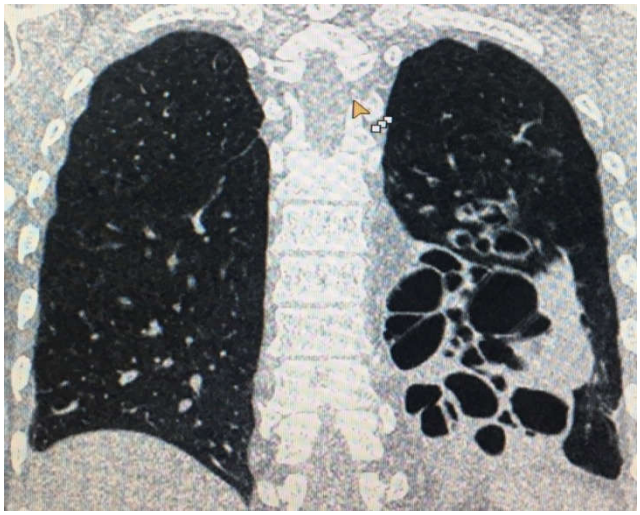


Image 2.

(Reich, 1992; Prince, 1989; Iseman, 1989). Reich and Johnson (Samjot Singh Dhillon, 2000) described 6 patients with MAC infection involving only the lingual or middle lobe of the right lung. Iseman *et al*. (1999) and Pomerantz *et al*. (1991) also described 12 women who had MAC infection of the middle lobe or lingual only. The middle lobe and lingual have in common long, narrow, and (in an upright position) dependent bronchi that predispose them to infection. Iseman *et al*. (5) and Pomerantz *et al*. (1991) described most of their patients as thin women for whom the incidence of skeletal abnormalities (including pectus excavatum, mild scoliosis, straight back, and mitral valve prolapse) was very high. On the basis of the frequent presence of a specific phenotype, thoracic abnormalities, and mitral valve prolapse, in addition to problems of collateral ventilation due to the frequent presence of complete or partially complete fissures, these researchers proposed that this syndrome might be a connective tissue disorder. They also believed that the thoracic skeletal abnormalities could result in decreased sputum clearance and ineffective cough mechanisms, also contributing to progression of the disease. Several studies have reported that most women with thoracic skeletal abnormalities and MAC pulmonary infection are thin (Samjot Singh Dhillon, 2000; Iseman, 1989; Huang, 1999). Reich and Johnson (Samjot Singh Dhillon, 2000) hypothesized that women are more likely to regard expectoration as socially unacceptable behavior and thus indulge in habitual voluntary cough suppression. This voluntary cough suppression leads to an inability to clear secretions (especially from the middle lobe and lingula), which results in a chronic nidus for inflammation that favors subsequent infection by MAC. Sufficient previously reported data on voluntary cough suppression are not available. However, it has been shown that this entity does exist and that cough induced by inhalation of capsaicin can be voluntarily suppressed (Pomerantz , 1996). Wells *et al*. (1993) described 4 young women with voluntary cough suppression; CT revealed bronchiectasis in 1 patient, which also developed during follow-up in another patient. Bronchoscope revealed copious secretions in the airways of 3 of these patients. Byrd *et al*. (1993) described 2 patients with middle lobe and lingular infiltrates due to MAC; these patients voluntarily suppressed their cough during physical examination. It is not clear whether they had a history of habitual cough suppression. Our patient was a retired schoolteacher and considered it impolite to cough in public. She had a lifelong history of suppressing her cough. She also had the typical phenotype described by Iseman *et al*. (1999) and Pomerantz *et al*. (1991), but there was no evidence of connective tissue disorder or mitral valve prolapse. It appears that the middle lobe and lingula are predisposed to nonspecific inflammation because of their particular anatomic structures and absence of collateral ventilation. Most of the patients with nonobstructive middle lobe syndrome due to MAC seem to be thin elderly women for whom the prevalence of abnormalities of the chest wall and thoracic spine is high. A diminished cough reflex due to the abnormal bony cage may further predispose the middle lobe and lingula to chronic inflammation and infection. The concept of voluntary cough suppression cannot be ignored. This entity occurs predominantly in women and may result in an inability to clear secretions from the middle lobe and lingula.

Our patient had a history of lifelong habitual cough suppression. To the best of our knowledge, we report the first case of bronchiectasis and middle lobe syndrome due to MAC in which a history of habitual voluntary cough suppression was elicited. Thus, she is the first patient with a bona fide diagnosis of Lady Windermere syndrome as defined by Reich and Johnson (Samjot Singh Dhillon, 2000). The exact prevalence of habitual cough suppression among such patients and, as a matter of fact, among all patients with no obstructive middle lobe syndrome is unknown and needs to be studied. It is possible that with appropriate behavior modification, this disease can be prevented.

REFERENCES

Byrd R, Payne JL, Roy TM. 1995. Lingular and middle lobe infiltrates in an elderly woman, Chest, vol. 108 (pg. 1156-7)

Byrd RP, Mirza S, Roy TM. 1995. Lady Windermere's syndrome: an atypical presentation of an atypical mycobacterium, *J Tenn Med Assoc.,* vol. 88 (pg. 303-5)

Huang JH, Kao PN, Adi V, Ruoss SJ. Mycobacterium avium-intracellulare pulmonary infection in HIV-negative patients without preexisting lung disease: diagnostic and management limitations, Chest, 1999, vol. 115 (pg. 1033-40).

Hutchings HA, Morris S, Eccles R, Jawad MSM. 1993. Voluntary suppression of cough induced by inhalation of capsaicin in healthy volunteers, *Respir Med*., vol. 87(pg. 379-82)

Iseman MD, Buschman DL, Ackerson LM. 1991. Pectus excavatum and scoliosis: thoracic anomalies associated with pulmonary disease caused by Mycobacterium avium complex, *Am Rev Respir Dis.*, vol. 144 (pg. 914-6)

Iseman MD. 1989. Mycobacterium avium complex and the normal host (editorial), *N Engl J Med.,* vol. 321 (pg. 896-8)

Pomerantz M, Denton JR, Huitt GA, Brown JM, Powell LA, Iseman MD. 1996. Resection of the right middle lobe and lingula for mycobacterial infection, *Ann Thorac Surg*., vol. 62 (pg. 990-3)

Prince DS, Peterson DD, Steiner RM. *et al*. 1989. Infection with Mycobacterium avium complex in patients without predisposing conditions, *N Engl J Med.,* vol. 321 (pg. 863 - 8)

Reich JM, Johnson RE. 1992. Mycobacterium avium complex pulmonary infection presenting as isolated lingular or middle lobe pattern: the lady Windermere Syndrome, Chest, vol. 101 (pg. 1605 - 9)

Samjot Singh Dhillon, Chatrchai Watanakunakorn, Lady Windermere Syndrome, 2000. Middle Lobe Bronchiectasis and Mycobacterium avium Complex Infection Due to Voluntary Cough Suppression, Clinical Infectious Diseases, Volume 30, Issue 3, March, Pages 572-575, doi: https://doi.org/10.1086/313726

Wells A, Rahman A, Woodhead M, *et al*., 1993. The clinical features of voluntary cough suppression associated with chronic pulmonary suppuration ("atussis nervosa"): a controlled evaluation (abstract), *Am Rev Respir Dis*, vol. 147 pg. A461

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