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Lady Windermere Syndrome; Rare Pulmonary Infection Secondary To Voluntary Suppression Of Cough.

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CLINICAL HISTORY: 65-year-old lady suffering from cough for the last one month with white colored scanty sputum. Past three weeks she has developed wheeze with shortness of breath. She also complains of on and off mid fever for last two weeks with evening raise of temperature. No evidence of chest pain or hemoptysis. No past history of pulmonary tuberculosis or allergic airway disease. Non diabetic and non-hypertensive. General physical examination was normal. On auscultation there was reduced air entry with crackles in the right mid and lower zones. Rest of the systems was normal.

IMAGES:



Figure no.1 Frontal chest Radiograph: coarse reticulations bilateral paracardiac regions with indistinct right cardiac border.

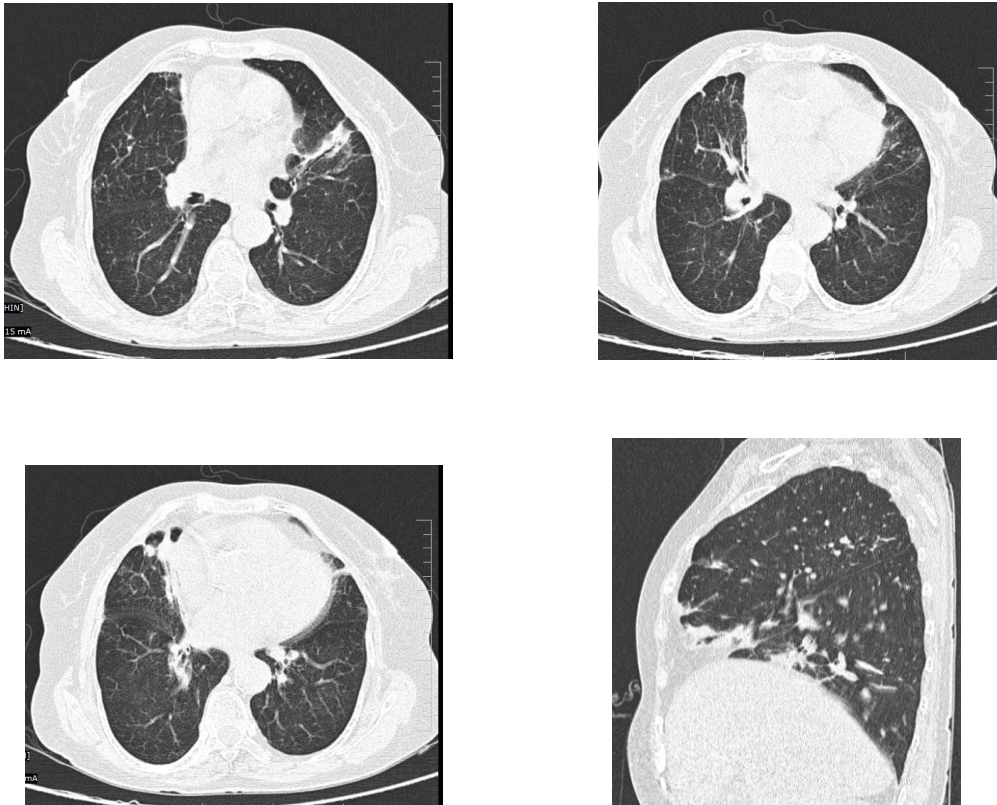


Figure no. 2 : Axial and sagittal reformatted CT scan of chest in lung window demonstrating atelectatic changes with bronchiectasis involving right middle and lingular lobes.

IMAGING FINDINGS:

Frontal radiograph depicted coarse reticulations in bilateral paracardiac regions with indistinct right cardiac border.

On HRCT there was cicatricial collapse of right middle lobe and lingular segments of left upper lobe with bronchiectatic changes. Few fibrotic bands are noted in the same segments. Focus of consolidation noted in the superior segment of right lower lobe. Subcentimetric mediastinal lymph nodes are noted.

DISCUSSION:

Introduction: Lady Windermere syndrome is eponymic name for Mycobacterium Avium Complex (MAC) pulmonary infection predominantly affecting right middle and left lingular lobes hypothesized secondary to voluntary suppression of cough¹. Lady Windermere in Oscar Wilde's play is a young married woman who never coughs or shows any signs of illness in public; for Victorian women, it is considered impolite to cough in public¹. The possibility of getting a lung infection may have increased as a result.

Epidemiology and Pathophysiology:

Lady Windermere syndrome has predilection for old age female patients who has tendency of voluntary suppression of cough.

The cause for infection could be multifactorial according to the various literatures. Narrow , long and dependent positions of right middle lobe bronchus and left lingular bronchus and poor collateral ventilation predisposes these lobes for chronic MAC infection. Some writers have also argued that thoracic skeletal deformities may render sputum clearance inefficient, resulting in persistent infection². According to Reich and Jhonson hypothesis, women are more prone to engage in habitual voluntary cough suppression because they view expectoration as socially unacceptable behaviour³.

Imaging features:

- a. **Plain radiograph:** Volume loss in middle and lingular lobes with coarse reticulations indicating bronchiectasis. Mucosal impactions may be additional findings.
- b. **CT:** MAC infection may manifest either as classic form or non-classic form. In classic form affected individuals have upper lobe fibro-cavitary lesions. The non-classic form manifest with features of Lady Windermere syndrome.

In Lady Windermere syndrome there is bronchiectasis involving right middle and left lingular lobe with volume loss. Solid or semisolid ground glass nodules with or without tree in bud configuration. No obvious obstructing lesion to the bronchi, making it one of the causes for right middle lobe syndrome.

Differential diagnosis:

The differentials could be other causes of middle lobe bronchiectasis like primary ciliary dyskinesia and acute respiratory distress syndrome. Both can be differentiated with their respective clinical symptomatology and other associated findings.

Teaching points: Lady Windermere syndrome is suspected in elderly women with bronchiectasis in the middle lobe and lingular distributions.

References :

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2. Pomerantz M , Denton JR , Huitt GA , Brown JM , Powell LA , Iseman MD. Resection of the right middle lobe and lingula for mycobacterial infection , *Ann Thorac Surg* 1996 vol. 62, 990-993.
3. Reich JM , Johnson RE. *Mycobacterium avium* complex pulmonary infection presenting as isolated lingular or middle lobe pattern: the lady Windermere Syndrome. *Chest* , 1992 , vol.101 (1605-1609).