**CLINICAL VIGNETTE**

# The Elusive Middle Lobe Syndrome

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***Case Summary***

A 67-year-old man with a history of chronic obstructive pulmonary disease (COPD) and obstructive sleep apnea was admitted to the medicine service with complaints of a productive cough and slightly increased shortness of breath compared to his baseline. He noted exertional dyspnea only with strenuous activity, as well as worsening of symptoms with cigarette use and no improvement with albuterol. Physical exam was notable for diminished breath sounds between the right anterior 4$^{th}$ to 6$^{th}$ rib. Initial infectious work up including sputum gram stain, culture, quantiferon gold and fungal serologies were negative. Evaluation for cardiac causes was negative. Chest radiograph showed right middle lobe collapse (Figure 1 and 2). CT of the chest showed complete right middle lobe atelectasis with multiple bilateral non-calcified pulmonary nodules (Figure 3). After specimens obtain from bronchoscopy were negative for infection and malignancy, the patient was diagnosed with Middle Lobe Syndrome (MLS).

***Discussion***

MLS was initially described by Graham in 1948 as recurrent or chronic collapse of the right middle lobe. [1] The process can also involve the lingula. There is limited literature on MLS since it is relatively rare, due to varied etiologies and presentations, and lacks a consistent definition. The epidemiology is not well described but it is known to occur in children and adults and is more common in females. [2]

Pulmonary symptoms are common but some patients are asymptomatic and identified with chest imaging alone. Presenting symptoms include chronic or recurrent cough, dyspnea, chest pain or symptoms from obstructing pneumonia.[2,3] Patients may also present with recurrent pneumonia or pneumonitis. A family history of atopy is common and up to half of patients report atopy, asthma or COPD. Chest radiographs may be normal in patients with intermittent collapse. On posterior-anterior chest film, the right heart border is obscured by a triangular opacity. The lateral film more clearly delineates an opacity where the horizontal and oblique fissures are brought close together. (see Figure 1 and 2) CT generally shows a triangular opacity with the apex pointing toward the hilum. (see Figure 3)

The etiologies are divided into obstructive and non-obstructive types.[1] Obstruction is identified on bronchoscopy or imaging and can be caused by endobronchial lesions (tumor, mucus plug, or foreign body) or extrinsic compression due to lymphadenopathy. Tumors can be either benign or malignant and are identified in about 25% of cases. Lymphadenopathy can be due to granulomatous infections such as histoplasmosis, other fungal or atypical mycobacterium (i.e. Lady Windermere syndrome).

Non-obstructive MLS is diagnosed when there is no obstruction identified on bronchoscopy. The unique anatomy of the right middle lobe, which is bordered by two fissures and has minimal collateral ventilation, is more prone to collapse and reduces the chance of reinflation once collapse occurs. Non-obstructive MLS can occur in adults and children with recurrent pneumonia, asthma, bronchitis or cystic fibrosis.[2,4] (see table 1)

Initial evaluation depends on the suspected underlying cause and can include a general infectious work-up including sputum gram stain and culture. If initial diagnostic evaluation is unrevealing, specific infectious processes should be considered (i.e. fungal pathogens and mycobacterium). Flexible bronchoscopy is important to evaluate the patency of the bronchus, collect specimens and exclude malignancy or other surgically correctible lesions. Endobronchial ultrasound (EBUS), can detect extrabronchial lesions. If no cause is found with bronchoscopy, additional imaging (high resolution CT or PET-CT) should be done to exclude neoplasm. [2]

Treatment depends on the underlying cause. In non-obstructive cases, medical management including antibiotics, bronchodilators, mucolytics with chest physiotherapy and postural drainage can be effective. There have been case reports of disease resolution after low dose macrolide administration.[5] In obstructive cases that cannot be managed by bronchoscopy alone, a surgical option can be explored, but there is currently no consensus on surgical indications. Lobectomy is considered if conservative management fails or in the setting of malignancy, bronchiectasis, or chronic infection that poses a substantial health risk to the patient. [2]

***Clinical Course***

This patient was initially treated with pneumatic physiotherapy for two days in conjunction with albuterol and acetylcysteine without improvement in symptoms or resolution on imaging. After bronchoscopy, complete extrinsic compression was suspected and no endobronchial lesions were seen at the point of obstruction. A trial of balloon dilation was attempted without any improvement. Transbronchial aspiration of a subcarinal lymph node, and right middle lobe bronchial nodule biopsies

were negative for malignant cells and cultures were negative. Repeat CT chest did not show any extrabronchial lesions. PET-CT did not identify abnormal uptake making neoplasm or infection unlikely. Thoracic surgery was consulted for possible lobectomy since his symptoms were bothersome and caused anxiety. The surgical team recommended observation and planned outpatient follow-up.

***Summary***

MLS has characteristic imaging findings but remains an elusive diagnosis due to a multitude of potential etiologies. Its causes are typically benign and due to infection and inflammation, though malignancy must be considered. Treatment includes conservative management for benign causes. Surgical management is considered for patients with malignancy, recurrent infections, or failure to respond to conservative management.

***Figures***

**Figure 1:** Posterior-anterior X-ray shows a patchy opacity obscuring the right heart border.


**Figure 2:** Lateral X-ray show right middle lobe collapse.

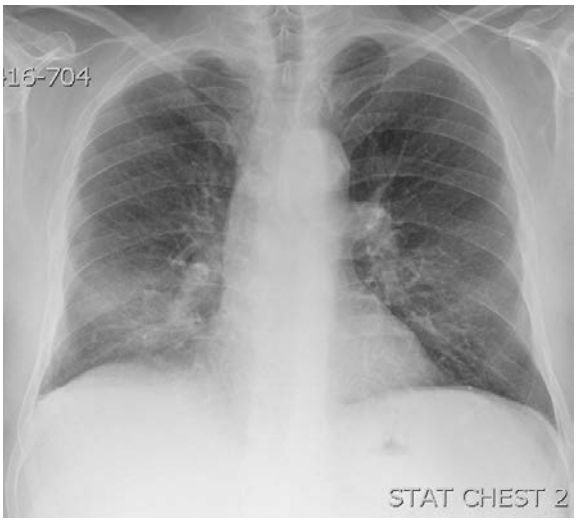


**Figure 3:** CT Thorax with Contrast.

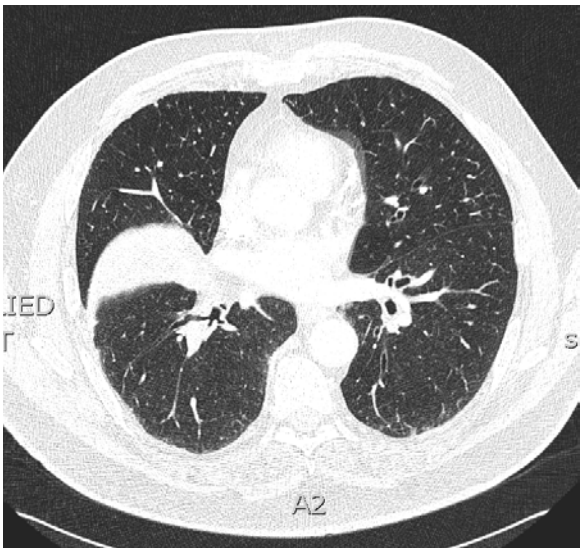


***Table 1***

| Obstructive | Non-obstructive |
|---|---|
| **Endobronchial**<br>Mucous plugging<br>Endobronchial tumor<br>- Malignant: primary lung cancer or metastasis<br>- Benign: hamartoma<br>Foreign body: common in children<br>Stenosis<br>Broncholith<br>**Extrabronchial:**<br>Lymphadenopathy<br>- Granulomatous infections<br>Primary tumor or metastasis<br>Sarcoidosis<br>Cardiovascular abnormalities | Recurrent pneumonia<br>Asthma<br>Bronchiectasis<br>Cystic fibrosis<br>Primary ciliary dyskinesia |

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