Middle Lobe Syndrome

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Abstract

Middle Lobe Syndrome (MLS) is a rare condition with significant clinical implications. MLS is characterized by a recurrent or chronic collapse of the right middle lobe of the right lung. MLS is more common in women, with a ratio ranging from 1.5 to 3. MLS is divided into two types: obstructive MLS, caused by enlarged peribronchial lymph nodes, and non-obstructive MLS, which is frequently associated with airway inflammation. Clinical symptoms of MLS include chest pain, chronic cough, excessive sputum, and dyspnea. Symptomatic medication such as mucolytics and bronchodilators, chest physiotherapy, fiberoptic bronchoscopy, and surgery are commonly used as a management of MLS.

Keywords: Medial Lobe Syndrome, Obstructive, Chronic Cough

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Introduction

Middle Lobe Syndrome (MLS) is a rare disease but has a great clinical importance. MLS is characterized by a recurrent or chronic collapse of the right middle lobe of the right lung. Although initially MLS occurs only in the middle lobe, the course of the disease can involve the lingula as well, which is often called lingula syndrome.[1]

MLS is more common in women, with a female-to-male ratio ranging from 1.5 to 3 in most studies. A family history of atopy is also common in MLS patients, with half reporting a history of atopy, asthma, or chronic obstructive pulmonary disease.[1]

To date, epidemiological data on MLS is still very limited. No studies clearly describe the incidence and mortality rates of MLS. Therefore, the epidemiological data of MLS only refers to existing retrospective studies. A Japanese one-year retrospective study reported that 0.17% (51 out of 30,588) of patients undergoing chest x-ray screening had MLS. Of these 51 people, age > 50 years, and female gender had a higher incidence of MLS.[2] Another study in Turkey involving asthmatic children reported an incidence of 1.62% (56 out of 3528), of which more than half were less than 6 years old.[3] A 20-year study of 157 obstructive and non-obstructive MLS patients undergoing surgery in Tunisia reported a mean age of 33.18 +/- 19.85 years.[4] In a nationwide study from Iceland on MLS patients requiring surgical resection, the incidence was 1.43 per million men per year and 2.94 per million women per year.[5] In Indonesia, there is no clear report on the incidence of MLS.

Etiology

Middle Lobe Syndrome is a long-term atelectasis that occurs when the right lung's middle lobe shrinks. Pressure on the bronchial airway caused by a tumor or enlarged lymph nodes is the most common cause of this condition. Congested and obstructed lungs can lead to chronic inflammation, scarring, bronchiectasis, and non-resolving pneumonia.[6]

Classification and Pathophysiology

There are two types of Middle Lobe Syndrome: obstructive and non-obstructive. An endobronchial lesion or extrinsic compression of the right middle lobe bronchus can cause the obstructive type. Tumors, whether benign or malignant, cause one-fourth of cases; however, many MLS cases occur in non-neoplastic patients. primary and metastatic lung cancer is the most common cause of obstruction in the middle airway. Enlargement of the peribronchial lymph nodes, caused by a granulomatous infection such as histoplasmosis, is another common cause of external compression of the right middle lobe bronchus. Other causes include sarcoidosis-related adenopathy or lymph node

enlargement caused by metastases and foreign bodies entering the bronchi.[7]

Non-ventilation of the distal airways is caused by bronchial obstruction, where gases are completely absorbed by pulmonary blood flowing through that area. Compression of the parenchyma and/or increased surface tension cause gas extrusion from the alveoli, reducing the ability of the involved parenchyma to re-inflate. Atelectasis ended up causing alveolar hypoxia and pulmonary vasoconstriction while causing minimal arterial hypoxia. If blood cannot be diverted, airflow disturbance by intrapulmonary shunting will occur.[8]

Non-obstructive MLS occurs in a patient with an intact right middle lobe bronchus. This type of MLS is not specific to the middle lobe; it can also be present in the lingula of the left upper lobe. The majority of individuals with MLS have the nonobstructive variant, which typically occurs in adults and children with recurrent pneumonia and is frequently associated with asthma, bronchitis, and cystic fibrosis.[7]

Diabetes, immunosuppression, bronchiectasis, and known alterations in right middle lobe anatomy, along with age-induced airway remodelings such as decreased lung and thoracic compliance, diminished respiratory muscle strength, and impaired mucociliary clearance, were thought to have caused pneumonia recurrence later.[9] When inflammation and/or edema are caused by lower respiratory tract infections, these variables may interfere with or impede the drainage of mucous secretions from the middle lobes. Mucus hypersecretion is the primary characteristic in some patients, which worsens their illness. This is a regular occurrence in children with asthma, cystic fibrosis, and ciliary dyskinesia. Airway inflammation and airway epithelial involvement in such patients can both impact peri-ciliary fluid outflow, lowering the action of surfactants and raising the likelihood of bronchial collapse. [8]

Table 1. Presenting symptoms of the MLS patients.[9]

	n	%
Cough	37	100
Sputum	35	94.6
Chest Pains	8	21.62
Dyspnea	6	16.22
Lung Infection	11	29.73
Fever	11	29.73
Hemoptysis	2	5.42

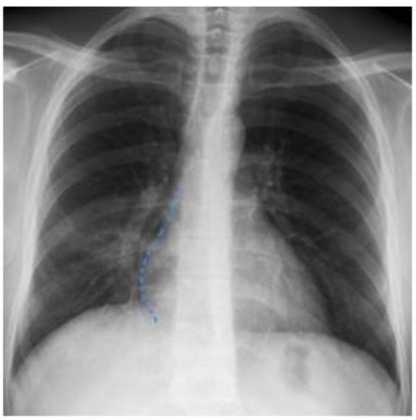
Clinical Manifestation

Patients with MLS present the general findings of chest pain, chronic cough, copious sputum, and dyspnea on clinical examination. Chronic productive cough, chest pain, and shortness of breath are symptoms that mimic bronchiectasis. The study showed chronic lung infection in 51.35% of patients, pneumonia in 21.62%, and tuberculosis sequelae in 18.92% of cases.[9] A detailed description of the percentage of clinical symptoms in MLS can be seen in table 1.

Diagnostic method

The majority of Middle Lobe Syndrome cases are diagnosed radiologically. Conventional chest radiographs from posteroanterior and lateral positions are useful for demonstrating atelectasis with increased density and sharpness of the border extending posteriorly. In the diagnosis of MLS, a simple plain chest X-ray is equally significant as computed tomography (CT). Bronchoscopy can also be used to diagnose the site of abnormality, collect the sample of microorganisms and treat MLS.[9]

A.



B.

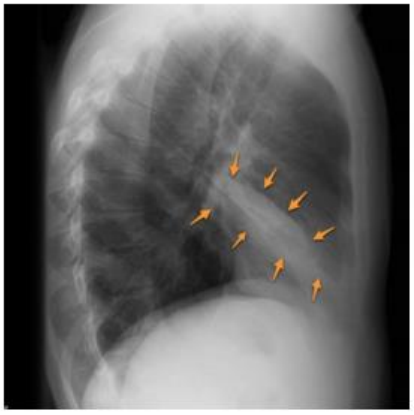
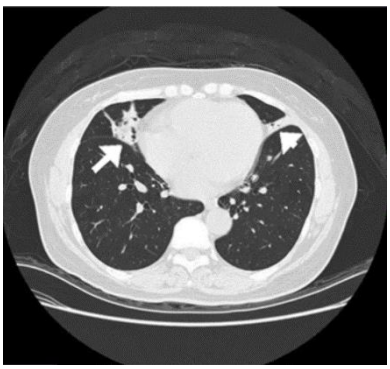


Figure 1. Plain chest x-ray of Middle Lobe Syndrome.[13] A. Posteroanterior view, B. Right lateral view.

Chest computed tomography (CT) scan with a thin section and high resolution will be helpful in identifying endobronchial or parenchymal abnormalities. This examination will clearly show bronchial patency due to calcification and compression from extrinsic masses. This



approach can also clear the right heart's border from the lung.[7] CT scan in MLS cases could also showed atelectasis and bronchiectasis in the middle lobe of the lung.[13,14]

Figure 2. *CT-Scan of Middle Lobe Syndrome*[13]

Management

The primary objective of dealing with middle lobe syndrome is to address the underlying cause of the problem. Infections that cause lung parenchymal damage should be treated appropriately based on the results of microbial cultures. To minimize the symptoms caused by the condition, symptomatic medication such as mucolytics and bronchodilators may be administered.[10]

Non-pharmacological treatment with chest physiotherapy can help to reduce the shortness of breath and is beneficially applied for all types of MLS. Fiberoptic bronchoscopy will help to identify the obstruction in the middle airway or any intrabronchial tumors in the obstructive type of middle lobe syndrome, and surgical intervention may play an important role if certain managements fail to provide favorable outcomes.[10]

Summary

Middle lobe syndrome is a long-term atelectasis occurs in the middle lobe of the right lung or lingula in the left lung. The cause of the disease is classified into obstructive and non-obstructive types. Patients often present the symptoms of shortness of breath, chest pain, chronic cough and mucoid sputum. Radiological diagnostic along with fiberoptic bronchoscopy could help to confirm the primary cause as well as to provide the treatment of MLS.

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