

Case report**Right Middle Lobe Syndrome**

Jawed Abu Bakar Warind *

Abstract:

Middle Lobe Syndrome is known to be caused by numerous etiologies with no consistent clinical picture. In this case report, we explain the importance of bronchoscopy with biopsy in identifying the exact etiology of middle lobe syndrome and to emphasize that pulmonary tuberculosis can present with atypical presentation.

Consultant Pulmonologist Liaqat National Hospital, Karachi

Introduction:

Middle Lobe Syndrome refers to atelectasis in the Right Middle Lobe of the lung and is normally suggested radiologically by Right cardiac "silhouette" sign. The term was coined by Graham & co-workers who reported 12 cases of middle lobe atelectasis of non-tuberculous origin secondary to bronchial compression by enlarged lymph nodes¹. The patients in their series presented with haemoptysis, chronic cough and recurrent pulmonary infections and were treated by lobectomy. The definition of middle lobe syndrome was subsequently modified to include all types of right middle lobe atelectasis, even when the bronchial compression was absent.

Middle lobe syndrome is divided into an *obstructive type*, with a demonstrable airway occlusion, and a *non-obstructive type*, with a patent right middle lobe bronchus apparent on bronchoscopy. The reported etiologies of RMLS in one series include inflammation (47%), malignant tumors (22%), bronchiectasis (15%), tuberculosis (9%), benign tumors (2%), and aspiration (2%)^{1,2}.

The clinical presentation of Middle Lobe syndrome is widely inconsistent and pulmonary tuberculosis can present in atypical way.

Case Report:

A 65 years old woman, with type 2 diabetes mellitus and hypertension for last 20 years, resident of Khanewal, admitted through OPD with complaints of; undocumented weight loss and anorexia for one and a half year, productive cough for 5 months, fever for 5 months and haemoptysis {(3 episodes) and last episode three days before presentation}. Patient and the family denied any history of contact with known tuberculous patient. Patient denied any history of smoking or any addiction to beetle nuts or tobacco.

Past medical history was significant for recurrent chest infections and multiple antibiotics use over the last one and a half year.

On physical examination, she was an elderly woman, of average height and built, lying comfortably in bed, not in acute distress, oriented in time, place and person. The vitals were;

Temp: 98.7°F

Pulse: 74/min, regular, normal volume

BP: 110/70

RR: 14/min

Chest examination demonstrated bilaterally symmetrical expansion with normal vesicular breathing and a central trachea. Apex beat was localized at the 5th intercostal space in the midclavicular line. Tactile vocal fremitus and Vocal resonance were normal. Percussion was resonant all over. The only abnormality was in auscultation, which revealed vesicular breathing with expiratory wheeze heard over the right nipple area.

Investigations:

Hb: 10.2g/dl (MCV: 72.60, MCHC: 30.50),

TLC: 6.3 (N: 65%, L: 25%, M: 8%)

Platelets: 376,

PT: 12/15, APTT: 24/30,

ESR: 40

Urea: 22mg/dl,

Creatinine: 0.73mg/dl

Chloride: 102meq/l

Sodium: 142meq/l

Potassium: 3.8meq/l

Bicarbonate: 29meq/l

Mg: 1.87,

Phosphorus: 2.67,

Calcium: 8.3mg/dl

Albumin: 4.16

ECG= normal sinus rhythm

Discussion:

This woman presented with haemoptysis, fever and weight loss associated with productive cough. For presumed recurrent pneumonias, she received multiple antibiotic therapies in the past. One has to enquire about the cause of recurrent pneumonias. Her chest radiograph demonstrated atelectasis of right middle lobe, which was further confirmed on a CT scan.

Without any evidence of main airway obstruction or significant adenopathy on CT, a diagnosis of MIDDLE LOBE SYNDROME is high on the list, and will need to be evaluated further to identify the etiology leading to this presentation.

Middle lobe syndrome (MLS) is a rare lung disorder concerning the right middle lobe and/or lingula and is characterized by a spectrum of clinical and pathological lesions ranging from recurrent atelectasis or pneumonias to bronchiectasis⁵.

The original view that bronchial compression was the pathophysiological abnormality leading to development of the syndrome has been rejected and the focus has now turned to the relative isolation of the middle lobe, especially when a complete minor fissure is present. This isolation prevents the aerating effects of collateral ventilation of the upper lobe from reaching the middle lobe and thus impairs the clearing of secretions from the middle lobe bronchus^{2,3}. Anatomic characteristics, such as the narrow diameter of the lobar bronchus and an acute takeoff angle, make the RML susceptible to transient, usually partial, obstruction. Such obstruction is considered to be the result of the poor drainage of secretions due to inflammation and/or edema of the RML bronchus. In addition, the relative anatomic isolation of the middle lobe and the poor collateral ventilation diminish the chance of re-expansion once atelectasis has been established. These mechanisms help to explain the vicious cycle of recurrent inflammation and obstruction that develops after repeated episodes of infection or asthma exacerbations^{4,5,9&10}.

Although patient is a non smoker without other identifiable risks for lung cancer. We would like to evaluate the etiology i.e. obstructive versus non obstructive and will need a bronchoscopy with biopsy.

The bronchoscopy demonstrated a patent but narrowed right middle lobe bronchus with significant swelling (fig 1). Endobronchial biopsy demonstrated Granuloma with giant cell and necrosis, suggestive of Tuberculosis (fig 2).

Although middle lobe syndrome is rare due to pulmonary tuberculosis but it is not uncommon⁵, and should be considered in areas with high tuberculosis burden. The usual course of middle lobe syndrome is recurrent symptoms and repeated antibiotics for recurrent bacterial infections ultimately requiring surgery. It is advisable to check for treatable causes before going to ultimate therapy.

Thus, if atelectasis persists after an adequate medical therapy, resection of the lobe is indicated⁶⁻⁸. Priftis et al in their study showed that timely medical intervention in patients with MLS that includes FOB with BAL prevents bronchiectasis that may be responsible for an ultimately unfavourable outcome⁵. In our patient the delay in the diagnosis was probably because of the fact that sputum results were negative, she was not worked up until she developed more drastic symptom i.e. haemoptysis; which is not unusual in a resource limited environment and atypical location of the disease.

Conclusions:

- A right sided cardiac Silhouette suggests disease in the middle lobe.
- Middle lobe syndrome needs bronchoscopic evaluation.
- Pulmonary tuberculosis can present with atypical presentation.
- Pulmonary tuberculosis can present with mildly elevated or even with normal ESR and negative sputum results
- Malignancy is not the rare cause of middle lobe syndrome

References:

1. Lindskog GE, Spear HC: Middle-lobe syndrome. *N Engl J Med* 1955; 253:489-495.
2. Bertelsen S, Struve-Christensen E, Aasted A, Sparup J. Isolated middle lobe atelectasis: aetiology, pathogenesis, and treatment of the so-called middle lobe syndrome. *Thorax*. 1980 Jun; 35(6):449-52.
3. Wagner RB, Johnston MR. Middle lobe syndrome. *Ann Thorac Surg*. 1983 Jun; 35(6):679-86. Review.
4. Prem Parkash Gupta, K.B. Gupta and Dipti Agarwal *Indian J Tuberc* 2006; 53:104-108.
5. Priftis KN, Mermiri D, Papadopoulou A, Anthracopoulos MB, Vaos G, Nicolaidou P. The role of timely intervention in middle lobe syndrome in children. *Chest*. 2005 Oct; 128(4):2504-10.
6. Kwon KY, Myers JL, Swensen SJ, et al. Middle lobe syndrome. A clinicopathological study of 21 patients. *Hum Pathol*. 1995; 26:302-307.
7. Middle lobe syndrome. *Cir Pediatr*. 1994 Apr; 7(2):105-7. Spanish
8. Ring-Mrozik E, Hecker WC, Nerlich A, Krandick G. Clinical findings in middle lobe syndrome and other processes of pulmonary shrinkage in children (atelectasis syndrome). *Eur J Pediatr Surg*. 1991 Oct; 1(5):266-72.
9. Sekerel BE, Nakipoqlu F. Middle lobe syndrome in children with asthma: review of 56 cases. *J Asthma*. 2004 Jun; 41(4):411-7.
10. Ayed AK. Resection of the right middle lobe and lingula in children for middle lobe/lingula syndrome. *Chest*. 2004 Jan; 125(1):38-42.