

Middle lobe syndrome: a retrospective analysis

Orta lob sendromu: Retrospektif bir inceleme

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Background: This study aims to evaluate the efficacy middle lobe syndrome (MLS) surgery in patients with recurrent pulmonary infections.

Methods: We retrospectively reviewed 37 cases (19 females, 18 males; mean age 12.3 years; range 6 to 55 years) who underwent surgery due to MLS in Dicle University, Faculty of Medicine, Department of Thoracic Surgery between January 1994 and January 2011. The underlying etiology, symptoms of the disease, bronchoscopic and radiological findings, and the outcome of surgery were evaluated.

Results: Coughing was the most common symptom. The mean duration of symptoms was 5.49 years (range, 6 months to 15 years). Indication for surgery was bronchiectasis in 33 cases, collapsed lungs in two case, and total atelectasis in two cases. No postoperative mortality was seen. The mean length of hospital stay was 7.14 days (range, 5 to 13 days). The mean follow-up was four years (range, 1 to 15 years). Infection, cough and mucus were absent after surgery.

Conclusion: In MLS, lobectomy is indicated for unresolving pulmonary infections and bronchial stenosis. Patients become asymptomatic following surgery.

Key words: Bronchiectasis; middle lobe syndrome; pneumonia.

Amaç: Bu çalışmada tekrarlayan pulmoner enfeksiyonlu hastalarda orta lob sendromu (OLS) cerrahisinin etkinliği değerlendirildi.

Çalışma planı: Ocak 1994 ve Ocak 2011 tarihleri arasında Dicle Üniversitesi Tıp Fakültesi, Göğüs Cerrahisi Anabilim Dalında OLS nedeniyle ameliyat edilen 37 hasta (19 kadın, 18 erkek; ort. yaş 12.3 yıl; dağılım 6-55 yıl) retrospektif olarak incelendi. Altta yatan etyoloji, hastalık semptomları, bronkoskopik ve radyolojik bulgular ve cerrahinin sonucu değerlendirildi.

Bulgular: Öksürük en sık görülen semptom idi. Semptomların ortalama süresi, 5.49 yıl idi (dağılım, 6 ay-15 yıl). Otuz üç hastada bronşektazi, iki hastada akciğer çökmesi ve iki hastada total ateletazi cerrahi endikasyonu idi. Ameliyat sonrası mortalite gözlenmedi. Ortalama hastanede kalış süresi, 7.14 gün idi (dağılım, 5-13 gün). Ortalama takip süresi, 4 yıl idi (dağılım, 1-15 yıl). Cerrahi sonrasında enfeksiyon, öksürük ve mukus gözlenmedi.

Sonuç: Orta lob sendromunda iyileşmeyen pulmoner enfeksiyonlarda ve bronşiyal stenozda lobektomi endikedir. Cerrahi sonrasında hastalar asemptomatik hale gelir.

Anahtar sözcükler: Bronşektazi; orta lob sendromu; pnömoni.

Isolated atelectasis of the middle lung lobe is known as middle lobe syndrome (MLS), and this can even extend its affects to the lingula, even though it is not located on the left side. This syndrome eventually appears when the middle lobe bronchus receives pressure from external lymph node enlargement, resulting in chronic recurrent pneumonia and postobstructive atelectasis. This process is more common in the middle lobe bronchus than elsewhere in the lungs because of its

narrowness and length. The absence of collateral ventilation in patients with complete fissures makes this process easy. Recurrent infections and middle lobe atelectasis are symptoms that precede MLS, and this can be seen with or without hilar lymph enlargement on chest X-rays. In these situations, endobronchial obstruction by slow-growing malignant neoplasms should be kept in mind, and attention must be paid to cases which yield the same view.^[1] Additionally,



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MLS is a chronic and suppurative disease, which can lead to much patient discomfort. In this report, we present 37 patients who were diagnosed with MLS after undergoing surgery for resection and compare the results with the related literature.

PATIENTS AND METHODS

In this study, we retrospectively reviewed the hospital data of 37 patients (19 females, 18 males; mean age 12.3 years; range 6 to 55 years) who were diagnosed with MLS between January 1994 and January 2011. Patients who did not undergo surgery and those with further pathologies in other areas in the middle lobe were excluded. We obtained the patient information related to age, gender, symptoms, etiology, morbidity, and mortality and also evaluated the data regarding postoperative hospital stay and duration of symptoms. In addition, we also took note of when each patient began having complaints. The results of chest X-rays, computed tomography (CT), high-resolution CT, and bronchoscopies were used for diagnosis (HRCT). We also investigated the presence of acid-fast, resistant bacilli in seven cases with tuberculosis (TB) sequelae. In addition, all of our patients had previously undergone a rigid bronchoscopy.

RESULTS

This retrospective study included 19 patients younger than 16 years of age and 18 patients older than 18 years of age. The most common symptoms were cough and sputum in all cases (Table 1), and inspiratory and expiratory rales over the middle lobe were present on auscultation in the majority of the patients. Middle lobe atelectasis or pneumonia, indicating an increase in density at the right border of the heart and the bronchovascular region, was seen on the chest X-rays of all of our subjects, and a narrowing of the middle lobe bronchus was identified on CT/HRCT (Figures 1a-c). Chronic pulmonary infections were the etiological source of the MLS in 51.35% of the cases (Table 2), and the symptomatic duration was 5.49 years (range 6 months-15 years). Furthermore, surgery was indicated by bronchiectasis in 33 patients, a destroyed lung in two patients, and total atelectasis in two others. A rigid bronchoscopy and bronchoalveolar lavage (BAL) were also performed on all of the patients (Table 3). Bacterial cultures were positive for β -hemolytic streptococci in seven patients as well as one patient with *Haemophilus influenza* on the BAL fluid (BALF) aspirate. The test for acid-fast, resistant bacteria was negative for all seven patients, and TB did not grow on any culture. In addition to postural drainage and physiotherapy, bronchodilator

and mucolytic therapy were prescribed along with antibiotics when the cultures were positive.

A right middle lobectomy was performed via a right thoracotomy on all of the patients. Enlarged lymph nodes around the middle lobe bronchus were present in 25 patients, and the thoracotomy revealed that they seemed to be depressing the bronchus in 12 of these patients. Furthermore, tissue stiffness was observed in 12 patients around the middle lobe bronchus, which was secondary to infections. A pathological study of the specimens revealed bronchiectasis in 32 patients, a destroyed lung in three patients, and organized pneumonia in two others. Postoperative complications included atelectasis in five patients and prolonged air leaks in four. Furthermore, pneumothorax failed to expand in one patient, and bleeding requiring a re-thoracotomy occurred in another. The mean length of hospital stay was 7.14 days (range 5-13 days). Analgesic and mucolytic treatment to prevent atelectasis due to secretion stasis as well as limited cough reflex caused by thoracotomy pain continued for up to one month. The cases were followed up for approximately four years (mean 1-15 years), and infection, cough, and sputum were not present at that time.

DISCUSSION

In 1948, Graham et al.,^[2] in a study composed of 12 patients, first defined MLS as right middle lobe bronchus compression secondary to enlarged lymph nodes.^[3] Since then, numerous studies have been undertaken to determine the etiology, pathophysiology, and management of this disease, and atelectasis has been easily identified as occurring more in the right middle lobe than in the other lobes.

The causes of MLS are the abnormal length and narrowness of the bronchus in the right middle lobe prior to segmental division. In addition, there is evidence that points to the bronchus being enclosed by lymph nodes. Collateral ventilation is also inadequate, which is another important factor. Furthermore,

Table 1. Presenting symptoms of the patients

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

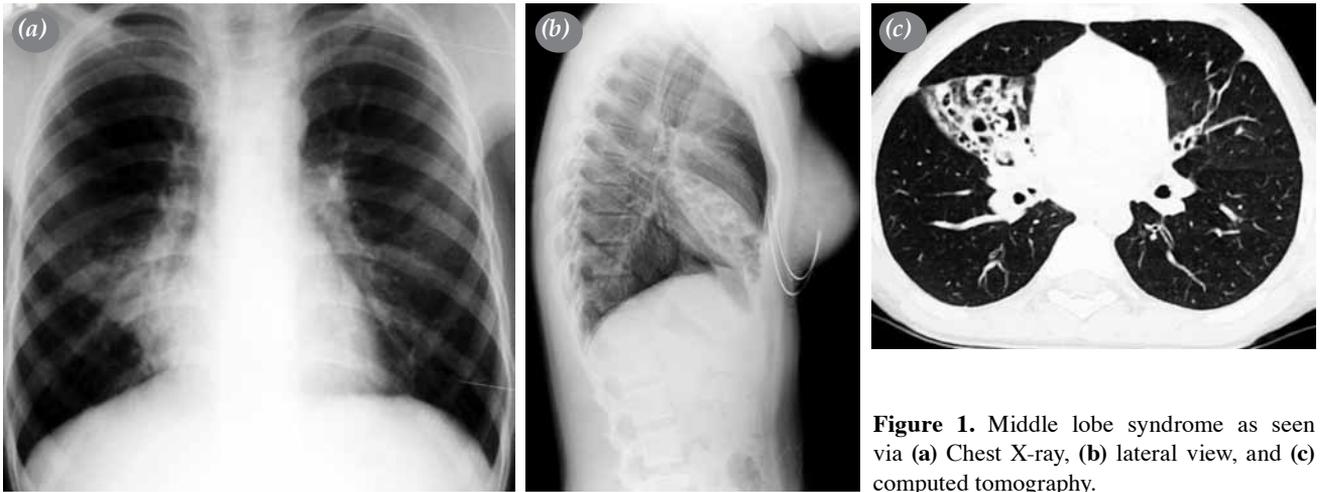


Figure 1. Middle lobe syndrome as seen via (a) Chest X-ray, (b) lateral view, and (c) computed tomography.

incomplete fissures around the right middle lobe prevent the rapid resolution of inflammation. Thus, the absence of collateral ventilation in patients with complete fissures reduces the risk of a more serious condition.

Obstructive or nonobstructive pathologies may be present in MLS. Nonobstructive causes include primary airway inflammation or impaired collateral ventilation. Additionally, diseases such as asthma, bronchopulmonary dysplasia, cystic fibrosis, and chronic bronchitis can also be responsible.

Obstructive causes may be intraluminal in nature with the occurrence of lymph nodes, tumors, and abnormal branching. However, they can also be extraluminal, as evidenced by foreign body aspiration, granulation tissue, and mucus plugs.^[4,5] In our research, most of the reported studies identified nonobstructive causes as being more common. In MLS, the symptoms are known to begin in early adolescence and start to subside with age if the etiology is nonobstructive. Moreover, both male and female predominance has been reported in the literature.

The etiology of MLS was determined to be inflammation in 47% of our cases, malignant tumors in 22%, bronchiectasis in 15%, TB in 9%, benign tumors in 2%, and aspiration in another 2% of the

patients. These results are consistent with those seen in a developing country. Other rare causes were amyloidosis, sarcoidosis, histoplasmosis, psittacosis, pertussis, cystic fibrosis, esophageal perforation, and bronchopulmonary aspergillosis.^[6] In addition, compression of the bronchus due to enlarged lymph nodes around the middle lobe bronchus was found in 25 patients intraoperatively, and tight tissue adhesion around the middle bronchus secondary to infections was found in 12 others in our series.

Patients with MLS demonstrate the common findings of chest pain, chronic cough, excessive sputum, and dyspnea on clinical examination.^[1,7] Chronic productive cough, chest pain, and shortness of breath are symptomatic of bronchiectasis. Our series revealed chronic pulmonary infection in 51.35% of the patients, pneumonia in 21.62%, and TB sequelae in 18.92% of the cases (n=7).

The etiology in 8.11% of the cases (n=3) was obscure. The literature has reported the most common cause of MLS as being benign inflammatory diseases, such as bronchiectasis and pneumonia.^[8,9] Bronchiectasis remains a serious problem in developing countries; however, it has been significantly reduced in developed countries. The etiology of this disease in developing countries is predominantly caused by bacterial, viral, and fungal infections, whereas immunodeficiency

Table 2. Probable etiologies of bronchiectasis

	n	%
Lung infections	19	51.35
Pneumonia	8	21.62
Tuberculosis sequelae	7	18.92
Idiopathic factors	3	8.11
<i>Total</i>	37	100

Table 3. Preoperative bronchoscopic findings

	n	%
Stenosis	12	32.43
Secretion	17	45.95
Granulation tissue	8	21.62
<i>Total</i>	37	100.0

syndromes along with genetic and metabolic defects are the foremost causes in developed countries. Surgery for bronchiectasis is performed once conservative medical management has become ineffective or when the condition remains unresolved upon investigation, in spite of at least six months of treatment,^[10] and the procedure consists of resecting the affected part of the lung (segment, lobe, or entire lung). Inflammation and bacterial agents are commonly known as causes of bronchiectasis, but in rare instances, viruses and TB have also been the source. Pulmonary TB was defined as a prevalent cause of MLS in a study from Korea.^[11] *Haemophilus influenzae* in one case and *Streptococcus pneumoniae* in seven cases were found in most of the bacterial cultures of our patients. However, active TB was not identified in the seven patients who had been previously diagnosed with TB.

Middle lobe syndrome is predominantly diagnosed radiologically. Chest X-rays of either side remain valuable, particularly for displaying atelectasis with its increased density and border sharpness stretching toward the posterior. Simple radiological tests are considered to be as important as CT in the diagnosis of MLS.^[12] We identified 33 cases of bronchiectasis, three cases of lung destruction, and two cases of organized pneumonia on CT in this study. A bronchoscopy is an important investigative procedure that is also useful for the diagnosis and treatment of MLS. Not only can it be used to obtain bacterial cultures, but it can also establish the location of the disease. We evaluated the inlet of the middle lobe bronchus and condition of the proximal part via this procedure. However, in some cases, granulation tissue, tumors, or foreign body aspiration can cause bronchial obstruction and these can go undetected with a bronchoscopy.

The bronchoscopy procedure, whether rigid or flexible, assists in clearing the retained secretions and can be used to collect biopsy samples for microbiological investigation. All of the cases in this study underwent a preoperative rigid bronchoscopy, and microbiological investigations of the BALF aspirate cultures were carried out. Sputum was found in 45.95% of our patients, stenosis was seen in 32.43% of the cases and granulation tissue was discovered in an additional 21.62% of the subjects near the inlet of the right middle lobe bronchus.

In their study, Onur et al.^[4] reported right middle lobe atelectasis was present in 3% of children who complained of chronic cough. They also found that allergic asthma was the most common underlying disorder in 23.9% of their cases. In addition, they

determined that TB was present in 11.9% of their study participants. De Boeck et al.^[13] reported that the long-term pulmonary results of MLS in children and its accompanying symptoms were prone to progress at the first diagnosis during follow-up. Furthermore, Priftis et al.^[14] determined that more than half of the children in their study had bacterial infection without pneumonia with middle lobe collapse. In this study, *Haemophilus influenzae* and *Streptococcus pneumoniae* were prominent. Positive results on bacterial cultures are associated with the pathological role of atelectasis. If the cause of obstruction is unclear when MLS is diagnosed, bronchiectasis, bronchitis, organized pneumonia, and atelectasis should be kept in mind.^[3] Many of our patients had often received medical treatment for pneumonia and chronic lung infection, and seven patients had been administered antituberculosis treatment.

The main principle for treating pediatric patients is to eliminate the underlying pathological condition. For this reason, antibiotic coverage, postural drainage, bronchodilator therapy, and a conservative approach with regard to allergic causes has been performed in practice.^[3,5,15,16]

Most of the causes of MLS are thought to be linked with asthma, and patients could possibly recover spontaneously. However, recovery from the acute symptoms of MLS might not take place due to the re-expansion of atelectatic pulmonary parenchyma following an asthma attack. Therefore, middle lobe atelectasis may remain undiagnosed, and the patient may experience severe attacks of recurrent infections, inflammation, and obstruction. This can lead to bronchiectasis on an ongoing basis; hence, surgical resection might be warranted.^[5,14,17]

According to our experience, the enlargement of lymph nodes due to infective or inflammatory causes impairs middle lobe drainage, which can result in recurrent infections in the middle lobe. Once this status becomes chronic, it commonly results in bronchiectasis or lung destruction.

Patients must be also be checked for TB, and symptomatic patients considering surgery should undergo pulmonary resection.^[1] Surgical removal of the middle lobe is advisable when inadequate re-expansion, bronchiectasis, complete bronchial obstruction, atelectasis with systemic symptoms, or a high risk of chronic infection in the remaining lung is present.^[5,13] Some authors have written that the main method of treatment for MLS or lingular segment syndrome can change.^[18] For example,

Livingston et al.^[19] reported a 33% cure rate with a bronchoscopy, which plays an important role in a conservative treatment plan. The patients in our study had intermittently been treated during the mean period of four or five years before any surgical procedure was performed. A lobectomy was carried out on those cases that were unresponsive to long-term medical treatment in which middle lobe atelectasis had become chronic and showed no evidence of aeration within the parenchyma. After surgery, atelectasis occurred in five of our patients, prolonged air leaks were seen in four, bleeding took place in one, and pneumothorax was also identified in one other case. However, no recurrence of pneumonia was present, and complete recovery from all symptoms was evident in all of the patients postoperatively. Attention should also be given to endobronchial obstruction caused by a malignant neoplasm since this yields the same radiological findings as MLS.

Conclusion

Middle lobe syndrome should be considered for patients complaining of chronic cough and recurrent pneumonia attacks, and chest X-rays should be used to evaluate the patients. Initially, the etiological factor must be confirmed in order to determine the proper course of treatment. We believe that a lobectomy is the treatment of choice when bronchiectasis, bronchial stenosis, re-expansion defect, or chronic, recurrent symptoms are present.

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