Case Report / Olgu Sunumu

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Severe Asthma Attack-associated Middle Lobe Syndrome in an Uncontrolled Asthma Patient

Uzun Süreli Takipsiz Astım Hastasında Ağır Astım Atağı İlişkili Orta Lob Sendromu

Muhammed Yusuf Mila[1], Talat Sürücü[1], İlyas Bingöl[1], Ceren Ören[1], Hakan Gemici[1], Burçin Beken[2]

[1]University of Health Sciences Turkey, Kanuni Sultan Süleyman Training and Research Hospital, Clinic Department of Pediatrics, İstanbul, Turkey
[2]University of Health Sciences Turkey, Kanuni Sultan Süleyman Training and Research Hospital, Clinic of Pediatric Allergy and Immunology, İstanbul, Turkey

Abstract

Middle lobe syndrome is a well-defined clinical and radiological entity in the pediatric literature, The causes include many pathologies such as asthma, lymphadenopathy, tumor, foreign body aspiration, granulation tissue, mucus plug, bronchopulmonary dysplasia, and cystic fibrosis. A nine-year-old girl with a diagnosis of asthma, who lack regular follow-up visits and a regular treatment, was admitted to the pediatric emergency department with respiratory distress. On physical examination; tachipnea, dyspnea, bilateral wheezing was revealed. Oxygen saturation was 80% in room air. Chest X-ray showed atelectasis in the right middle lobe and bilateral pneumonic infiltrates, more prominently in the right lung. The patient was started a non-invasive mechanical ventilation support, a broad-spectrum antibiotic therapy, asthma medication, N-acetylcysteine nebule therapy and chest physiotherapy. The chest X-ray showed improvement in atelectasis on the 5th day of treatment. Middle lobe syndrome has a very good prognosis when diagnosed early and treated appropriately. It should be kept in mind in patients with recurrent or persistent respiratory symptoms. Asthma is one of the most common causes of middle lobe syndrome in children, and uncontrolled asthma patients constitute a high risk. It is very important for asthma patients to continue their regular follow-up and to receive appropriate treatment according to their disease control.

Keywords: Asthma, atelectasis, middle lobe syndrome

Öz

Orta lob sendromu sıklıkla sağ orta bronşun dıştan bası ya da içeriden mukus tıkacı ile tıkanması sonucu ortaya çıkan bir klinik tablodur. Etiyolojide astım, lenfadenopati, tümör, yabancı cisim aspirasyonu, granülasyon dokusu, mukus tıkacı, bronkoakciğer displazi, kistik fibrozis gibi pek çok patoloji yer almaktadır. Astım tanılı ancak takipsiz olan ve düzenli tedavi almayan dokuz yaşında kız hasta çocuk acil servise solunum sıkıntısı şikayetiyle başvurdu. Oksijen satürasyonu oda havasında %80 olan hastanın fizik muayenesinde; takipne, dispne, akciğerlerde sağda daha belirgin olmak üzere yaygın ronküsler ve ekspiryum uzunluğu mevcuttu. Akciğer grafisinde sağ akciğer orta lobda atelektazi ve sağda daha belirgin olmak üzere bilateral pnömonik infiltrasyon olduğu görüldü. Non-invaziv mekanik ventilasyon desteği verilen hastaya geniş spektrumlu antibiyotik tedavisi ve astım atağına yönelik tedavinin yanı sıra N-asetilsistein nebül tedavisi verildi ve göğüs fizyoterapisi uygulandı. Tedavini beşinci gününde çekilen akciğer grafisinde atelektazinin düzeldiği görüldü. Orta lob sendromu erken tanı ve uygun tedavi ile prognozu oldukça iyi olan bir tablo olup, tekrarlayıcı ya da persistan solunum semptomu olan hastalarda mutlaka akılda tutulmalıdır. Astım çocuklarda orta lob sendromuna en sık yol açan tablolardan birisidir ve kontrolsüz astım hastaları riskli gurubu oluşturmaktadır. Astım hastalarının düzenli kontrollerine devam etmeleri ve hastalık kontrolüne göre uygun tedavi almaları oldukça önemlidir.

Anahtar Kelimeler: Astım, atelektazi, orta lob sendromu

Address for Correspondence/Yazışma Adresi: Burçin Beken, University of Health Sciences Turkey, Kanuni Sultan Süleyman Training and Research Hospital, Clinic of Pediatric Allergy and Immunology, İstanbul, Turkey
E-mail: burcinbeken@gmail.com **ORCID ID:** orcid.org/0000-0001-7677-7690
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Introduction

Middle lobe syndrome (MLS) is a well-defined clinical and radiological condition in the pediatric literature.[1] The features of lung involvement were first described in 1937 by Brock et al.[2] in children with tuberculous lymphadenitis, but the term MLS was first used by Graham et al.[3] in a series of 12 cases published in 1948, in which atelectasis developed as a result of compression of the peribronchial lymph nodes to the middle lobe bronchus. Culiner[4] stated in 1966 that the main cause of MLS was bronchial compression and that this picture emerged as isolated in the right middle lobe due to the absence of collateral ventilation. Then, Wagner and Johnson[5] expanded the definition of this syndrome by stating that atelectasis could develop without external pressure on the bronchi. It has been observed over time that although this clinical picture is mostly in the right middle lobe, it can also occur in other areas of the lung.

It is usually seen with a history of asthma or atopy in the childhood age group. Also, in etiology, many pathological conditions such as lymphadenopathy, tumor, foreign body aspiration, granulation tissue, mucus plug, bronchopulmonary dysplasia, cystic fibrosis play a role, and the exact prevalence is unknown. It is more common in the preschool age group in children, and the median age of diagnosis varies between 3.3 and 5.5 years.[4,5] Middle lobe syndrome is a clinically overlooked diagnosis, and delay in diagnosis is responsible for both the high economic burden of drug overuse and potentially poor long-term outcomes. This case was presented and discussed in the light of the literature, in order to draw attention to severe asthma attack and related MLS in asthma patients without follow-up.

Case Report

A nine-year-old female patient was admitted to the pediatric emergency department of our hospital with the complaint of respiratory distress. It was learned from the patient's history that she had recurrent wheezing attacks since she was 2 years old, she received nebular treatments in the emergency department at the time of the attack, she never received regular inhaled corticosteroid treatment, and she did not apply to the hospital because she had no attacks for 3 years. Her vital signs at the time of admission were as follows; temperature: 37.6 °C, respiratory rate: 60/min, pulse: 140/min, blood pressure: 115/50 mmHg, oxygen saturation: 80% in room air, 94% with oxygen. In the physical examination, her general condition was moderate-poor, dyspneic, with diffuse rhonchi and expiratory length in the lungs, more prominently on the right side. In the laboratory tests of the patient whose other system examinations were normal, hemoglobin was 12.7 g/dL, leukocyte was 16300/mm^3, neutrophil was 14780/mm^3 lymphocyte was 1100/mm^3, platelet was 331000/mm^3 and crp was 2.1 mg/dL. There was mild lactic acidosis in blood gas. Chest X-ray showed atelectasis in the middle lobe of the right lung (Figure 1A).

In the thorax computed tomography, there was an increase in density consistent with diffuse atelectasis, including air bronchograms observed as extending from the posterior of the upper lobe apical segment in the right lung to the hilar section and to the middle lobe to the lower lobe anterobasal segment on the right and at the level of the lower lobe anterobasal segments on the left, and there were findings consistent with prominent bilateral bronchopneumonic infiltration on the right (Figure 2).

The patient, whose Coronavirus type-19 (Coronavirus disease-2019) polymerase chain reaction test was performed twice and was found to be negative, was hospitalized in the pediatric intensive care unit with the preliminary diagnosis of asthma attack and right middle lobe syndrome. Meropenem and teicoplanin antibiotics were started. Respiratory support was given with Bilevel positive airway pressure. 2 mg/kg/day methylprednisolone, continuous salbutamol nebulization, ipratropium bromide nebulization, intravenous (iv) magnesium sulfate ($MgSO_4$), and iv aminophylline infusion treatments together with N-acetylcysteine nebules were started for asthma attack. Chest physiotherapy and postural drainage were applied to the patient. According to the improvement in the patient's auscultation findings, iv aminophylline and $MgSO_4$ treatments were discontinued on the 2nd day of hospitalization, respectively. On the fifth day of hospitalization, chest X-ray showed improvement in atelectasis (Figure 1B). Antibiotherapy and steroid treatment of the patient, who was transferred to the ward after a seven-day intensive care follow-up, was discontinued on the tenth day, and nebular salbutamol and ipratropium treatments were gradually decreased and discontinued on the eleventh day.

In the bronchoscopy performed on the patient for the differential diagnosis of foreign body aspiration or intrabronchial obstructive lesion, edema and occasional sticky occlusive secretions were observed in both bronchial systems, and no foreign body, external compression appearance or mass was detected. Purified protein derivative test performed for tuberculosis exclusion, and acid-fast bacillus staining taken from fasting gastric juice for 3 days and tuberculosis cultures were negative. The patient's immunoglobulin A, G, M levels were 1.18 g/L, 10.84 g/L, and 1.37 g/L, respectively, and were within normal limits for her age. It was observed that IgE level was 569 IU/L, *Dermatophagoides pharynea* specific IgE level was 22 kU/L, and *Dermatophagoides pteronyssinus* specific IgE level was 40.2 kU/L.

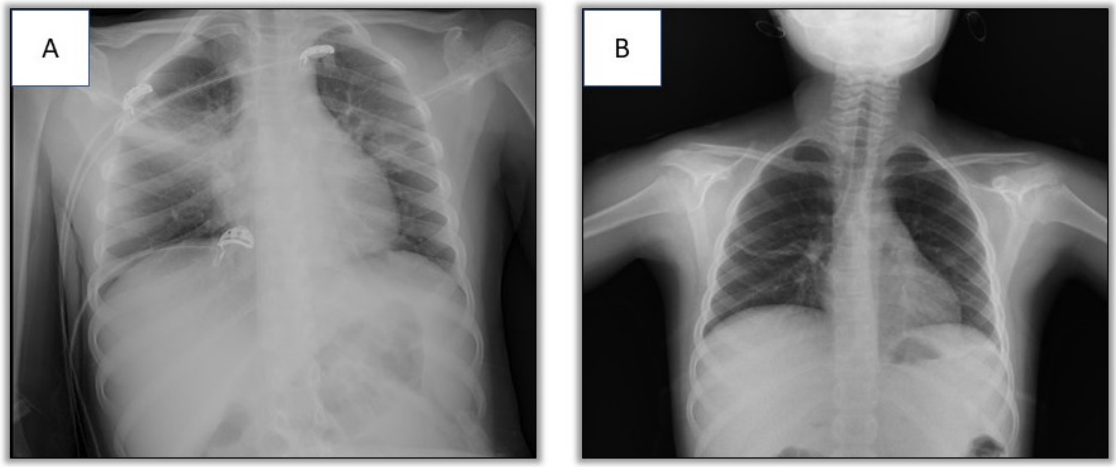

Figure 1. A) On the patient's chest X-ray taken at the time of admission, view of atelectasis in the middle lobe of the right lung and bilateral paracardiac infiltration, more prominently on the right, **B)** On the chest X-ray taken on the fifth day of the patient's treatment, it is seen that atelectasis has improved

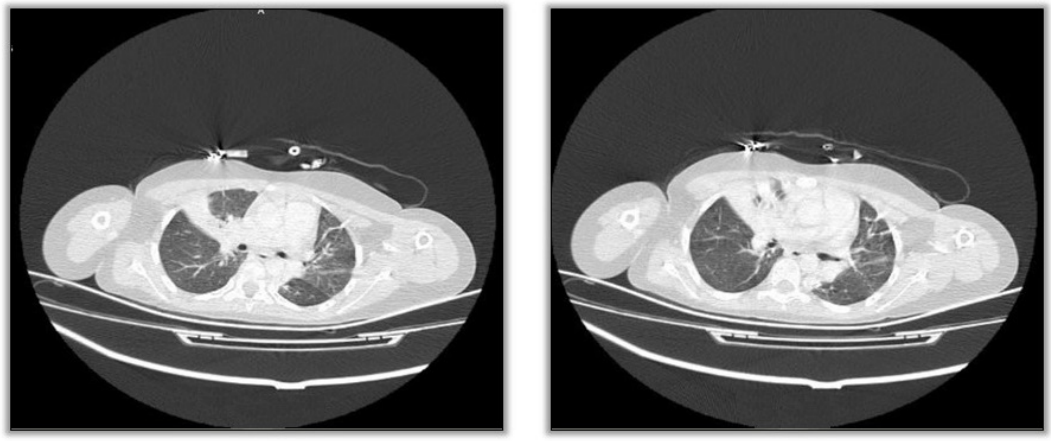

Figure 2. In the thorax computed tomography, there was an increase in density consistent with diffuse atelectasis, including air bronchograms observed as extending from the posterior of the upper lobe apical segment in the right lung to the hilar section and to the middle lobe to the lower lobe anterobasal segment on the right and at the level of the lower lobe anterobasal segments on the left, and there were findings consistent with prominent bilateral bronchopneumonic infiltration on the right

Fluticasone propionate (250 mcg) + salmeterol (50 mcg) metered dose inhaler treatment was started during discharge, and no pathology was detected in the physical examination and chest X-rays in the 3rd and 6th month control examinations, and she is being followed by the department of pediatric immunology and allergy diseases.

Discussion

Although middle lobe syndrome is a complication of childhood asthma, its exact prevalence and incidence are unknown. Studies have reported that 5-10% of patients hospitalized with acute asthma attack have radiological findings of lobar collapse.[6,7] However, these studies are quite old and were conducted before preventive anti-inflammatory treatments for asthma were begun to be widely used.

In a study conducted in Turkey in 2004, 3.528 patients who applied to the pediatric allergy clinic for a two-year period were evaluated and it was observed that the middle lobe syndrome was found in 56 (1.58%) patients, emerging only once in 50 patients, 2 times in 5, and 3 times in 1 patient.[8] It was stated that the symptoms of the patients at the time of admission were respiratory distress, cough, sputum, fever and wheezing.[8] Our patient applied to the emergency department with respiratory distress. Again, in the same study, it was observed that atelectasis continued for an average of 45 days in 36.5% of the patients, and although it was not statistically significant, patients who took systemic corticosteroids for 10 days recovered faster than those who did not receive corticosteroids. The authors have stated that the most important clinical clue for middle lobe syndrome is prolonged asthma symptoms and that the complaint or recovery period lasting more than 2 weeks can be used as a limit to investigate the complication of atelectasis. Early initiation of corticosteroid therapy was thought to be effective in the rapid recovery of atelectasis in our patient.

Reasons suggesting that young children are more prone to atelectasis are that in early childhood, the airways tend to be closed with smaller and larger peripheral airway resistance, the chest wall is more compliant, and collateral ventilation is not fully developed.[9] Although the clinical findings in middle lobe syndrome are variable and may be asymptomatic, chronic/recurrent cough, sputum, recurrent wheezing or recurrent/persistent pneumonia are the most common findings.[10] Since the symptoms are quite non-specific, the diagnosis is made late in about half of the patients.[10] Delays in taking chest X-ray in patients with non-specific, mild persistent symptoms may result in missed diagnosis of long-standing middle lobe syndrome. Findings such as mild fever, weight loss, malaise, chest pain or hemoptysis suggest complications associated with suppurative infection. The history usually includes having taken antibiotics, anti-mucolytic or anti-asthmatic drug therapy several times. The physical examination of the patients is usually normal, and rales and rhonchi localized to the middle lobe can be heard.[11]

In asthmatic patients, the airway lumen may be partially or completely obstructed due to viral infections, poor clearance of inflammatory debris, smooth muscle contraction, and edema in the bronchial wall.[1] Since the right middle lobe bronchus is separated from the main bronchus at a steeper angle and is shorter, drainage is more difficult. Therefore, the most affected lobe is the right middle lobe. In the differential diagnosis of patients with middle lobe syndrome, foreign body aspiration, cystic fibrosis, primary ciliary dyskinesia, primary immune deficiencies, neuromuscular diseases, mass causing external compression to the bronchus, lymphadenopathy or endobronchial tumors should be kept in mind.[1] No external or internal compression was detected in the bronchoscopy of the patient, no foreign body was observed, her tomography revealed no pathology in the lung parenchyma, bronchiectasis or enlarged lymph node, there was no pathology in the examinations carried out for tuberculosis and immune deficiency.

In the presence of concomitant infection in middle lobe syndrome, treatment is in the form of antibiotherapy including bacteria such as "*Streptococcus pneumoniae, Haemophilus influenzae, Moraxella catarralis*", bronchodilator and inhaled corticosteroid treatments for asthma patients. It is very important to ensure airway clearance in patients with chronic productive cough or mucus plug findings. Inhalation of 3-14% hypertonic saline or dry powdered mannitol together with chest physiotherapy may be beneficial. While flexible bronchoscopy is diagnostically important, recovery is seen in the majority of patients after bronchoalveolar lavage. Although mucolytic therapy is not recommended in asthma, there are studies showing that it is beneficial in middle lobe syndrome. In addition to its mucolytic effect, N-acetyl-cysteine also has strong antioxidant properties, and it has also been shown to have positive effects on preventing airway hypersensitivity, inflammation and goblet cell hyperplasia in asthma mouse models.[12] In our patient, in addition to the treatments for asthma attack, acetylcysteine nebul and postural drainage techniques applied together were thought to have an effect on the early recovery of atelectasis.

The prognosis of middle lobe syndrome is quite good. In a study in which 17 patients were evaluated, it was reported that mild obstructive airway symptoms persisted in 1/3 of the patients, and cylindrical bronchiectasis developed in only 1 patient in a 10-year follow-up.[13] In a study of 55 patients by Priftis et al.[10], the rate of bronchiectasis was reported as 27.3%. The development of bronchiectasis was found to be less in patients who underwent early bronchoscopy and bronchoalveolar lavage. In the 3rd and 6th month follow-ups of our patient, the physical examination was normal, no atelectasis or bronchiectasis was detected in the chest radiographs, and her regular follow-up is being continued.

In conclusion, middle lobe syndrome is a picture with a very good prognosis with early diagnosis and appropriate treatment, and it should definitely be kept in mind in patients with recurrent or persistent respiratory symptoms. It is especially important to regulate the treatment of asthma patients for disease control.

Ethics

Informed Consent: Permission was obtained from the patient's family.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Concept: B.B., Design: B.B., Data Collection or Processing: M.Y.M., T.S., İ.B., C.Ö., H.G., B.B., Analysis or Interpretation: B.B., Literature Search: B.B., Writing: M.Y.M., T.S., İ.B., C.Ö., H.G., B.B.

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