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Middle Lobe Syndrome in Children

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ABSTRACT

Background: Atelectasis of the middle lobe or lingula of the lung is defined as middle lobe syndrome. On chest x-ray it is demonstrated as a wedge shaped density with anterior-inferior extension from the hilum. Although many etiologies have been implicated, this syndrome is one of the most common complications of asthma.

Materials and Methods: A simple descriptive study was conducted on 11 patients with an age range of 0-18 yrs. They were admitted to Masih Daneshvari Hospital during 2000-2007 with the diagnosis of lingula or middle lobe atelectasis (of more than one month duration) and / or recurrent consolidation (2 times or more).

Results: The study group consisted of 6 boys (54.5%) and 5 girls (45.5%). All patients were clinically symptomatic at the time of admission. Cough was the chief complaint (7patients, 63.6%). The mean age at the time of initial diagnosis was 7.3 yrs (SD: 1.6). The most common findings on pulmonary CT-scan were infiltrations (3 cases, 27.3%) and atelectasis (3 cases, 27.3%).

Non-obstructive causes were the most frequent etiologies which included asthma (n=3, 27.3%), pneumonia (n=2, 18.2%) and bronchiectasis (n=2, 18.2%). Among the obstructive causes, an undefined tumor (1 case, 9.1%) was to mention.

Nine cases (81.8%) had negative blood cultures and 9 cases (81.8%) had AFB negative sputum smears (3x). Bronchoscopy was performed in 4 (36.4%); which showed rapid improvement after fiberoptic bronchoscopy (FOB). Medical treatment was planned for 9 children who demonstrated quick recovery. Surgery (lobectomy) was conducted in only 1 patient.

Conclusion: Patients with right middle lobe syndrome (RMLS) had airway hypersensitivity, which is supported by the fact that asthma is very severe in this group of patients. Despite its low incidence, it should be considered very carefully and cautiously since it is associated with many severe complications. Therefore in undiagnosed suspected cases, in addition to a meticulous history taking, detailed diagnostic and therapeutic measures are recommended. (*Tanaffos 2009; 8(1): 50-55*)

Key words: Middle lobe syndrome, Children, Asthma, Atelectasis

INTRODUCTION

Right middle lobe syndrome (RMLS) presents as

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a group of clinical symptoms consisting of chronic cough, intermittent wheezing and dyspnea (1). On CXR it is demonstrated as recurrent atelectasis of the right middle lobe and / or lingula which may be resistant to treatment (2, 3).

Because of the nonspecific symptoms, it is easily

misdiagnosed in children (3). A large number of children with right middle lobe infiltrations or atelectasis receive various antibiotic regimens and inhaler corticosteroids for persistent recurrent pulmonary infections. Some even go further and experience surgical interventions (such as lobectomy) for irreversible pulmonary changes (4). Thus, bronchoscopy and bronchoalveolar lavage must be performed along with other diagnostic measures as early as possible; it should not be delayed more than 3-7 months after the appearance of clinical features (5, 6).

The surgical indications of lobectomy for RMLS include association with bronchiectasis, bronchial stenosis and failure of lung re-expansion (7).

According to various reports, 15% of the reported cases occur in children (8).

We evaluated 11 children with RMLS during a 7-year period.

MATERIALS AND METHODS

A simple descriptive study was conducted to assess 11 children with RMLS admitted to the pediatric ward of Masih Daneshvari Hospital from 2000 to 2007. The medical files of patients who had atelectasis on CXR (n=40) were chosen and those with RMLS were selected.

Right middle lobe syndrome was implicated if atelectasis (of more than 1 month duration) and / or recurrent consolidation (2 times or more) of the right middle lobe or lingual were present. The inclusion criterion was age less than 18 yrs of age. Exclusion criteria consisted of age > 18 yrs and known causes of purulent bronchitis (such as cystic fibrosis and primary ciliary dyskinesia).

Medical treatment consisted of a combination of antibiotics, mucolytics, inhaler bronchodilators, inhaler corticosteroids and chest physiotherapy.

Lobectomy of one or more lobes constituted the surgical approach. Also, atopy (shown as high levels

of IgE for the studied age group) was evaluated. Data were analyzed by SPSS software version 16.5.

RESULTS

Eleven patients including 6 boys (54.5%) and 5 girls (45.5%) with a mean age of 7.4 yr (SD=1.6) were studied. All cases were symptomatic; symptoms in decreasing order of frequency included chronic cough (7 cases, 63.6%), wheezing (3 cases, 27.3%), dyspnea (2 cases, 18.2%), hemoptysis (1 case, 9.1%) and weight loss (1 case, 9.1%).

On clinical examination, diffuse rhonchi in 4 cases (36.4%) was the most common finding. The most frequent CXR and CT-scan findings included atelectasis and patchy infiltration, each in 3 cases (total of 6 cases, 54.6%). Other manifestations included mass consolidation (1 case, 9.1%) and bronchiectatic changes (1 case, 9.1%) (Figures 1, 2).



Figure 1-The PA radiograph of the chest revealing infiltration, collapse and consolidation of RML.

Laboratory investigation showed an eosinophil count of > 400 (>5%) in 3 (27.3%) and a count of <400 (<5%) in 7 patients.

Blood culture was performed in 9 patients; after 24 and 48 hrs both cultures were negative in all cases (81.8%).

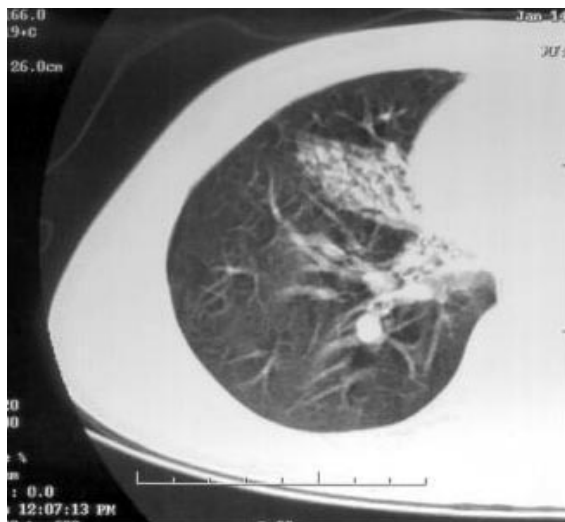


Figure 2. Chest CT-scan showed bronchiectatic changes in medial and lateral segments of RML.

Similarly, sputum smears for AFB (3×) prepared in 9 patients were all negative (81.8%).

Immunologic evaluation demonstrated a high IgE titer in only one case who had asthma (9.1%). Pulmonary function test (PFT) demonstrated obstructive pattern in 5 (45.5%) patients. FEV₁ (forced expiratory volume in 1 sec) was equal or less than 60% of the predicted level in 2 cases (18.2%), while in 3 patients (27.3%) it was greater than 80% of the predicted value.

Also, in this study, vital capacity and FEV₁ showed a significant decrease for values predicted according to age and gender of the patients.

Nine patients (81.8%) received medical treatment (antibiotics, mucolytics, bronchodilators, corticosteroids and chest physiotherapy). Surgery was performed in one case (9.1%). One patient (9.1%) was discharged on parents' consent without continuing the treatment. All 4 patients (36.4%) who underwent bronchoscopy showed gradual improvement. In addition to its diagnostic value, bronchoscopy was also used as a therapeutic tool.

Definite diagnoses were made in 9 patients (81.8%) of whom 7 (63.6%) had reversible

obstructive etiologies (asthma 3 cases , 27.3% ; pneumonia 2 cases , 18.2% ; bronchiectasis 2 cases , 18.2%) . The remaining 2 cases had obstruction due to tumor (1 case, 9.1%) and a cardiovascular anomaly (1 case, 9.1%)(Figure 3).

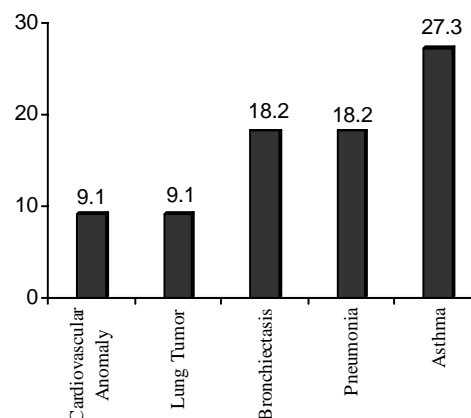


Figure 3- The etiology of patients with RMLS.

DISCUSSION

In our studies, common manifestations of RMLS ranged from recurrent atelectasis of middle lobe and lingula to pneumonia and bronchiectasis. The present study evaluated 11 children (0-18 yrs) with this syndrome who were admitted to the pediatric ward of Masih Daneshvari Hospital during 2000-2007. Akilov in Russia in 2003 evaluated 242 cases of RMLS during a 10-year period (9).

In 1995 Kwon et al. reported 21 RMLS cases in the age range of 5-80 yrs (10). The small number of patients in our study may be due to early diagnosis and early treatment.

A research conducted by Karakoc et al. in Turkey on 56 children with RMLS and bronchiectasis demonstrated the high prevalence of this syndrome among girls (twice as much as boys) (11). However, in the present study boys and girls constituted 54.5% and 45.5% of the cases respectively. Also, mean age at the time of initial diagnosis was 7.4 yrs (SD=1.6).

In Greece, Priftis et al. performed a study on 55 children with RMLS, showing a mean age of 5.5 yrs at the time of initial diagnosis (12).

Sekerel and Nakipoglu studied the medical files of 3528 asthmatic children in 2004 to assess RMLS as a complication of asthma. They observed that the incidence of asthma in RMLS patients was 1.62% (56 cases) and half the patients were below the age of 6 yrs. They also found that complicated cases were more frequent among non-atopic individuals (13). However, in the present evaluation only 3 RMLS patients suffered from asthma and complicated cases were more prevalent among atopic patients.

With regard to the etiology of RMLS, reversible obstructive airway disorders such as asthma comprised 27.3% of the cases in this study. Meanwhile, in a study performed by Nemr in 2007 in the USA, 44% of RMLS cases were due to primary respiratory disorders such as asthma, cystic fibrosis and bronchopulmonary dysplasia (1).

Rollan et al. in Spain demonstrated only 2 cases with obstruction as the etiology of RMLS (foreign body and endobronchial tumor) (8). We had 2 cases of obstruction as the cause of RMLS (a case of tumor existing outside the airway and one case of cardiovascular anomaly).

Infective agents such as recurrent lobar pneumonia were observed in 18.2% of our cases while Youssef Fasheh showed a 66.7% rate in his study conducted during 1992-1996 in Spain (14). In his research, bronchiectasis was detected in 7 out of 27 patients; this figure was 2 cases in our study.

In our study, the most common findings on CXR and pulmonary CT scan were atelectasis (27.3%) and pathologic infiltration (27.3%) and 9.1% of the patients exhibited bronchiectatic changes.

Eastham et al. in their study on 55 patients with RMLS showed that 27.3% had bronchiectatic changes on HRCT (15).

Sekerel and Nakipoglu in 2004 in Turkey demonstrated that 36.5% of the sick children had atelectasis (13).

A study performed by Wong in Taiwan compared pulmonary CT-scan and bronchoscopy to determine the etiology of atelectasis and obstruction. The results showed that lung scan must be performed in

all patients while bronchoscopy must be saved for recommended cases only (16). As it is clear, CT-scan was performed for all 11 cases in our study while bronchoscopy was performed in only 4 (36.4%), among which abnormal findings were observed in 3 (27.3%).

Based on various researches, bronchoscopy and CT-scan are essential for further evaluations if atelectasis and / or clinical features persist despite supportive treatment for at least one month.

Kawamura in Japan in 2001 reported 2 patients with RMLS that showed complete clinical and radiological recovery after receiving low dose roxithromycin (17). One of our patients in the present research was cured after receiving a combined regimen of azithromycin and ceftriaxone.

A 10-year research (1995-2005) conducted by Priftis et al. in Greece, evaluated the therapeutic response in children with bronchiectasis and those without it by HRCT. Their results demonstrated a more significant therapeutic response in patients who did not have bronchiectasis (18). Likewise, despite receiving medical treatment and demonstrating clinical improvement, patients with bronchiectatic changes in the present study did not show any positive change in radiologic features; a fact that necessitated urgent treatment.

Ayed in Kuwait in 2004 recommended bronchoscopy in the early stages of the syndrome. They also considered bronchoscopy with CT-scan as a tool in determining the progression of the syndrome towards bronchiectatic changes (7). In addition, in their study lobectomy was performed in 53% while in the present study only 9.1% of the patients were treated by surgery who had no bronchiectatic changes on radiographs.

In Perez-Ruiz et al. (19) and Chang et al. (20) studies, diagnostic bronchoscopy of patients showed that although at present, rigid bronchoscopy is the most common method for treating respiratory problems, flexible bronchoscopy is a more useful tool for evaluating the airways and detecting respiratory problems in children.

CONCLUSION

Clinical features of RMLS are nonspecific and may lead to misdiagnosis. Carefully conducted diagnostic procedures such as CXR are valuable when facing vague nonspecific clinical pictures in children. They can help to reach an early diagnosis and treatment and preventing the progression of this syndrome towards bronchiectasis. Since the main aim of treatment is to prevent the appearance of bronchiectasis in this group of patients, early on-time diagnosis and treatment are essential.

When confronting patients with RMLS, in addition to asthma and foreign body aspiration, other etiologies such as cardiovascular anomalies, tumors, and GI reflux should be considered.

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