Original Article

Surgical treatment of right middle lobe syndrome in children

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Abstract:

OBJECTIVE: Right middle lobe syndrome is a rare entity in children, causing high morbidity. Our experience of these patients including their clinical and laboratory characteristics, indications forsurgical management, postoperative courses, and follow-up results was evaluated.

METHODS: A retrospective analysis was performed involving 20 children with right middle lobe syndrome who were hospitalized and treated with surgical resection of the right middle lobe in Van Training and Research Hospital and Yüzüncüyıl university hospital, Turkey, between January 2002 and January 2011.

RESULTS: The mean age of the patients was 10.5 years (range, 5 to 15 years). Twelve patients were boys and eight were girls. The most frequent symptom was chronic cough (75%). Hemoptysis was present in two (10%) patients. One patient was being treated for asthma. 25% positive cultures were identified among the patients. *Streptococcus pneumoniae* was the most frequently identified agent in sputum. All patients underwent chest computed tomography. There were bronchiectasis in 11 (55%) patients, atelectasis and bronchiectasis in five (25%) patients, and destroyed lung in four (20%) patients. A narrowed middle lobe bronchus was shown in 15 (75%) patients. Bronchoscopy was performed in 18 (90%) patients. Stenosis due to external compression was seen in 12 (60%) patients, hyperemia and bronchitis in two (10%) patients, granulation tissue in two (10%) patients, and dense secretions in two (10%) patients. A history of doctor-diagnosed tuberculosis was present in two (10%) patients. These patients had completed antituberculous treatment. The patients had been symptomatic for the last 1 to 10 years (mean, 4 years) and had received several medical treatments. All patients (totally 20 patients) underwent right middle lobe resection. In one patient, a bronchial abnormality was found intraoperatively. One patient died on postoperative day 10 due to a brain abscess. Three other patients had postoperative complications (15%). Mean duration of follow-up of the patients was 4.5 years (range, 2 months to 12 years). Seventeen patients were asymptomatic, and two patients had improved.

CONCLUSIONS: Children with right middle lobe syndrome unresponsive to medical treatment should undergo early lobe resection to avoid serious complications and the progression of the disease to other segments or lobes.

Key words:

Atelectasis, bronchiectasis, children, pulmonary resection, right middle lobe

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Right middle lobe syndrome is a rare entity in children, causing high morbidity. Right middle lobe syndrome is characterized by a spectrum of diseases from recurrent atelectasis and pneumonitis to bronchiectasis of the middle lobe.[1] Nonobstructive atelectasis due to poor collateral ventilation is the most likely mechanism for the development of the right middle lobe syndrome.[2-5] The term right middle lobe syndrome was originally coined by Graham *et al*.[6,7] They reported on 12 patients with right middle lobe atelectasis and bronchial compression caused by nontuberculous enlarged lymph nodes, and all patients were treated by lobectomy.

We aimed to examine the specific group of these patients. The purpose of this study was to analyze our experience of these patients with their clinical and laboratory characteristics, indications for surgical management, postoperative courses, and follow-up results.

Methods

A retrospective analysis was performed of 20 children with right middle lobe syndrome who were hospitalized and treated with surgical resection of the right middle lobe in Van Training and research Hospital and Yüzüncüyıl University hospital, Turkey, between January 2002 and January 2011. The data obtained included age, sex, presentation and symptoms, etiologic factors, radiologic and bronchoscopic findings, microbiologic examinations, surgical treatments, postoperative complications, and results. Follow-up of the patients lasted from 2 months to 12 years.

Results

The mean age of the patients was 10.5 years (range, 5 to 15 years). Twelve patients were boys and eight were girls. The most frequent symptom was chronic cough (75%). Hemoptysis was present in

two (10%) patients. One patient was being treated for asthma. *Streptococcus pneumoniae* was the most frequently produced agent in sputum cultures. All patients underwent thorax computed tomography (CT) or high-resolution CT (HRCT). The results revealed bronchiectasis in 11 (55%) patients, atelectasis and bronchiectasis in five (25%) patients, and destroyed middle lobe in four (20%) patients. A narrowed middle lobe bronchus was seen in 15 (75%) patients. Bronchoscopy was performed in 18 (90%) patients. Stenosis due to external compression was revealed in 12 (60%) patients, hyperemia and bronchitis in two (10%) patients, granulation tissue in two (10%) patients, and dense secretions in two (10%) patients. A history of doctor-diagnosed tuberculosis was present in two (10%) patients. These patients had completed antituberculous treatment. The patients had been symptomatic for the last 1 to 10 years (mean, 4 years) and had received several medical treatments. The treatments included antibiotics, mucolytic agents, bronchodilators, steam application, chest physiotherapy, and postural drainage. All patients (20 patients) underwent right middle lobe resection. The decision criteria for the surgical resection of the right middle lobe were as follows: Repeated episodes of infection with lobar collapse, evidence of bronchostenosis, bronchiectasis [Figures 1 and 2], and destroyed lobe. In one patient, a bronchial abnormality was found intraoperatively (middle lobe bronchus was short with very early branching of the medial and lateral segments). One patient died on postoperative day 10 due to a brain abscess. Three other patients had postoperative complications (15%). Postoperative complications included atelectasis in one patient, brain abscess in one patient, and reoperation after hemorrhage in one patient. Atelectasis was treated with respiratory exercise and nasotracheal aspiration, no bronchoscopy was needed. The indication for reoperation was oozing 300 ml blood a day through the chest tube in a 7- year-old child. Intercostal artery ligation was performed. Mean follow-up of the patients lasted 4 years (range, 2 months to 12 years). Seventeen patients were asymptomatic, and two of three patients who developed complications improved (bleeding and atelectasis). The results are summarized in Table 1.

Discussion

Graham *et al*. in 1948 used the term middle lobe syndrome to describe middle lobe atelectasis resulting from bronchial compression (12 patients).[3] Anatomic characteristics, such as the narrow diameter of the lobar bronchus and an acute takeoff angle, make the right middle lobe susceptible to transient, usually partial, obstruction. Such an obstruction is considered to be the result of poor drainage of secretions due to inflammation and/or edema of the right middle lobe bronchus. In addition, the relative anatomic isolation of the middle lobe and the poor collateral ventilation decrease the chance of reinflation 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.[1,8-10] Subsequent reports suggest that ineffective collateral ventilation is a major factor in the pathogenesis of middle lobe syndrome.[11] Our experience indicates that the middle lobe is prone to develop persistent total or partial atelectasis with infection. Five patients in this series had atelectasis and bronchiectasis, and infection was a manifestation of the syndrome in all patients.

Wagner and Johnston published a review of 933 cases reported in the literature. In their review, the following etiologic causes of right middle lobe syndrome were noted: Inflammation, 47%; bronchiectasis, 15%; malignant tumors, 22%; benign tumors, 2%; tuberculosis, 9%; aspiration, 2%; and miscellaneous, 3%. It was not documented whether the atelectasis was caused by active disease or nodal compression.[3] The main bronchus to the middle lobe showed cicatricial stenosis at the point of maximum compression. This point correlated with the enlarged nodes on the external surface of the bronchus. Histological examination of the lymph nodes revealed changes only of a chronic, nonspecific lymphadenitis.[7] We also intraoperatively saw enlarged and scarred lymph nodes around the middle lobe bronchus in most of our cases. Stenosis due to external compression was revealed in 12 patients, hyperemia and bronchitis in two patients, granulation tissue in two patients, and dense secretions in two patients.

It was previously reported that swelling of lymph nodes due to either acute or chronic inflammation narrows the bronchus, leading to further retention of bronchial secretions, atelectasis, chronic bronchitis, or bronchiectasis. Healing of these recurrent episodes by fibrosis and scarring of the lymph nodes further contributes to collapse and narrowing of the

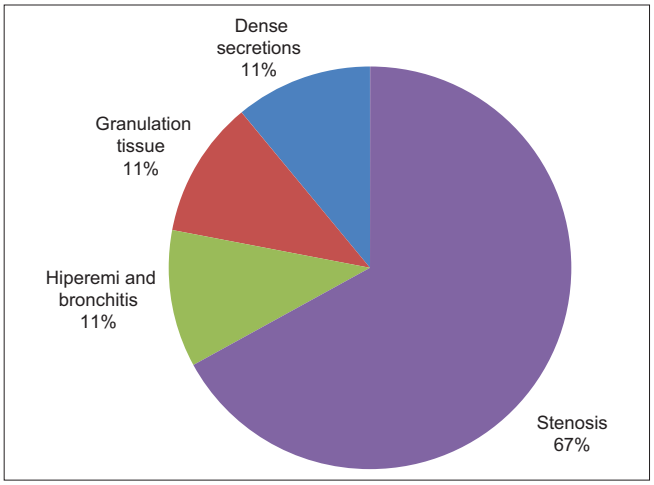

Figure 1: Bronchoscopic findings in 18 patients with middle lobe syndrome

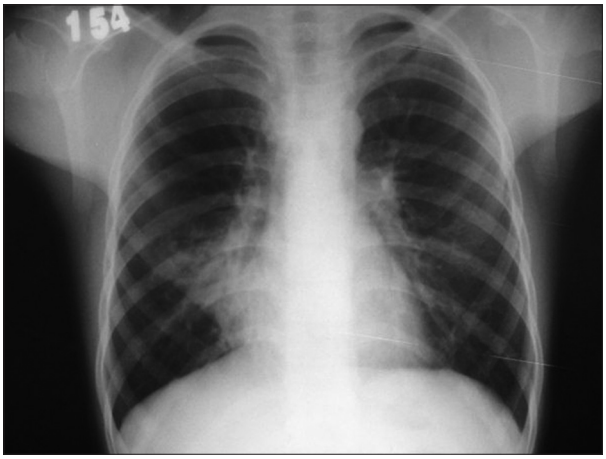

Figure 2: Posteroanterior radiograph of a 6-year-old child with bronchiectasis of middle lobe syndrome

Table 1: Summary of patient data

Patient no.	Year of operation	Age, yr	Sex	Period between onset of symptoms and surgery	Area resected	Bronchoscopy		
						Bronchoscopy (RML)	CT scan	Postoperative complications
1	2002	8	Male	8 mo	RML	Stenosis	Atelectasis	
2	2003	7	Female	1 yr	RML	Stenosis	Atelectasis and bronchiectasis	Hemorrhage
3	2004	15	Female	5 mo	RML	Secretions	Bronchiectasis	
4	2004	15	Male	2 yr	RML	Stenosis	Atelectasis and brocchiectasis	
5	2005	12	Female	1 yr, 6 mo	RML	Secretions	Bronchiectasis	
6	2005	11	Female	8 mo	RML	Granulation tissue	Atelectasis and brocchiectasis	
7	2005	14	Male	4 yr	RML	Secretions	Destroyed RML	
8	2006	14	Female	3 yr	RML	Stenosis	Atelectasis	
9	2006	6	Male	6 mo	RML	Stenosis	Bronchiectasis	
10	2007	6	Male	2 yr	RML	Stenosis	Atelectasis	
11	2007	13	Female	1 yr	RML	Stenosis	Atelectasis	
12	2007	7	Female	1 yr, 4 mo	RML	Secretions	Atelectasis	Brain abscess
13	2008	6	Male	1 yr	RML	Granulation tissue	Bronchiectasis	
14	2008	10	Female	2 yr	RML	Stenosis	Atelectasis	
15	2008	6	Male	1 yr	RML	Stenosis	Atelectasis	Atelectasis
16	2009	11	Male	6 mo	RML	Stenosis	Bronchiectasis	
17	2009	9	Female	3 yr	RML	Stenosis	Atelectasis	
18	2009	5	Male	6 mo	RML	Secretions	Atelectasis and brocchiectasis	
19	2009	9	Female	1 yr	RML	Secretions	Destroyed RML	
20	2010	7	Female	6 mo	RML	Stenosis	Bronchiectasis	

RML = Right middle lobe, CT = Computed tomography

bronchial divisions.[3] However, with repeated episodes of infection, through the resulting vicious cycle of recurring bouts of inflammation, the right middle lobe may eventually be destroyed completely.[2]

The reasons for obstruction can be intraluminal or extraluminal. Although the lymph nodes are the extraluminal reason for tumors and abnormal branching, object aspiration is the intraluminal reason for granulation tissue and mucous blockage.[2,12] Nonobstructive reasons were observed etiologically in most of the studies.

The existence of a nonobstructive etiology in middle lobe syndrome was especially indicated by the fact that the symptoms started in the early infantile period and decreased with age. In some series, the proportion of males was higher than that of females, whereas in other series, the opposite was found. The average age of these patients was 6.0 ± 1.1 years. The common reason in children for right middle lobe syndrome was inflammatory diseases such as bronchiectasis or pneumonia.[13,14]

The etiology of the infection is commonly bacterial and occasionally viral, and in some cases, it is caused by tuberculosis and histoplasmosis. Two patients in this series had tuberculosis. The diagnosis was made by a smear and culture of the bronchial aspirates. The microorganisms detected are those usually isolated in children, most frequently *S. pneumoniae*, followed by *Haemophilus influenzae* and *Staphylococcus aureus*.[9,15] In our study, *S. pneumoniae* was isolated in frequencies that were similar to those of other studies.

Clinical application in children with middle lobe syndrome was not found to be associated with the respiratory symptoms.[8,15] Conditions such as chronic cough, snuffles, dyspnea, repetitive pneumonia attacks, asthma, or atopy can be found in the anamnesis. If the nonspecific and mostly moderate respiratory symptoms of a patient were delayed with a chest graphy, it can be inadequate for diagnoses for middle lobe syndrome existing for a long time.

Posterior-anterior (PA) and lateral lung X-ray films, CT, HRCT, and bronchoscopy are used in the diagnosis.[1,12] HRCT is currently the modality of choice in the diagnosis of bronchiectasis, with only a 2% false-negative rate and 1% false-positive rate.[16] HRCT was performed in all patients in this series to document the presence of bronchiectasis. Bronchoscopy is important for diagnosis and treatment.[1] The situation of the proximal entrance of the middle lobe can be examined by bronchoscopy. Bronchial obstruction caused by the granulation tissue, a tumor, or a foreign body can be accurately excluded. At the same time, bronchoscopy enables microbiological examination and the cleaning of remaining secretions.[17]

Among the 18 patients, 66% had specific endoscopic findings of stenosis. Abnormal but nonspecific changes were present in 34% of these patients. Thus, the diagnostic yield by bronchoscopy in middle lobe syndrome appears to be almost 66%. The treatment of middle lobe syndrome is directed at the underlying cause [Figure 3].

For this purpose, antibiotic treatment, postural drainage, and bronchodilator treatment were applied, and it was suggested to avoid from allergen factors.[1-4]

For obstructing lesions, restoration of bronchial patency may be curative. Surgery may also be indicated in patients without malignancy who have scarring, fibrosis, or abscess formation. Broncholiths and foreign objects can often be removed with

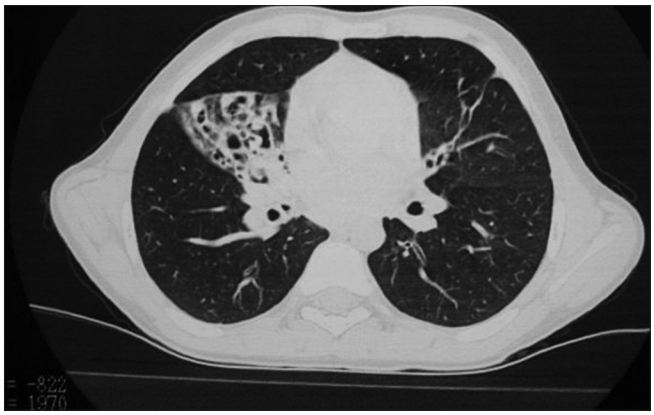

Figure 3: CT of 6-year-old child with bronchiectasis of the middle lobe

flexible fiberoptic or rigid bronchoscopy.[7] Intensive medical therapy is quite capable of resolving atelectasis and healing the patient. If endoscopic examination and CT scan did not show abnormalities after one month of the development of atelectasis, follow up with conservative treatment for another month is recommended.

If the middle lobe fails to re-expand, or recurrent atelectasis is observed, or when bronchiectasis is documented, surgical excision of the right middle lobe is indicated. Operative treatment also should be offered for patients with complete bronchial obstruction.[2]

Our patients were treated medically for 4.5 years interruptedly before surgical treatment. Middle lobectomy was applied in these cases that did not respond to medical treatment for a long time, in which the middle lobe atelectasis turned chronic and aeration of lung tissue was not observed.

One patient died on postoperative day 10 due to a brain abscess. A relapse of pneumonia symptoms was not observed in any of the other patients in the postoperative follow-up.

In conclusion, middle lobe syndrome must be kept in mind for children presenting with chronic cough and repetitive pneumonia attacks and the patient must be assessed with PA and lateral lung X-ray films. It is primarily important to determine the etiologic factors carefully before starting the treatment. In middle lobe syndrome patients who have bronchiectasis, bronchial stenosis, lung re-expansion defect, despite the medical treatment, curative results can be obtained by lobectomy in the cases of recurrent symptoms.

References

1. Livingstone GL, Holdinger LD, Luck SR. Right middle lobe syndrome in children. Int J Pediatr Otorhinolaryngol 1987;13:11-23.
2. Ayed AK. Resection of the right middle lobe and lingula in children for middle lobe/lingula syndrome. Chest 2004;125:38-42.
3. Wagner RB, Johnston MR. Middle lobe syndrome. Ann Thorac Surg 1983;35:679-86.
4. 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;1:266-72.
5. Fraser RS, Muller NL, Colman N. Radiologic signs of chest disease: Atelectasis. In: Fraser and Pare's diagnosis of diseases of the chest. 4th ed. Philadelphia, PA: WB Saunders; 1999. p. 513-62.
6. Graham EA, Burford TH, Mayer JH. Middle lobe syndrome. Postgrad Med 1948;4:29-34.
7. Miller JI. Bacterial infections of the lungs and bronchial compressive disorders. IN: Shields TW, editor. General Thoracic Surgery. 5th ed. Philadelphia: Lippincott Williams and Wilkins; 2000. p. 1039-52.
8. Sekerel BE, Nakipoglu F. Middle lobe syndrome in children with asthma: Review of 56 cases. J Asthma 2004;41:411-7.
9. Kwon KY, Myers JL, Swensen SJ, Colby TV. Middle lobe syndrome: A clinicopathological study of 21 patients. Hum Pathol 1995;26:302-7.
10. Springer C, Avital A, Noviski N, Maayan C, Ariel I, Mogel P, *et al*. Role of infection in the middle lobe syndrome in asthma. Arch Dis Child 1992;67:592-4.
11. Saha SP, Mayo P, Long GA, McElvein RB. Middle lobe syndrome: Diagnosis and management. Ann Thorac Surg 1982;33:28-31.
12. Onur BG, Can D, Asilsoy S, Gülle S, Alper H, Bak M. Right middle lobe syndrome in that to have recourse to cases with chronic cough: Retrospective study. Türk Toraks Dergisi 2006;7:104-8.
13. Youssef Fasheh W, Esquinas Rychen G. The middle lobe syndrome in pediatrics. A study of 27 cases. An Esp Pediatr 1998;49:582-6.
14. Franchini F, Pitta D, Pozzessere A, Cocchi C, Casini T. Middle lobe syndrome in children. Critical review. Pediatr Med Chir 1995;17:135-8.
15. Priftis NK, Mermiri D, Papadopoulou A, Anthracopoulos MB, Vaos G, Nicolaidou P. Lobe syndrome in children the role of timely intervention in middle. Chest 2005;128;2504-25.
16. Yung K, Aspestrand F, Kolbenstvedt A. High resolution CT and broncography in the assessment of bronchiectasis. Acta Radiol 1991;32:439-41.
17. Türkyılmaz A, Aydın Y, Ermancık M, Erdem AF, Eroğlu A. The surgical treatment of isolated middle lobe syndrome. Eurasia J Med 2007;39:15-18

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