Tuberculosis and Respiratory Diseases 결핵 및 호흡기질환, Vol. 55, No. 5, Nov, 2003

□ 증 례 □

Pneumothorax as the First Clinical Manifestation of Systemic Sclerosis : A Case Report of Multiple Cystic Lung Lesions in Systemic Sclerosis

Chang Hoon Lee, Seung Pyo Lee, Hee Suk Lee, Jin Young Oh, Woo Jin Kim, Jae Joon Yim, Chul Gyu Yoo, Sung Koo Han, Young Soo Shim, Young Whan Kim

Division of Pulmonary and Critical Care Medicine, Department of Internal Medicine and Lung Institute of Medical Research Center, Seoul National University College of Medicine

=국문초록=

기흉으로 발현한 전신성 경화증의 첫 증례

서울대학교 의과대학 내과학교실 및 의학연구원 폐연구소

이창훈, 이승표, 이희석, 오진영, 김우진, 임재준, 유철규, 한성구, 심영수, 김영환

26세 여자환자가 우측 흉통을 주소로 응급실을 방문하였다. 흉부방사선검사에서 우측 기흉을 진단받고 흉관삽입술을 시행하였다. 환자는 6년전부터 레이노드 현상이 있었고 양손에 피부경화증이 있었으며 항 DNA 국소이성화효소 I 항체가 양성으로서 전신성 경화증을 진단받았다. 고해상력 컴퓨터단층촬영에서 양쪽 폐야에 낭종성 병변이 관찰되었고 기흉은 낭종의 파열에 의한 것으로 판단되었다. 전신성 경화증은 폐를 포함한 여러 장기를 침범하는 질환이다. 이 질환에서 기흉과 낭종성 폐병변이 발생하였던 증례들은 1954년 이래로 보고되어 왔는데 모든 증례에서 기흉은 전신성 경화증을 진단받은 뒤에야 발견된 것이었다. 본 증례는 기흉으로 발현한 전신성 경화증의 첫 증례라는 점에서 임상적 의의가 있다고 여겨진다. **(Tuberculosis and Respiratory Diseases 2003, 55:522-525)**

Key words : Systemic sclerosis, Pneumothorax, anti DNA topoisomerase I.

Address for correspondence :
Young Whan Kim, M.D., Ph.D
Division of Pulmonary and Critical Care Medicine, Department of Internal Medicine and Lung Institute of Medical Research Center, Seoul National University College of Medicine
28 Yongon Dong, Chongno Gu, Seoul, 110-744, Korea
Phone : 02-760-2856 Fax : 02-762-9662 E-mail : ywkim@snu.ac.kr

Introduction

Systemic sclerosis is a disorder of the connective tissue that is characterized by the involvement of multiple organs including lungs, and the pulmonary disease is now the leading cause of morbidity and a principal source of mortality[1]. A number of pneumothorax cases with cystic lung lesions in systemic sclerosis have been reported since 1954[2-8]. In all reports, the pneumothorax in each case was detected after being diagnosed as a systemic sclerosis. But in our case, the pneumothorax was the first clinical manifestation and the patient was diagnosed with systemic sclerosis based on her medical history and, positive anti-DNA topoisomerase I, which is known to be correlated with the high prevalence of pulmonary involvement in systemic sclerosis.

Case

A 26 year old Korean female was brought to the emergent room suffering from sudden right pleuritic pain. The right breath sound was decreased, and a pneumothorax was detected in the right chest and a chest tube was inserted. High resolution CT(HRCT) showed multiple cystic lesions in both upper and lower lobes(Fig. 1). The cystic lesions were distributed bilaterally and in both upper and lower lobes, although they are predominant in the base of the lung. There was no evidence of interstitial lung disease in the HRCT. The patient was a non smoker, and the α_1-antitrypsin concentration was within the normal range(338mg/dL). From her medical history, she had noticed Raynaud's

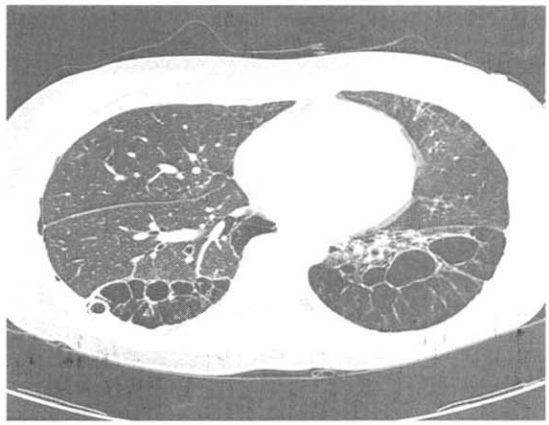

Fig. 1. HRCT shows multiple cystic lesions in both lower lobes. A chest tube is seen in the right pleural space. The cystic lesions are also located in the upper lobes although they are predominant in the lower lobes. There are no interstitial fibrotic lesions found in the lung parenchyme.

phenomenon and a hardening of the fingers since the age of 20. The physical examination showed thin, hard skin on both upper extremities and the forehead (fingers ++/++, hand dorsum +/+, forearm 0/+, forehead +), and a pitting ulcer on the left 3rd finger. The ANA was positive (mixed pattern with a speckled and homogeneous type), the anti DNA topoisomerase I antibody test was also positive. However, the anti-centromere antibody test was negative. A pulmonary function test revealed the presence of a restrictive ventilatory abnormality (FEV_1 1.64L, 49%, FVC 1.97L, 48%, FEV_1/FVC 83%). Several attempts were made to remove the chest tube from her after verifying no air leakage, but the pneumothorax recurred at those times of the clamping chest tube. It was thought that the recurrent pneumothorax might be due to ruptures of multiple bullae. It was recommended

that both lower lobes with multiple lesions should be resected, but this was rejected by the patient. It took 13 days to free the chest tube from her. After a pleurodesis, she was released and is currently being observed in the outpatient department.

Discussion

Systemic sclerosis is a disorder of the connective tissue. It is characterized by a thickening and fibrosis of the skin, and by distinctive forms of organ involvement, notably the heart, lungs, kidneys, and gastrointestinal tract. The extent and severity of the internal organ involvement determines its prognosis. Since the use of ACE inhibitors, pulmonary disease is now the leading cause of morbidity and a principal source of mortality.[1]

Cystic lung lesions in systemic sclerosis has been reported since Dostrovsky reported three cases in 1947.[2-8]

In these cases, cystic involvement is mostly observed bilaterally[2,5,8] and in the lung bases[2,4,6,8]. However, because there are also cases with upper lobe involvement[2,3,7], and computed tomography was not available in most case reports, it is believed that cystic lung lesions can be observed in any lobe of the lung in systemic sclerosis. In our case, as showed in (Fig. 1), the cystic lung lesions were also present in the bilateral upper and lower lobes, although these lesions were prominent in both lung bases.

Since Boyd reported the first case in 1954, there have been 4 cases of systemic sclerosis with cystic lung lesions where a pneumothorax was detected[4-7]. In each of these cases the symptoms, which required hospitalization, differed from each other. In addition, the pnemothorax in those cases were detected after they had been diagnosed as systemic sclerosis. However, our case is different from those cases because the patient had pleuritic pain as the chief complaint and a diagnosis of systemic sclerosis, based on review of the medical history and the laboratory findings, was preceded by the detection of pneumothorax. That is, a pneumothorax was the first clinical manifestation in the diagnosis of systemic sclerosis.

It is believed that these kinds of pneumothorax are formed by a rupture of the cysts, which is a life threatening manifestation. In most cases with cystic lung lesions in systemic sclerosis, a pulmonary interstitial fibrosis is the main lung manifestation. These pulmonary lesions in systemic sclerosis are thought to be the result of inflammation mediated processes. Moreover, recent studies have suggested that reactive oxidant species and proteolytic enzymes injure the connective tissue structures of the alveolar septa and the normal alveolar walls are replaced by thin avascular hyalinized walls, which are prone to rupture. Pulmonary interstitial fibrosis is known to have a higher risk in diffuse systemic sclerosis than the limited type[1]. However, in several cases with limited systemic sclerosis there are fibrotic and cystic lung lesions.

Anti-DNA topoisomerase I is known to correlate with the cutaneous extent (6.5% in limited type vs. 70.6% in diffuse type) and a

high prevalence of pulmonary involvement in systemic sclerosis[9]. On the other hand, anti-centromere antibodies, another antinuclear antibody, have a high correlation with limited systemic sclerosis and a lower correlation with pulmonary involvement[1]. In recent reports[7,8], which were able to measure the level of these autoantibodies, all their 3 cases tested positive to anti DNA topoisomerase I antibodies but tested negative to anti-centromere antibodies. In addition, all were classified as limited systemic sclerosis. Our case was belonged to the limited type but tested positive to anti-DNA topoisomerase I antibodies and negative to anti-centromere antibodies. Yoko et al. thought the lung involvement, which could be specified by cystic lung lesions and fibrosis, had a relationship with the positive anti-DNA topoisomerase I antibody test.[7] Therefore, this possibility was considered in this case.

However, in our case, no definite fibrotic lesions were observed in the lung parenchyme although a pulmonary fibrosis can develop anytime. The patient is currently being followed up to determine whether or not a pulmonary fibrosis develops.

References

1. James RS. Scleroderma and mixed connective tissue diseases. In: Shaun R, Edward DH, Clement BS, editors. Kelly's textbook of rheumatology. 6th ed. Philadelphia: W.B. Saunders company; 1997. p.1211-39.
2. Dostrovsky A. Progressive scleroderma of the skin with cystic sclerodermal changes of the lungs. Archives of Dermatology and Syphilology. 1947;55:1-11.
3. Church RE, Ellis AR. Cystic pulmonary fibrosis in generalized sclerodema. Lancet. 1950;1:392-4.
4. Boyd JA, Patrick SI, Reeves RJ. Roentgen changes observed in generalized scleroderma. Archives of Internal Medicine. 1954;94:248-58.
5. Israel MS, Harley BJS. Spontaneous pneumothorax in scleroderma. Thorax. 1956;11:113-8.
6. Edwards WG, Dines DE. Recurrent spontaneous pneumothorax in diffuse scleroderma, Disease of the Chest. 1966;39:96-8.
7. Yoko K, Masashi A, Shunji Y, Tsuneyo M, Mitsuo H. Large cystic lesions of the upper lobes of the lungs in two patients with CREST syndrome. Arthritis and Rheumatism. 1984;27:935-8.
8. Anette B, Mohammed T. Cystic lung disease in sytemic sclerosis: A case report with high resolution computed tomography findings. Rev. Rhum. 1996;63:213-5.
9. Diot E, Giraudeau B, Diot P. Is anti topoisomerase I a serum marker or pulmonary involevement in systemic sclerosis?. Chest. 1999;116(3):715-20.