Fibrothorax and severe lung restriction secondary to lupus pleuritis and its successful treatment by pleurectomy

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Pleural disease is a common pulmonary manifestation of systemic lupus erythematosus (SLE) that usually responds to corticosteroids and other immunosuppressive agents. In the present report, a new approach, pleural decortication, was used in a patient with medically refractory chronic pleuritis secondary to severe SLE. A 26-year-old woman with known SLE developed progressive dyspnea and pleuritic chest pain over several months. The other systemic manifestations of her lupus were controlled with cyclophosphamide and prednisone. A computed tomography scan revealed a persistent, small, loculated right pleural effusion; pleural thickening; and atelectasis of the right middle and lower lobes. Pulmonary function tests showed a severe restrictive defect. The patient was disabled by her severe dyspnea despite maximal medical therapy, and, therefore, surgery was considered. A right thoracotomy revealed entrapment of the right lung by dense visceral pleura. Decortication was performed. On pathology, pleuritis with vascular pleural adhesions was found. No lupus pneumonitis was noted. Postoperatively, a significant clinical improvement in dyspnea was evident within several weeks. On a 6 min walk test, the patient achieved 384 m with a Borg dyspnea scale rating of 2 compared with 220 m and a Borg dyspnea scale rating of 4 preoperatively. Her forced vital capacity improved from 24% predicted to 47% predicted, and her total lung capacity improved from 35% predicted to 54% predicted. Medical therapy of systemic lupus erythematosus has been proven to be effective in controlling pleuritis in most cases. However, in the event of refractory pleuritis or pleural thickening, decortication may be a viable alternative.

Key Words: *Pleurectomy; Pleuritis; Systemic lupus erythematosus*

Le fibrothorax et la restriction pulmonaire grave secondaire à une pleurésie lupique et son traitement réussi par pleurectomie

RÉSUMÉ : La pleuralgie est une manifestation pulmonaire courante du lupus érythémateux aigu disséminé (LEAD), qui réagit généralement aux corticoïdes et à d'autres immunosuppresseurs. Dans le présent rapport, une nouvelle méthode, la décortication pleurale, a été utilisée chez une patiente présentant une pleurésie chronique réfractaire aux médicaments, secondaire à un LEAD. Une femme de 26 ans atteinte d'un LEAD connu a développé une dyspnée évolutive et des douleurs pleurétiques en l'espace de plusieurs mois. Les autres manifestations systémiques de son lupus étaient contrôlées au moyen de cyclophosphamide et de prednisone. Une tomodensitométrie a révélé la présence d'une petite effusion pleurale loculée droite persistante, un épaississement pleural et une atélectasie des lobes pulmonaires droits moyen et inférieur. L'exploration fonctionnelle respiratoire a démontré une grave défaillance restrictive. La patiente était handicapée par sa dyspnée grave, malgré une pharmacothérapie maximale. C'est pourquoi une intervention chirurgicale a été envisagée. Une thoracotomie droite a révélé la compression chronique du poumon droit par une plèvre viscérale épaissie. Une décortication a été exécutée. À l'examen pathologique, on a découvert une pleurésie accompagnée d'adhérences pleurales vasculaires. Aucune pneumonite lupique n'a été observée. Après l'opération, une importante amélioration clinique de la dyspnée est devenue évidente au bout de quelques semaines. D'après un essai de marche de 6 minutes, la patiente a atteint 384 m et une échelle de dyspnée de Borg de 2, par rapport à 220 m et une échelle de dyspnée de Borg de 4 avant l'opération. Sa capacité vitale forcée est passée de 24 % prévisibles à 47 % prévisibles, et sa capacité pulmonaire totale, de 35% prévisibles à 54 % prévisibles. Il est démontré que la pharmacothérapie permet de contrôler la pleurésie dans la plupart des cas de lupus érythémateux aigu disséminé. Cependant, en cas de pleurésie réfractaire ou d'épaississement pleural, la décortication constitue peut-être une solution pratique.

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Pulmonary involvement may occur in 50% to 75% of patients with systemic lupus erythematosus (SLE) (1,2). The pleura is the most commonly affected site, and pleuritis and pleural effusion are the most common manifestations (3,4).

Most pleural effusions in patients with SLE resolve spontaneously; others require treatment with corticosteroids or a combination of corticosteroids and immunosuppressive agents. Although pleuritis may result in pleural thickening, progressive pleural fibrosis leading to fibrothorax has not been previously reported. A case of fibrothorax secondary to lupus pleuritis and its successful treatment by pleurectomy is reported.

CASE PRESENTATION

A 26-year-old woman presented with symptoms of arthralgia, alopecia, fatigue, weight loss and malar rash at the age of 19 years. Diagnosis of SLE was considered, and confirmed by low complement levels (C4), an antinuclear antibody titre of 1:10,240, and elevated single- and double-stranded DNA levels at 75 and 52 Bethesda units/mL, respectively. She was treated with prednisone and intervenous cyclophosphamide. Over the next one to two years, she developed pleuritic chest pain and exertional dyspnea, which progressed to the point that she became dyspneic at rest. There was no history of paroxysmal nocturnal dyspnea, palpitations, abdominal pain or peripheral edema. A physical examination showed her to be tachypneic at rest, with a respiratory rate of 32 breaths/min. Thoracic examination showed a marked reduction in thoracic excursion, especially of the right hemithorax; the results of a cardiovascular examination was normal.

The results of her pulmonary function tests were forced vital capacity (FVC) 1.06 L/s (24% predicted), forced expiratory volume in 1 s (FEV_1) 0.95 L/s (27% predicted), FEV_1/FVC ratio 90%, total lung capacity 2.11 L (35% predicted), lung diffusing capacity for carbon monoxide 7.8 mL/min/mmHg (29% predicted), maximum inspiratory pressure 194 cm/H_2O and maximal expiratory pressure 104 cm/H_2O. A chest radiograph and computed tomography scan of the thorax revealed small right pleural effusion, pleural thickening and partial atelectasis of the right lower and right middle lobes (Figures 1 and 2). There was no evidence of interstitial lung disease. The 6 min walk test was discontinued because of dyspnea. A walking distance of 220 m was achieved; however, no desaturation was demonstrated.

Because the patient did not respond to medical therapy, surgical options were considered. A right thoracotomy, decortication of right visceral pleura and an open lung biopsy were performed. At the time of the surgery, dense adhesions were seen trapping the right lower lobe; thick visceral pleura limited the expansion of the right upper and middle lobes. A thick plural peel was excised by decortication. The lung biopsy confirmed plural fibrosis, but did not show evidence of lupus pneumonitis. Four months postoperatively, the patient's dyspnea markedly improved. The results of a pulmonary function test showed FVC 2.03 L/s (47% predicted), FEV_1 1.58 L/s (46% predicted), FEV_1/FVC ratio 84%, total lung capacity 3.04 L (51% predicted) and lung diffusing capacity for carbon monoxide 12.5 mL/min/mmHg (47% predicted). The patient achieved 400 m on the 6 min walk test, and there was no desaturation and no dyspnea noted. Since the surgery, the patient has continued to be asymptomatic (Figure 3).

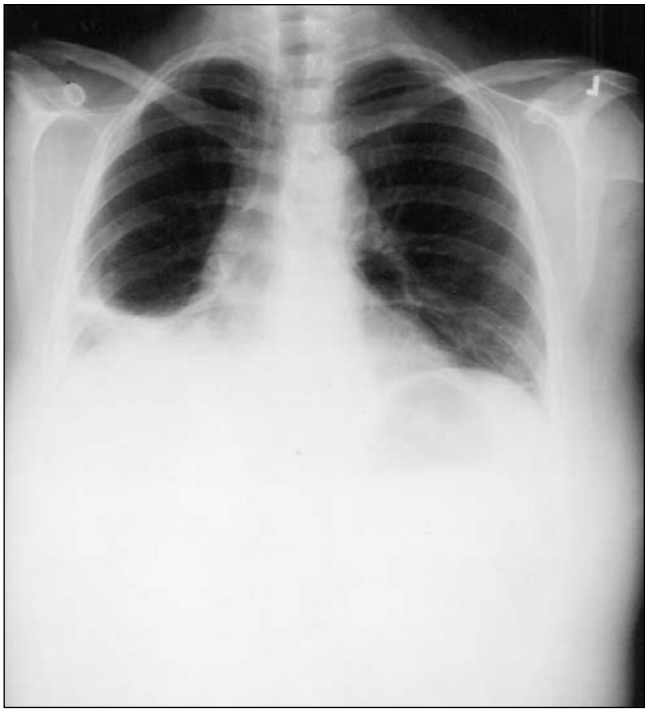

Figure 1) *Chest radiograph showing marked restriction of right lung inflation during inspiration*

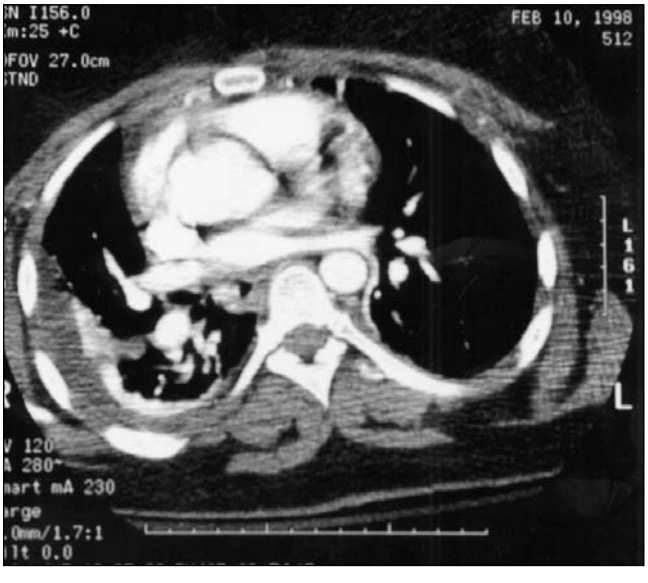

Figure 2) *Computed tomography scan of the chest showing visceral pleural thickening, volume loss and a small pleural effusion*

DISCUSSION

Pulmonary involvement occurs in more than one-half of patients with SLE during the course of their illness. Pulmonary manifestations include infectious pneumonia, pleuritis with or without effusion, acute lupus pneumonitis, alveolar hemorrhage, chronic interstitial pneumonitis or fibrosis, respiratory muscle weakness, pulmonary hypertension, pulmonary thromboembolism and upper airway dysfunction (1,2). The most common site of respiratory involvement in patients with SLE is the pleura. The presence of pleural effusion was noted in 7% of patients with SLE by echocardiography (3) and in 9% of patients with SLE by high resolution computed tomography scan (4). Effusions are typically small to moderate in size and are commonly bilateral, but may be unilateral. Pleural effusions in patients with SLE may resolve spontaneously or, alternatively, may respond well to therapy with corticosteroids (5). On rare occasions, chest tube drainage and pleurodesis with a sclerosing agent may be necessary in a few patients who are unresponsive to systemic anti-inflammatory therapy. Thickening of the visceral pleura has been previously reported in patients with SLE, but is rather rare (6,7). Progressive pleural fibrosis leading to fibrothorax has not been previously described in patients with SLE. We report, for the first time, the treatment of such a complication by decortication.

Bell and Lawrence (6) reported two cases of chronic pleurisy in patients with SLE treated with pleurectomy; these patients had unremitting pleural pain as the dominant symptom. Both of these patients failed conservative medical treatment, but pleurectomy resulted in a significant improvement of chronic pleural pain. In another article, Passero and Myers (8) reported two cases of hemopneumothorax in patients with SLE. Chest tube drainage failed to re-expand the lungs in these patients; therefore, surgical decortication led to the expansion of the collapsed lung and resolution of pneumothorax. Another case report in 1986 described the case of a 34-year-old man who developed recurrent, massive pleural effusions (9). Medical treatment with corticosteroids and azathioprine improved the manifestations of SLE, except for the pleural effusions. They were successfully treated by pleurectomy. In a similar report by Elborn et al (10), refractory massive pleural effusion was treated by pleurectomy. In our patient, severe restrictive pulmonary disease and disabling dyspnea resulted secondary to fibrothorax induced by lupus pleuritis. Aggressive medical therapy with corticosteroids and cyclophosphamide failed to improve the patient's symptoms. A pleurectomy resulted in a marked improvement in lung mechanics and resolution of symptoms.

Pleuritis and pleural effusion are the most common pulmonary manifestations of SLE. Although previously unreported, pleural thickening secondary to SLE may progress to fibrothorax. Previously published case series and the present report establish pleurectomy as a logical and definitive treatment for a variety of pleural disorders that may cause severe disability and are unresponsive to medical therapy.

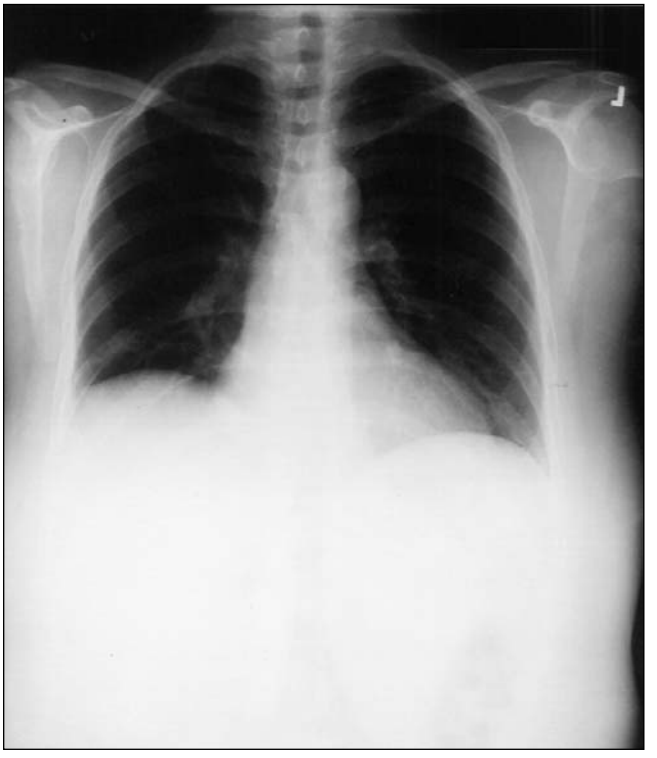

Figure 3) *Postpleurectomy chest radiograph demonstrating a marked improvement in right lung expansion*

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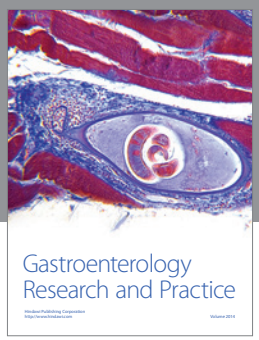

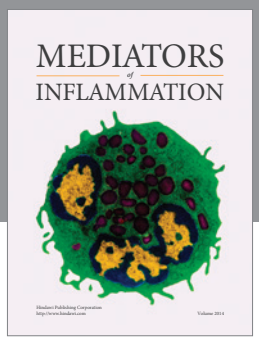

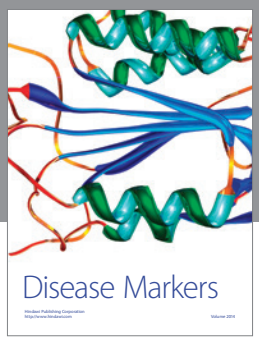

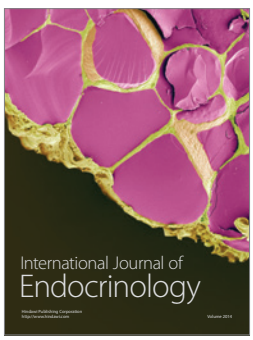

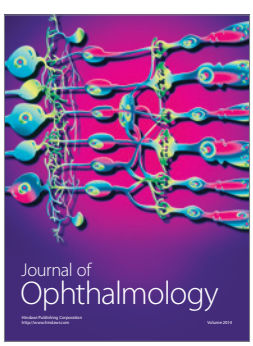

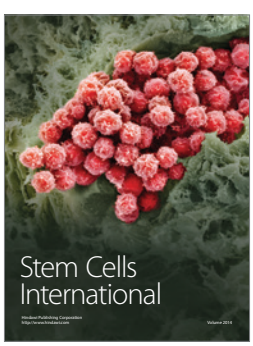

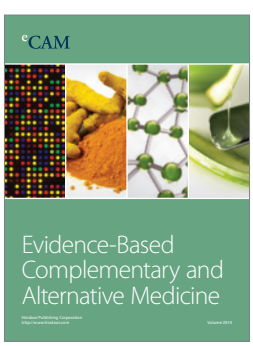

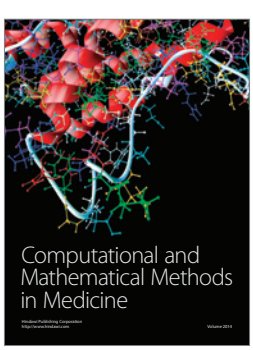

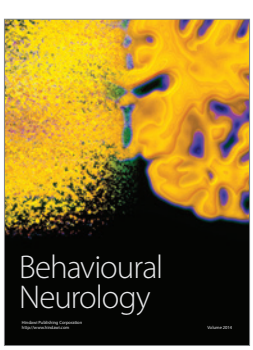

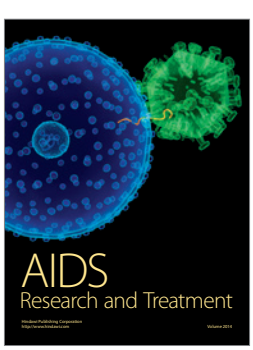

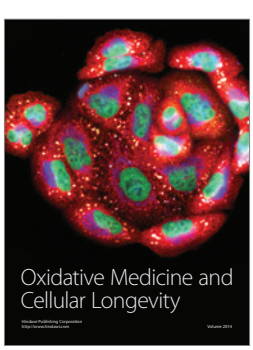

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