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CASE REPORT

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A case of polymyositis complicated with interstitial pneumonitis and pneumomediastinum

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Abstract Pneumomediastinum as a complication of interstitial pneumonia with leakage of air into the mediastinum or subcutaneous tissues is a rare complication of dermatomyositis (DM). Herein we report a case of pneumomediastinum complicating polymyositis (PM), which is usually associated with DM. A 61-year-old man was hospitalized in our department because of deterioration of interstitial pneumonia. Treatment with high-dose corticosteroid and cyclosporin A steadily improved his interstitial pneumonia. Two weeks later, he developed subcutaneous emphysema and chest X-ray showed pneumomediastinum. Both subcutaneous emphysema and pneumomediastinum improved gradually without any additional treatment.

Key words Corticosteroid · Cyclosporin A · Interstitial pneumonitis · Pneumomediastinum · Polymyositis

Introduction

It is widely known that polymyositis (PM) and dermatomyositis (DM) are often accompanied by progressive interstitial pneumonia, which in DM tends to be more resistant to therapy and is sometimes fatal. In some cases of DM, the interstitial pneumonia has been complicated by pneumomediastinum.

We report here the case of a 61-year-old man who suffered from PM that was complicated by progressive interstitial pneumonia. Although the interstitial pneumonia gradually improved with steroids and cyclosporin A therapy, his clinical course was complicated by the appearance of pneumomediastinum. Without any additional therapy, however, the pneumomediastinum gradually disappeared. To our knowledge, this is the first reported case in the English literature of pneumomediastinum complicating interstitial pneumonia in a patient with PM.

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Case report

A 61-year-old man who developed fever, dyspnea, and myalgia in his lower extremities visited a general practitioner in his neighborhood. Chest X-ray showed interstitial changes in both lower lung fields and he was referred for admission to a hospital. After admission, he had progressive dyspnea and rapid deterioration of pulmonary interstitial changes on X-ray despite the administration of antibiotics. As methylprednisolone pulse therapy was initiated intravenously, he was transferred to our hospital for further evaluation and treatment. On initial examination he was afebrile. Cardiac, abdominal, and dermatologic features were normal but coarse crackles were heard in both lower lung fields. On physical examination, there was no joint deformity or cyanosis in his fingers, but he had profound proximal muscle weakness in both lower extremities.

Initial laboratory tests revealed a white blood cell count of 14800/mm^3, elevation of serum C-reactive protein value (4.8 mg/dl), aspartate aminotransferase, alanine aminotransferase, lactate dehydrogenase (LDH), and creatine kinase (CK) (2450 IU/ml) levels, and a low PaO_2 level. The KL-6, SP-D, and SP-A values were also increased, indicating the severity of the interstitial pneumonia.

Antinuclear antibody was negative but anti-Jo-1 antibody level was elevated to 164 U/ml. Chest X-ray showed a reticulonodular pattern involving mainly the middle and lower lobes, and chest computed tomography. (CT) scan showed ground-glass opacity which had progressed rapidly compared with the former CT. Transbronchial lung biopsy specimen showed inflammatory changes in the pulmonary interstitium infiltrated mainly with lymphocytes, without formation of hyaline membrane or granuloma (Fig. 1), which was consistent with nonspecific interstitial pneumonia (NSIP). Muscle biopsy showed changes typical for myo-

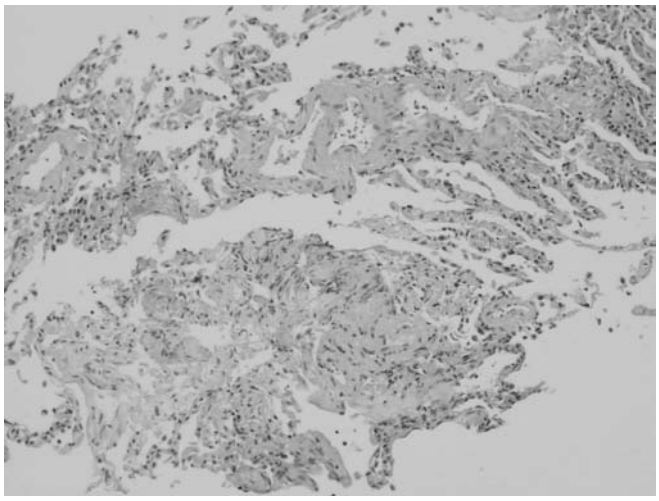

Fig. 1. Pulmonary interstitium is infiltrated with lymphocytes without formation of hyaline membrane or granuloma (H&E, ×20). Obtained from transbronchial lung biopsy on October 30, performed in former hospital

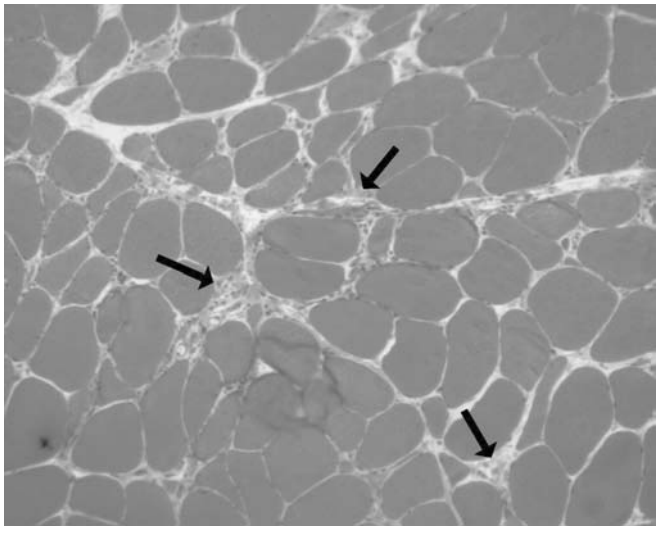

Fig. 2. Biopsy specimen of the left biceps reveals prominent infiltrates of mononuclear cells around small vessels (*arrows*) and muscle fibers showing difference in size (H&E, ×500)

sitis, including infiltration with lymphocytes and difference in size of muscle fibers due to degeneration (Fig. 2). From these findings, including weakness of proximal muscles, elevation of muscle-related enzymes, anti Jo-1 antibody, findings from transbronchial lung biopsy, and muscle biopsy, we diagnosed him as having PM accompanied by interstitial pneumonia. Prednisolone (PSL) 60 mg and cyclosporin A (CsA) 50 mg per day were commenced initially and CsA was increased up to 100 mg/day. Although his chest X-ray findings and values of creatine phosphokinase, LDH, and KL-6 had rapidly normalized within a few days, 14 days after initiating treatment the patient noticed subcutaneous emphysema and chest X-ray and CT showed pneumomediastinum (Fig. 3). Without any additional

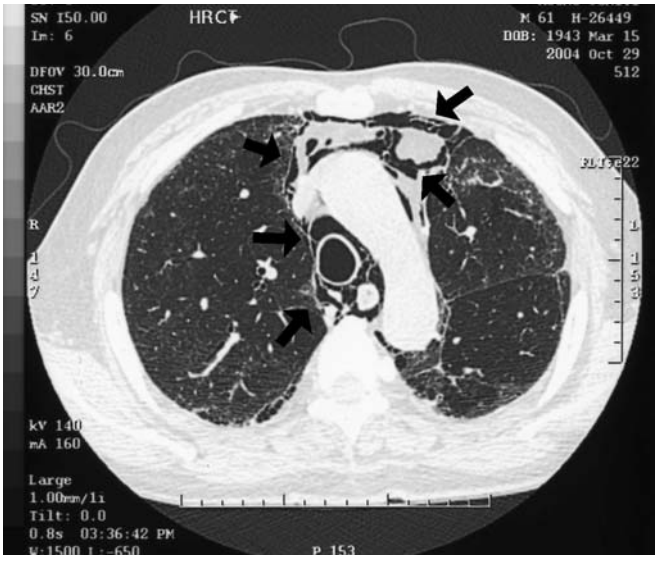

Fig. 3. Chest computed tomography performed 14 days after initiating treatment on November 8 shows diffuse pneumomediastinum (*arrows*)

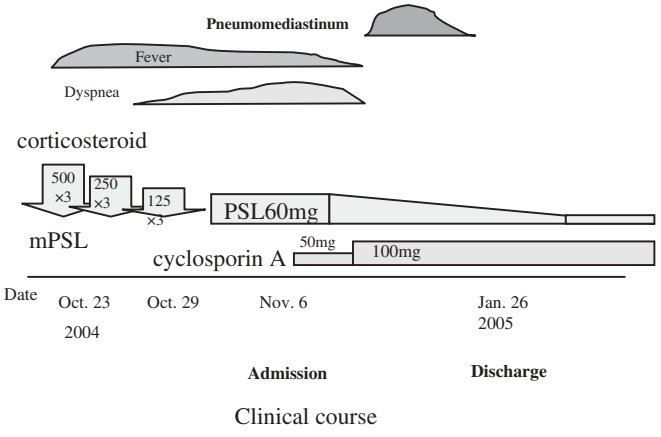

Fig. 4. Clinical course. *PSL*, prednisolone; *mPSL*, methylprednisolone

therapy other than keeping him at rest, the subcutaneous emphysema and pneumomediastinum gradually disappeared, and the interstitial pneumonia and muscle symptoms also gradually improved on treatment. Prednisolone had been tapered down to 20 mg/day when he was discharged on the 30th day following admission. He is now treated as an outpatient and the PSL dose has been decreased to 10 mg a day (Fig. 4).

Discussion

The frequency of interstitial pneumonia as a complication of DM and PM is reported as 5%–42% worldwide.[1,2] According to another report surveyed in Japan, 58.1% of DM and 54.6% of PM were complicated by interstitial pneumonia. Due to the poor prognosis of interstitial pneumonia in these cases, thorough evaluation and careful follow-up of

these patients are very important. Rapid progression of interstitial pneumonia had often been reported in DM but only occasionally reported in PM.[3] In these cases, pulmonary interstitial infiltration with lymphocytes was seen in the biopsy specimen of the lung and most of them were diagnosed as NSIP, which was similar to our case. On the other hand, pneumomediastinum is a rare complication that had been reported mainly in DM up until now. Pneumomediastinum reported in a series of DM cases tended to be resistant to conventional therapy and 56% of those cases were fatal.[4] The mechanism of pneumomediastinum was thought to be the result of air bursting from the alveolus and spreading through the neighboring tissues along the pulmonary vessels toward the mediastinum.[5]

Kono et al. described in their series of pneumomediastinum in DM[6] that the bursting of the alveolus might have some connection with the existence of vasculitis because the pneumomediastinum cases tend to have severe skin lesion manifestations. Other than DM, pneumomediastinum can develop not only in connective tissue diseases such as polyarteritis and systemic lupus erythematosus, but can also occur in traumatic and metabolic disorders. These cases are not necessarily accompanied by skin lesions, so the severity of the pulmonary involvement might directly account for the occurrence of pneumomediastinum. The reason our case developed pneumomediastinum may have been mechanical disruption or severe pulmonary involvement.

There is an alternative view concerning DM with pneumomediastinum. Satomi et al. showed that in fatal cases of DM with pneumomediastinum in Japan, their CK level was normal or slightly higher and anti-Jo-1 antibody was negative.[4] In contrast, improved cases showed elevation of myogenic enzymes, such as CK or creatine and anti-Jo-1 antibody titer.

In most of the fatal cases, pneumomediastinum occurred concurrently with exacerbation of interstitial pneumonia, whereas in improved cases, development of pneumomediastinum was delayed until after the improving stage of interstitial pneumonia. The difference in laboratory features and clinical course between fatal cases and improved cases could possibly indicate an essentially different pathological process, even though both groups had pneumomediastinum in DM.

The favorable outcome in our patient might be attributed to having good prognostic factors such as elevation of both myogenic enzyme and anti-Jo-1 antibody titer. Because of the rapid progression of his pulmonary involvement, we administered CsA as initial therapy. Cyclosporin A was reported as a good choice of treatment for interstitial pneumonia.[7,8] There is also a report of DM complicated with pneumomediastinum that was successfully treated with CsA.[9] Initiating treatment with CsA might have brought about the favorable effect in our patient.

We think it is noteworthy that pneumomediastinum has occurred as a complication of PM. This report might lead to the identification of other cases in the future.

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