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□ Case Report □

A Case of Dermatomyositis Complicated with Pneumomediastinum Successfully treated with Cyclosporine A: a Case Report and Review of Literature

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Dermatomyositis (DM) is a systemic inflammatory disease affecting skeletal muscles and other organs. Spontaneous pneumomediastinum (PnM) has been previously reported as a rare complication of DM and it is known to occur more frequently in patients with interstitial lung disease (ILD). Here we report on a case of a 52-year-old woman with DM who developed spontaneous PnM, which was treated successfully with high-dose steroid pulse therapy and cyclosporine A (CsA). This case suggests that CsA can be an effective therapeutic agent in DM refractory to glucocorticoid therapy, with ILD or pulmonary fibrosis accompanied by DM. CsA should be considered as an initial immunosuppressive agent for patients with PnM in DM.
Key Words. **Dermatomyositis, Pneumomediastinum, Interstitial lung disease, Cyclosporin**

Introduction

Dermatomyositis (DM) is a systemic inflammatory disease affecting skeletal muscles and other organs. In 10～22% of cases, pulmonary signs are related to aspiration pneumonia or respiratory muscle weakness (1). Clinically apparent respiratory diseases developed in up to 50% of patients with DM and interstitial lung disease (ILD) is present in 10～43% of cases and are associated with a poor outcome (1,2). Spontaneous pneumomediastinum (PnM) has been previously reported as a rare complication of DM and it is known to be more frequent in patients with ILD (3). Spontaneous PnM in DM patient is life-threatening, and aggressive high-dose steroid pulse therapy with immunosuppressive agents is recommended. Here we report a case of a 52-year-old woman with DM who developed spontaneous PnM which was successfully treated with high-dose steroid pulse therapy and cyclosporine A (CsA).

Case Report

A 52-year-old woman visited our hospital with complaints of general weakness, swallowing difficulty, arthralgia, and skin erythema for 5 months. There were periungual erythema (Figure 1A) and typical Gottron's sign on her knuckles of the proximal interphalangeal joints (Figure 1B) and extensor area of her elbows. On physical examination, muscle strengths of her extremities were lowered to grade III. No Raynaud phenomenon was shown. Laboratory studies showed hemoglobin of 10.0 g/dL (12～16 g/dL), white blood cell count of 5,630 /mm^3 (4,800～10,800 /mm^3), platelet count of 305×10^3 /mm^3 (130～450×10^3 /mm^3). Blood chemistry showed glutamate-oxaloacetate transaminase/glutamate-pyruvate transaminase of 84/38 IU/L (12～33/5～35 IU/L), creatine kinase level of 225 IU/L (50～200 IU/L), lactate dehydrogenase of 1,229 IU/L (218～472 IU/L), and aldolase of 14.6 IU/ml (＜7.6 IU/mL) Antinuclear antibody was positive with titers of 1 : 20 with homogenous pattern. Rheumatoid factor, anti-SS-A, anti-SS-B, anti-Sm, anti-RNP, anti-Scl-70, and anti-Jo-1 antibody were

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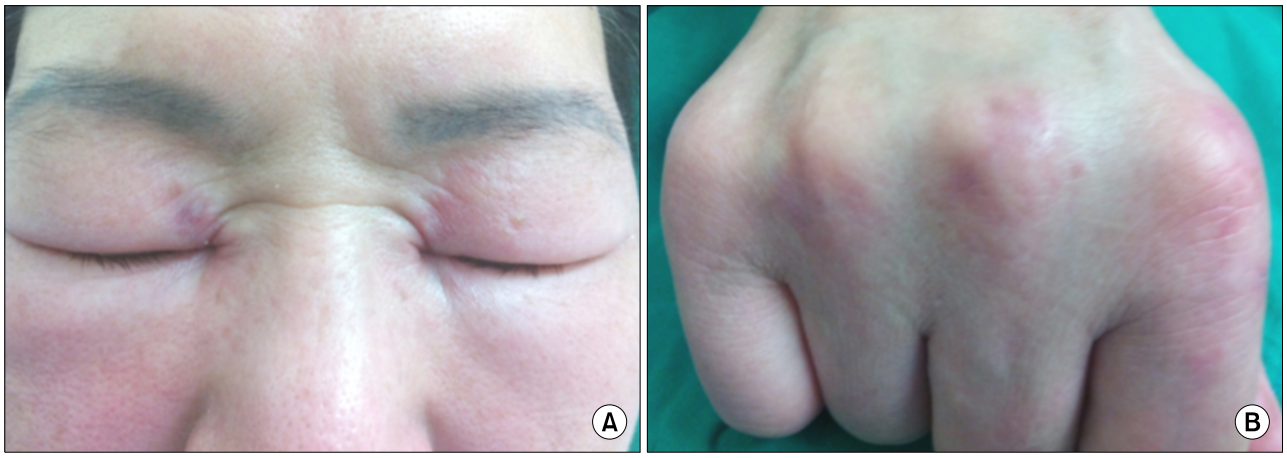

Figure 1. Periungual erythema (A) and Gottron's sign (B).

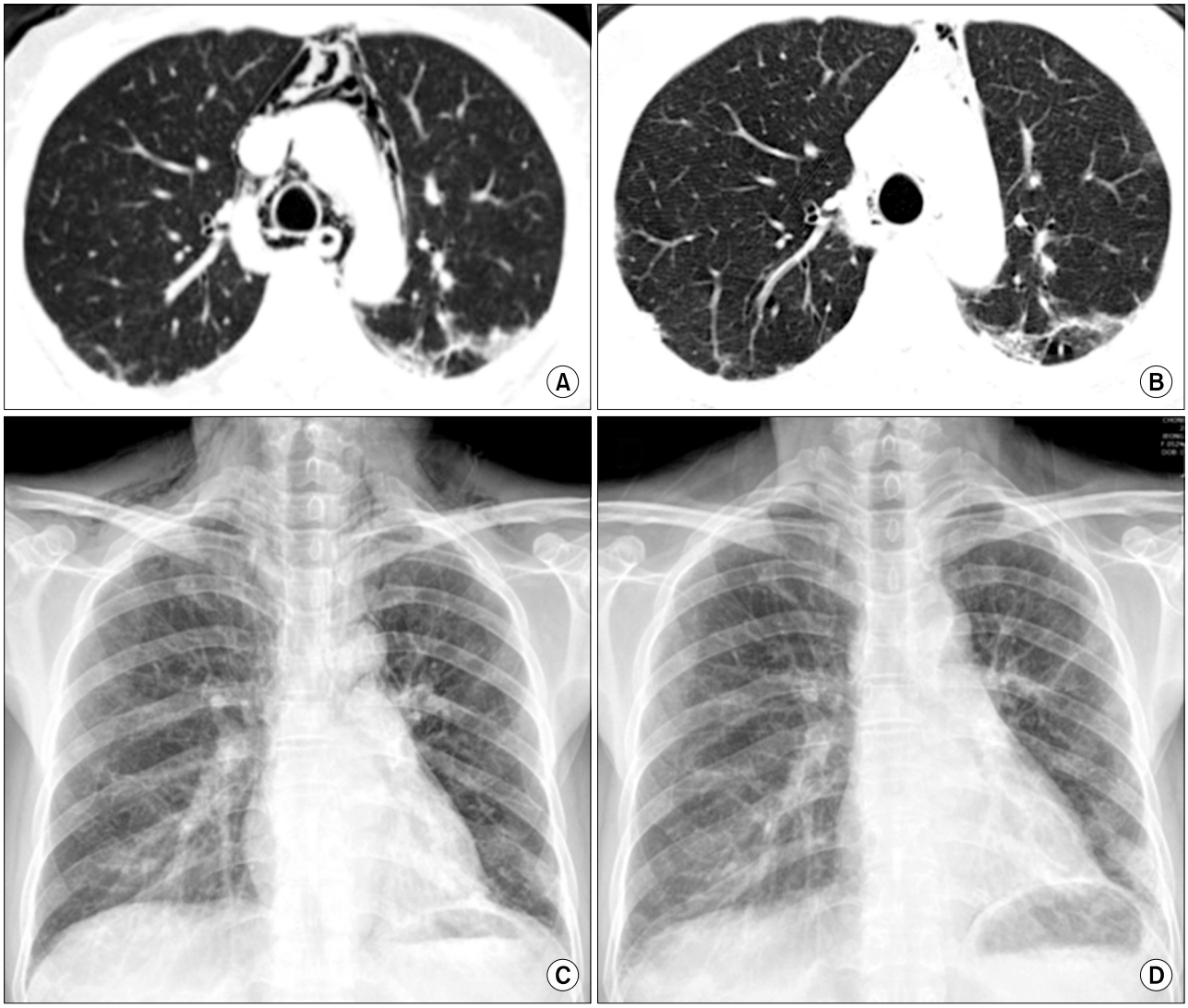

Figure 2. Spontaneous pneumomediastinum (A) with subcutaneous emphysema (C) and resolved state (B and D) after the treatment with high dose steroid and cyclosporine.

all negative. She was diagnosed with DM based on proximal muscle weakness, cutaneous manifestations, elevated muscle enzymes, and positive electromyography. On high resolution computed tomography, it showed subpleural reticular pattern density with traction bronchiectasis, honeycombing predominatly on lower lobes of both lung. Pulmonary function test on admission showed FEV1/FVC 90%, FEV1 1.87 L (89%), FVC 2.07 L (74%), TLC 3.2 L (77%), DLco 9.1 mL/mmHg/min (52%). No echocardiography was done. Treatment began with high-dose oral prednisolone (1 mg/kg/day) and azathioprine (100 mg/day). Creatine kinase lowered on one week laboratory follow-up from the admission day (225 → 79 IU/L), but skin lesions and muscle weakness didn't get improved with onging hair loss. Two weeks later, she complained of sudden chest discomfort and dyspnea. On physical examination, a subcutaneous crepitus around neck was detected and PnM was diagnosed by X-ray finding of subcutaneous emphysema with free air along cardiac border (Figure 2A and 2C). High-dose steroid pulse therapy (1 g of IV methylprednisolone) was started with oral CsA (100 mg twice a day). High dose oxygen supply and absolute bed rest were ordered. After treatments described above, subcutaneous emphysema and PnM gradually disappeared (Figure 2B and 2D), and muscle weakness was also improved. Prednisolone was tapered and the patient discharged for the outpatient department.

Discussion

DM is an inflammatory connective tissue disease of unknown etiology with onset most commonly in the fifth decade of life characterized by inflammatory involvement of muscle and typical skin manifestations such as violaceous rash over extensor aspects of interphalangeal joints, elbows, and knees (Gottron papules); heliotrope discoloration of the eyelids, of-

Table 1. Review of dermatomyositis with spontaneous pneumomediastinum

No	References	Year	Sex/Age	Anti-Jo1 Ab	ILD	Skin lesion	Management after PnM	Outcome
1	Bradley et al. (3)	1986	M/42	NA	NA	Yes	MPD+AZA	Resolution
2	Kobayashi et al. (16)	1989	49/F	NA	Yes	Yes	High dose PDS+mediastinal drainage	Death
3	Matsuda et al. (16)	1993	41/F	NA	Yes	(−)	Pulse MPD+PDS+AZA	Death
4	Santos et al. (16)	1995	41/F	NA	Yes	(−)	PDS+AZA	Death
5	Santiago et al. (11)	1998	M/10	(−)	Yes	NA	Pulse MPD+HCQ	Death
6	JH Park et al. (9)	1999	56/F	(−)	Yes	Yes	O2 therapy	Death
7	HY Lee et al. (17)	2000	36/F	(−)	Yes	Yes	MPD+HCQ+cyclophosphamide	Death
8	Kono et al. (4)	2000	M/30	(−)	Yes	Yes	Pulse MPD+CsA	Resolution
9	Kono et al. (4)	2000	M/25	NA	Yes	Yes	Pulse MPD+CsA+AZA	Death
10	Kono et al. (4)	2000	M/23	NA	Yes	Yes	Pulse MPD	Resolution
11	Kono et al. (4)	2000	F/59	NA	Yes	(−)	Pulse MPD	Resolution
12	Nonomura et al. (8)	2001	M/39	NA	Yes	NA	CsA	Resolution
13	Kuroda et al. (8)	2003	M/46	(−)	Yes	No	PDS+CsA	Resolution
14	Bin Yoo et al. (14)	2006	F/40	(−)	Yes	Yes	MPD+IVIG+CsA	Resolution
15	Terao et al. (8)	2007	M/16	(−)	Yes	Yes	PDS+CsA	Resolution
16	HY Lee et al. (17)	2007	45/F	(−)	Yes	Yes	Pulse MPD+cyclophosphamide	Death
17	Powell et al. (11)	2007	M/34	(−)	Yes	NA	Pulse MPD+IVIG	Death
18	Park et al. (11)	2009	F/35	(−)	Yes	Yes	PDS+AZA+cyclophosphamide	Resolution
19	Yoo et al. (8)	2009	F/38	(−)	Yes	Yes	MPD+CsA	Resolution
20	Masrouha et al. (12)	2009	M/66	NA	Yes	NA	Pulse MPD	Resolution
21	Miyazaki et al. (11)	2011	M/29	(−)	Yes	NA	Pulse MPD+CsA	Resolution
22	Miyazaki et al. (11)	2011	F/41	(−)	Yes	NA	Pulse MPD+CsA+cyclophosphamide+IVIG	Resolution
23	Miyazaki et al. (11)	2011	F/43	(−)	Yes	NA	Pulse MPD+CsA+cyclophosphamide	Resolution
24	Hutchinson et al. (13)	2010	F/25	(−)	Yes	Yes	Rituximab+MPD+cyclophosphamide	Resolution
25	Kim et al. (15)	2010	F/59	(−)	Yes	(−)	Pulse MPD+CsA	Resolution
26	Kim et al. (15)	2010	F/51	(−)	Yes	(−)	Pulse MPD+CsA	Resolution
27	Kim et al. (15)	2010	F/42	(−)	Yes	(−)	Pulse MPD+CsA	Resolution
28	Kim et al. (15)	2010	F/43	(−)	Yes	(−)	PDS+HCQ+IVIG+CsA	Resolution
29	Kim et al. (15)	2010	F/44	(−)	Yes	(−)	PDS+CsA	Resolution
30	Kim et al. (15)	2010	F/54	(−)	Yes	(−)	PDS+IVIG+Tacrolimus	Resolution
31	Present case	2013	F/52	(−)	Yes	Yes	Pulse MPD+CsA	Resolution

AZA: azathiorpine, CsA: cyclosporin A, HCQ: hydroxychloroquine, IVIG: intravenous immunoglobulin, MPD: methylprednisone, PDS: prednisolone.

ten with periorbital edema; macular erythema of posterior shoulder and neck; and hyperpigmentation, hyperkeratosis and fissuring and scaling of hands (mechanic hand). Extramuscular manifestations may be present to a varying degree in patients with DM, including systemic symptoms, joint contractures, dysphagia, gastrointestinal symptoms, and cardiac and pulmonary dysfunctions.

It has been estimated that clinically apparent lung disease develop in up to 50% of patients after diagnosis (1,2). Common patterns of respiratory involvement include ILD, bronchiolitis obliterans, organizing pneumonia, diffuse alveolar damage, aspiration pneumonia from esophageal dysfunction, infectious pneumonia caused by drug-induced immunosuppression, pneumonitis or pulmonary fibrosis caused by therapy with methotrexate or cyclophosphamide, atelectasis due to respiratory muscle weakness, lung cancer, pulmonary edema caused by DM-induced congestive cardiomyopathy and very rarely PnM.

Pneumothorax, PnM, and subcutaneous emphysema are rare complications of DM-related ILD, and carry a poor prognosis. It has been showed that subpleural blebs rupture, resulting air dissection around perivascular sheaths, through the mediastinum, and into the pleural space and subcutaneous tissue planes. It has been suggested that DM vasculopathy may cause subpleural infarctions and interstitial emphysema (4). These complications have a special relationship with DM as they occur less commonly in patients with other connective tissue-related lung diseases.

The risk factors for the PnM in patients with DM include ILD (5,6), cutaneous vasculopathy (4), mild or absent CK elevation (4,7), young age (4), and steroid treatment (4). Our patient had features of periungual erythema and typical Gottron's sign on her knuckles as cutaneous vasculopathy, ILD shown in HRCT, and previous steroid therapy before development of PnM.

DM complicated with PnM manifesting similar characteristics as mentioned above have been rarely reported (8-11,14,15). Furthermore, DM-associated pneumothorax presents difficult therapeutic challenges. A chest tube may fail to reexpand the lung because of decreased lung compliance from ILD. Persistent air leaks commonly occur. Despite that maintenance therapy with steroid has been reported as possible cause of PnM or pneumothorax in DM patients (16), high dose prednisolone and methylprednisone pulse therapy were generally adopted as baseline management for DM complicated with PnM 2 decades ago, most of the DM cases with PnM or pneumothorax showed fatal outcomes (Table 1). Recently, CsA has been reported to be an effective treatment for DM complicated with PnM. After CsA was actively applied to DM patients with spontaneous PnM in early phase, the clinical courses of them have been significantly improved (Table 1).

Summary

We herein reported a rare case of a patient with DM, who developed PnM and who was treated with CsA. This case suggests that CsA can be an effective therapeutic agent in DM with ILD or pulmonary fibrosis refractory to glucocorticoid therapy. CsA should be considered as an initial immunosuppressive agent for patients with PnM in DM.

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