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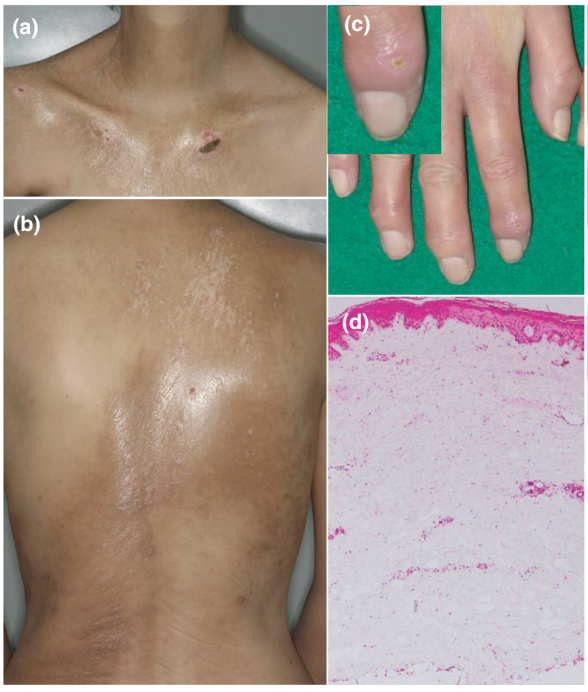
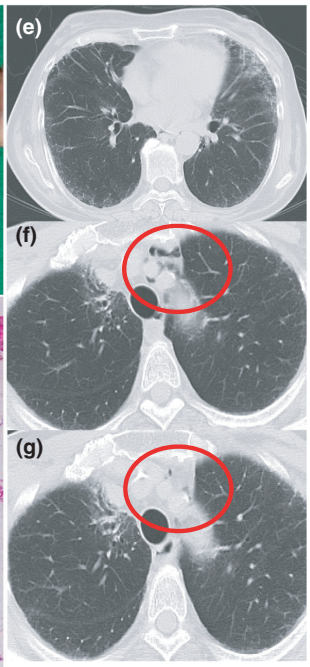
LETTER TO THE EDITOR

Spontaneous pneumomediastinum developed after steroid pulse therapy in diffuse cutaneous systemic sclerosis patient: A case report

Dear Editor,

A 59-year-old Japanese woman was referred to our department with a 4-month history of rapidly progressing skin sclerosis and dyspnea. She was diagnosed with breast cancer 11 years prior, and had tumor resection with radiotherapy. Letrozole and palbociclib therapy were commenced 6 months prior due to multiple metastasis. Physical examination revealed diffuse scleroderma with well-defined shiny plaques of thickened skin on the trunk (Figure 1a,b). The total skin thickness score was 43. Digital ulcers and sclerodactyly were also observed (Figure 1c). Abnormality in the laboratory data included white blood cell count (2900/μL), hemoglobin level (6.9 g/dL), C reactive protein (0.22 mg/dL), creatinine (1.23 mg/dL), KL-6 (854 U/mL). Anti-nuclear antibody and anti-RNA polymerase III antibody (860 U/ml) were positive. A skin biopsy showed thickened homogenous collagen fibers in the dermis (Figure 1d). Based on these findings, a diagnosis of cutaneous systemic sclerosis (SSc) with morphea-like skin lesions was made.





(h) Characteristics of systemic sclerosis cases copmlicared by pneumomediastinum[1-5]

Age (mean ± SD)	29 - 61 years (48.3 ± 13.3 years)
Sex (n=6)	3 male/3 female
SSc duration (mean ± SD)	0 - 8 years (4.6 ± 5.4 years)
SSc subtype	2 diffuse/2 limited
Autoantibody profile	
Anti-topoisomerase 1 Abs	2/5 cases
Anticentromere Abs	1/5 cases
Anti-RNA polymerase III Abs	2/5 cases
Clinical feature	
digital ulcer or digital pitting scar	4/5 cases
interstitial pneumonia	6/6 cases
gastrointestinal involvement	5/6 cases
Previous steroid therapy	5/6 cases
Dyspnea	3/6 cases
Subcutaneous emphysema	4/5 cases
Conservative therapy	6/6 cases
Cure	5/6 cases

FIGURE 1 (a–c) shows the clinical characteristics of the skin lesions; (a and b) show sclerotic shiny skin, with hypo- and hyper-pigmented areas on the trunk; (c) shows the dorsal side of the distal interphalangeal joint, with sclero-edematous fingers and ulcer formation; (d) shows histopathology of the skin from the forearm. Proliferated and hyalinized collagen bundles with inflammatory cells infiltration around atrophic sweat glands embedded in collagen bundles are seen using hematoxylin and eosin (original magnification: ×40). The image is stained by hematoxylin-eosin. (e) High-resolution computed tomography (CT) of the chest shows reticulation and ground-glass opacity in the subpleura. (f) CT of the chest shows pneumomediastinum in the anterior mediastinum. (g) CT image shows a decrease in air volume one week after the conservative therapy started. (h) Summary of the published SSc cases complicated by spontaneous pneumomediastinum, as well as the present case. Abs, antibodies; SD, standard deviation; SSc, systemic sclerosis

A high-resolution computed tomography (HRCT) of the chest revealed new reticulation and ground-glass opacity in the sub-pleural area were noted (Figure 1e). She was treated with methylprednisolone 500 mg pulse therapy for subacute progressive interstitial pneumonia and rapidly progressing skin sclerosis with dyspnea, followed by oral prednisolone (PSL) 30 mg (0.6 mg/kg). A follow-up HRCT performed 2 weeks later revealed free air in the mediastinum (Figure 1f); hence, she was diagnosed with pneumomediastinum (PNM) without any history of trauma or new respiratory symptoms. The patient was given bed rest, PSL was tapered to 20 mg, and continuous positive airway pressure (CPAP) used for sleep apnea syndrome was discontinued. After one week, most of the free air had disappeared (Figure 1g).

Pneumomediastinum is associated with an interstitial lung disease (ILD) due to collagen diseases especially dermatomyositis (DM). However, cases of PNM coexisting with SSc are extremely rare. Only six cases have been reported to date. The five previous cases described in detail and the present case are summarized in Figure 1h.[1–5] There were no tendencies in subtypes and autoantibody profile. All the patients had ILD. Except for one patient whose PNM was caused by pneumatosis cystoides intestinalis,[3] they all recently had a lung disease. Five patients had a history of steroid administration.[2–4] In patients with DM, relations between weakened alveolar walls due to corticosteroids and development of PNM has been pointed. Steroid administration, including pulse therapy, may be involved in the PNM developed in SSc cases. Three of the patients had no respiratory symptoms of PNM.[1,3] All patients were managed conservatively, and five of them were cured. One patient developed respiratory failure and septic shock.[5]

Radiation therapy for lung cancer can result in PNM development. In our patient, palbociclib administration may have partially resulted in ILD development, and CPAP may have contributed to the onset of PNM. However, to the best of our knowledge, there are no reported cases of PNM induced by tangential radiation therapy for breast cancer, palbociclib administration or CPAP for sleep apnea syndrome in adults. PNM possibly develops in SSc-ILD cases when specific risk factors are present.

CONFLICT OF INTEREST

None declared.

Yukie Endo
Jain Kim
Akihiko Uchiyama
Masahito Yasuda
Kenichiro Hara
Sei-ichiro Motegi

Department of Dermatology, Gunma University Graduate School of Medicine, Maebashi, Japan

Correspondence
Sei-ichiro Motegi, Department of Dermatology, Gunma University Graduate School of Medicine 3-39-22, Showa, Maebashi, Gunma 371-8511, Japan.
Email: smotegi@gunma-u.ac.jp

ORCID

Yukie Endo https://orcid.org/0000-0003-3058-6279
Jain Kim https://orcid.org/0000-0001-8225-2743
Akihiko Uchiyama https://orcid.org/0000-0002-2169-2427
Masahito Yasuda https://orcid.org/0000-0002-2907-640X
Sei-ichiro Motegi https://orcid.org/0000-0001-8286-0669

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