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Case Report

Pulmonary Erdheim-Chester disease: A response to predonisolone

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ABSTRACT

Erdheim-Chester disease (ECD) is a rare non-Langerhan's cell histiocytosis of unknown origin, involving multiple organs. The patient with ECD described here is a 38-year-old man who was admitted to the hospital with dyspnea on exertion. His chest radiograph revealed a diffuse reticulonodular shadow. After the video-assisted thoracoscopic surgery was performed, he was diagnosed as having ECD. A brown eruption on his left temple, when tested by skin biopsy, proved to be ECD. No lesions other than these on the lung and skin were identified, and oral administration of predonisolone successfully treated both of them. Although recovery has followed the administration of predonisolone and chemotherapy for several patients with pulmonary ECD, this is the first report that predonisolone alone provided clinical and objective recovery from pulmonary ECD. This outcome indicates that, of all the many treatments tried for ECD, steroids may become the first-line therapy for pulmonary involvement.

1. Introduction

Erdheim-Chester disease (ECD), which was first described by William Chester in 1930,[1,2] still has no known cause. The most commonly affected organs in ECD are the long bones, but more than 50% of the patients suffer extraskeletal involvements, including orbits, pituitary glands, retroperitoneal spaces, skin, lungs, kidneys and hearts.[1,3,4] The prognosis of this disease is often poor when extraskeletal pathology results in cardiomyopathy, respiratory or renal failure and central nerve involvement.[2,5,6] Various treatments have been tried, such as corticosteroids, interferon, chemotherapy (cyclophosphamide, vinblastine, adriamycin, etoposide, etc.), cyclosporine, radiotherapy and surgical resection.[1,7,8] However, no standard treatment for ECD has been established as yet. In addition, owing to the rarity of ECD patients whose organ involvement is limited mainly to the lungs, few treatments for pulmonary ECD have been reported in the literature. Accordingly, neither the most effective therapy for pulmonary involvement nor which type should be initiated first has been established.

In this report, we describe a 38-year-old man with ECD limited mainly to the lungs. Oral administration of predonisolone relieved dyspnea on exertion and improved pulmonary function as documented in radiological findings. This successful outcome suggests that steroid is an effective first-line treatment for early-phase pulmonary ECD.

2. Case report

A 38-year-old man undergoing his regular health examination in July 2007 had a chest radiograph that showed an abnormal shadow. Within a few months, he began to feel dyspnea that gradually worsened on exertion. He was admitted to our hospital for evaluation on April 2008. His history noted that he worked as a plumber and had smoked 30 cigarettes daily for 20 years until he stopped because of the dyspnea. The only abnormality recorded during his physical examination was a brown swollen eruption on the temple (Fig. 1A). However, the chest radiograph showed diffuse reticulonodular shadows that were distributed principally in the bilateral lower lung fields (Fig. 2A). Computerized tomography (CT) of the patient's chest revealed thickening of the pleura, interlobular septa and peribronchovascular interstitial markings as well as fine nodules and small cysts scattered predominantly in the bilateral lower lobes (Fig. 2B,C). No swelling of lymph nodes was evident in the mediastinum. Laboratory tests showed a slight elevation in C-reactive protein (0.6 mg/dL) and soluble IL-2 receptor (925 U/mL). Arterial blood gas analysis was normal. The pulmonary function tests showed reductions in vital capacity (VC) of 3.1 L (75.1% of

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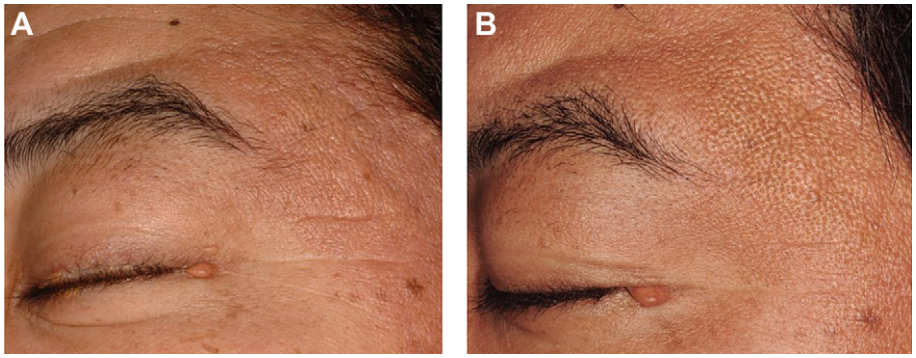

Fig. 1. Skin involvement identified in the left temple. Brown maculae and elastic hard eruption were identified on the left temple prior to the administration of predonisolone (A). The lesion gradually softened and flattened within several months after the initiation of predonisolone treatment (B). Fig. 1B is a matching photo taken a year and two months after the treatment began.

the predicted), forced expiratory volume in 1 s (FEV_1) of 2.01 L (68.1% of the predicted) and diffusing capacity for carbon monoxide (DLCO) of 11.1 mL/min/mmHg (39.9% of the predicted). His 6-min walking test revealed the desaturation of SpO_2 from 96% to 88% with a substantial walking distance of 427 m.

Sarcoidosis, chronic hypersensitive pneumonia, Langerhans' cell histiocytosis and lymphoproliferative disorder were considered as differential diagnoses. Bronchoalveolar lavage fluid contained an increased percentage of lymphocytes (28%) and an elevated CD4/CD8 ratio (6.3). Transbronchial lung biopsy was performed twice, but the histopathological examination of a tissue specimen showed only a slight increase of collagen fibers in part of the alveolar wall. Because sarcoidosis was suspected, a skin biopsy of the brown eruptions on the left temple was performed; xanthomatous changes were found but no evidence of epithelioid cell granuloma. To complete the diagnosis, video-assisted thoracoscopic surgery was performed. Subsequent histopathological examination revealed fibrous thickenings of the pleura and the interlobular septa along with marked infiltration of foamy histiocytes in the fibrotic areas (Fig. 3A). Elastica van Gieson staining of the resected specimen demonstrated that

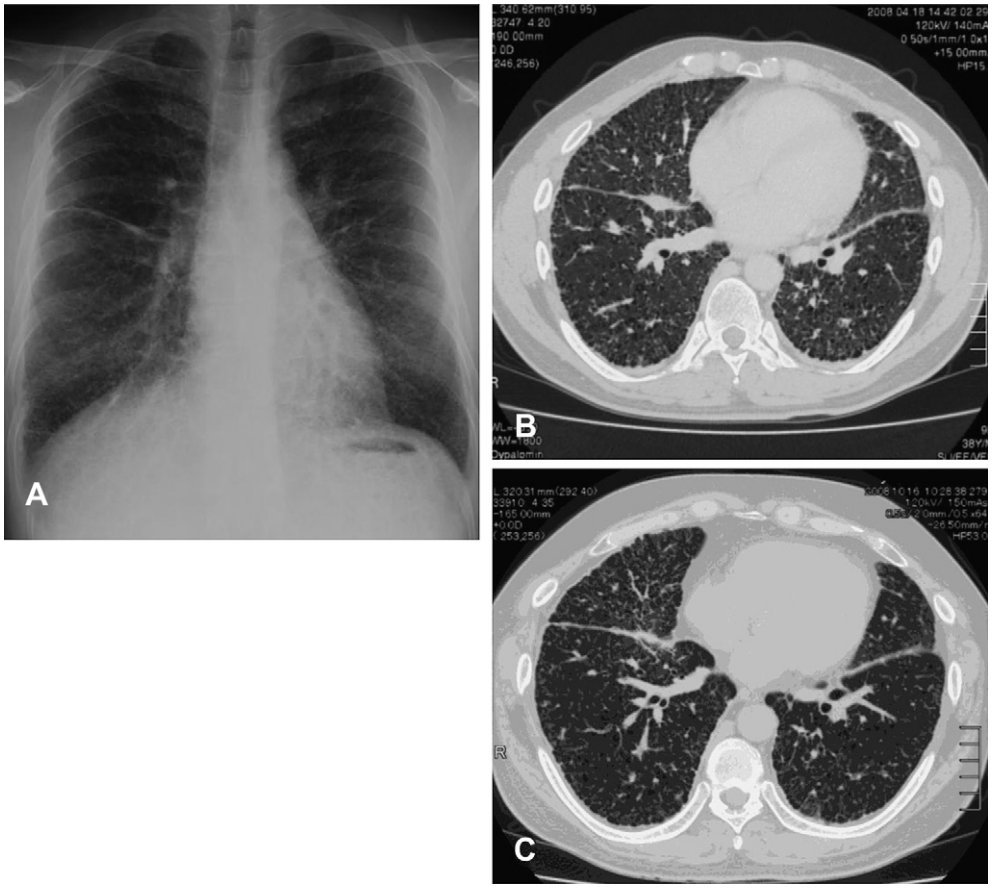

Fig. 2. Radiologic findings in the lungs. Chest radiograph taken on the admission showed a diffuse reticulonodular pattern predominantly distributed in bilateral lower lung fields and a thickening of a minor fissure line in the right lung (A). The thickening of interstitial markings and interlobular septa, fine nodules and small cysts scattering dominantly in bilateral lower lobes (B), representative image of the lower lung field. The chest CT taken 4 months after the initiation of an oral administration of predonisolone demonstrated that the thickening of interstitial markings and interlobular septa had lessened, and the fine nodules had disappeared. (C), representative image of the lower lung field.