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CORRESPONDENCE

A rare case of folliculotropic mycosis fungoides with eosinophilic pneumonia

A 51-year-old male with no history of allergies presented with a 5-year history of pruritic erythematous papules and plaques on his trunk previously diagnosed as prurigo nodularis. He was treated with topical steroids. The nodules and erythema gradually spread to his face, scalp, and body (Figure 1A,B). Facial skin biopsies showed dense atypical lymphocytic infiltration with prominent folliculotropism (Figure 1C–E). On immunohistochemical staining, the lymphocytes were positive for CD3 and CD4 (Figure 1F), but negative for CD8, CD20, CD30, and CD56. Positron emission tomography-computed tomography (CT) showed left cervical lymphadenopathy with atypical lymphoid cell infiltration on biopsy. The rearrangement of the T-cell receptor Cß1 chain was detected on skin biopsy by the southern blot technique. The patient was diagnosed with folliculotropic mycosis fungoides (FMF), stage IV A2 T3N3M0B0. He received narrow-band ultraviolet B irradiation and vorinostat, followed by cyclophosphamide, adriamycin, vincristine, and prednisolone (CHOP) therapy. After two CHOP cycles, he complained of cough and mild dyspnea with significantly elevated soluble interleukin-2 receptor (sIL-2R; 9421 U/ml) and eosinophil (2500/μl) counts. Chest radiography and CT scan revealed diffuse intraalveolar and interstitial infiltrates (Figure 1G,H). However, his respiratory symptoms were stable despite the severe pneumonia. The patient underwent bronchoscopy with bronchoalveolar lavage (BAL). The BAL fluid (BALF) contained eosinophils, lymphocytes, and neutrophils. Atypical cells were not revealed clearly in the BALF. These findings were consistent with eosinophilic pneumonia (EP), which was treated by steroid pulse therapy with methylprednisolone (1 g daily, 3 days) (Figure 1I). However, it recurred six times. Interestingly, gemcitabine and etoposide temporarily improved the symptoms as confirmed by chest radiography before myelosuppression. The patient was diagnosed with EP associated with infiltration of FMF given that chemotherapy was effective against pneumonia. Hypereosinophilic syndrome was ruled out with no detection of *FIP1L1-PDGFRA* fusion gene. Finally, he received allogeneic bone marrow transplantation. However, the FMF recurred with inguinal lymph node infiltration, and the patient died from infection.

To our knowledge, this is the first reported case of FMF involving EP. EP is a heterogeneous disease characterized by an increase in eosinophils in the lung tissue or BALF.[1] Malignant tumors are considered as a possible cause of EP. FMF is a variant of mycosis fungoides, which expresses CCR4 in cutaneous T-cell lymphoma (CTCL) and presents as cutaneous folliculotropism.[2] A TH1 to TH2 profile shift is observed in advanced CTCL stages, which involves interleukin-5 (IL-5) release. A patient with CTCL developing EP reportedly presented with increased IL-5.[3] However, the patient's details were not described. The previous report and our case show the correlation between EP, eosinophil count in peripheral blood, and serum sIL-2R. The mechanism behind EP involves the release of IL-5 and cytotoxic granular proteins from eosinophils.[4] Chemotherapy was transiently effective for EP in our case, indicating the possibility of the aforementioned mechanism. If EP with CTCL was caused by cytokines from tumor cells or associated eosinophils, it would be difficult to treat EP without CTCL

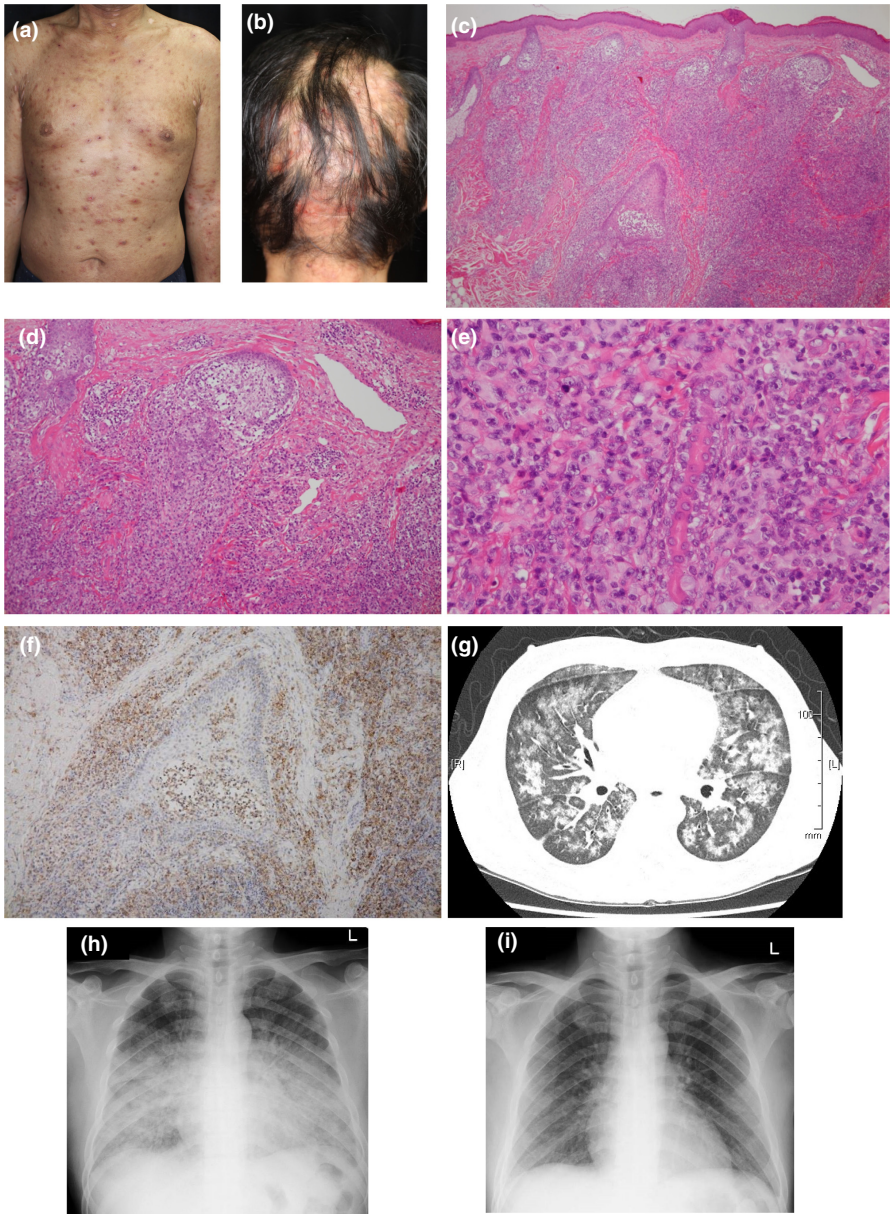

FIGURE 1 (a, b) Papules and erythema on the patient's body and back of the head at the first consultation. (c–e) Histopathological findings showed a dense atypical lymphocytic infiltrate with prominent folliculotropism (hematoxylin-eosin; [C] ×40; [D] ×100; [E] ×400). (f) Immunohistochemical staining showed that the atypical lymphocytes were positive for CD4, ×100. (g, h) Chest radiography and computed tomography demonstrated diffuse intra alveolar and interstitial infiltrates. (i) A chest radiography showed improvement of eosinophilic pneumonia after steroid pulse therapy

treatment. The anti-IL-5 antibody might also be helpful in EP management.

DECLARATION SECTION

Approval of the research protocol: No human participant was involved in this study.
Informed consent: N/A.
Registry and the registration No.: N/A.
Animal studies: N/A.

CONFLICT OF INTEREST

The authors declare no conflict of interest. Dr. Manabu Fujimoto is the Editor in Chief for the Journal of Cutaneous Immunology and Allergy. Management of the peer review process, and all editorial decision-making, for this article was undertaken by an Associate Editor.

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