Eosinophilic folliculitis in a patient with chronic myelomonocytic leukemia

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Key words: chronic myeloid leukemia; chronic myelomonocytic leukemia; eosinophilic dermatosis of hematologic malignancy; eosinophilic folliculitis; paraneoplastic dermatoses.

INTRODUCTION

Eosinophilic folliculitis (EF) is a rare, recurrent dermatosis characterized by folliculocentric papules, pustules, or plaques affecting the head, trunk, and extremities. It comprises 4 subtypes: classic, infantile, HIV-associated, and hematologic malignancy-associated EF. Diagnosis of EF in association with hematologic malignancy can be challenging owing to variable morphologies and histopathologic findings. Here we describe a patient with EF in association with chronic myelomonocytic leukemia (CMML).

CASE REPORT

A 79-year-old man with a recent diagnosis of CMML presented with a 1-month history of a persistent, highly pruritic eruption involving the forehead, chest, abdomen, and upper extremities. The lower extremities were spared. Physical examination found follicular papules and nodules (Fig 1). The initial differential diagnosis included folliculitis, prurigo, Grover disease, folliculotropic mycosis fungoides, and leukemia cutis. Skin biopsies found a perifollicular mixed infiltrate composed of lymphocytes and eosinophils, with follicular spongiosis (Figs 2 and 3). Examination of multiple level sections highlighted prominent exocytosis of eosinophils in the follicular epithelium; rare eosinophils were also identified within the infundibular epithelium and sebaceous lobule (Fig 4). The infiltrate was largely composed of CD3+ T cells, with a CD4/CD8 ratio of approximately 4:1. Cytologic atypia was absent within lymphocytes. Myeloperoxidase, CD117, and lysozyme were negative. Alcian blue stain failed to demonstrate intrafollicular mucin. Complete blood count with differential demonstrated 1.6% eosinophils (300/mm³, with total white blood cell count of 16,600/mm³). Enzyme-linked immunosorbent assay for antibodies against HIV was negative. After diagnosis of EF in the setting of hematologic malignancy, the patient was treated with topical tacrolimus and mometasone ointments along with narrow-band ultraviolet B phototherapy. Only 10% to 20% resolution was observed after 1 month, so therapy with isotretinoin (0.5 mg/kg/d) was initiated. After 2 weeks, there was resolution of pruritus and approximately 75% of lesions.

DISCUSSION

The classic variant of EF, Ofuji disease, presents as a papulopustular eruption distributed over the face, torso, and arms primarily in Japanese women and lasting for approximately 7 to 10 days with relapses. The HIV-associated variant, an AIDS-defining illness, presents with erythematous, follicular papules and urticarial plaques in patients with a CD4 cell count below 300 cells/mL. Infantile EF, the rarest variant, presents with recurrent crops of sterile papules on the scalp of boys between 5 and 14 months of age and tends to resolve spontaneously. Hematopoietic disorders associated...
with EF include acute myeloid leukemia, chronic lymphocytic leukemia, Hodgkin lymphoma, non-Hodgkin lymphoma, Sézary syndrome, and myelodysplastic syndrome. EF has also been described following chemotherapy, bone marrow transplantation, and peripheral blood stem cell transplantation. In the context of hematologic malignancy, EF presents with follicular papules and vesicles above the waist. EF is markedly pruritic in all forms.

The pathogenesis underlying EF is likely immune dysregulation in response to a follicular antigen, with subsequent production of cytokines, chemotactic factors, and expression of intercellular adhesion molecules. In association with B-cell neoplasms, clonal expansion of T helper 2 cells with interleukin-5 production leads to recruitment of eosinophils. Hypersensitivity to Demodex may also be contributory.

Diagnosis of EF in association with hematologic malignancy can be challenging because of variable morphologies and histopathologic findings. Folliculocentric papules, vesicles, pustules, and urticarial plaques may all be observed, distributed over the head and neck, upper trunk, and arms. EF

Fig 1. Eosinophilic folliculitis. Excoriated papules and nodules on the chest and abdomen (A) and cheek (B).

Fig 2. Eosinophilic folliculitis. Perifollicular mixed infiltrate with lymphocytes and eosinophils. (Hematoxylin-eosin stain; original magnification: ×100.)

Fig 3. Eosinophilic folliculitis. Eosinophils adjacent to infundibular epithelium with spongiosis. (Hematoxylin-eosin stain; original magnification: ×400.)
can be confused with insect bite reaction, bacterial folliculitis, folliculotropic MF, and leukemia cutis. Eosinophilic dermatosis of hematologic malignancy (EDHM) is a term that may encompass insect bite–like persistent hypersensitivity reactions and eosinophilic dermatosis of myeloproliferative disease. Histopathology in EF and EDHM shows a lymphocytic infiltrate, perivascular and interstitial eosinophils, and intraepidermal and intrafollicular eosinophilic spongiosis. EDHM and EF in association with hematologic malignancy likely represent the same diagnosis: these 2 entities share the same clinical context, morphology, distribution, and natural history. Although neoplastic clonal cells may be identified in EF and EDHM, most of the inflammatory infiltrates comprise reactive T cells. Dermatopathologists should avoid overdiagnosis of leukemia cutis in this context. Multiple-level sections to identify follicular eosinophilic spongiosis and careful correlation with clinical findings are helpful.

Treatment of EF is based on associated systemic conditions, patient preference, and response. Classic EF usually responds to indomethacin with more than 80% efficacy. Topical corticosteroids are described as first-line therapy for the other 3 EF subtypes. Narrow-band ultraviolet B therapy is also effective for EF associated with hematopoietic disorders, especially in controlling pruritus. Topical and systemic retinoids, dapsone, and oral tetracycline class antibiotics have also been used for EF. Treatment of underlying disease states may also result in improvement.

EF should be considered in the differential diagnosis for a patient with hematologic malignancy who presents with pruritic, folliculocentric papules, pustules, or plaques affecting the head and neck, trunk, and upper extremities. Eosinophilic spongiosis and eosinophils within sebaceous lobules or the infundibular epithelium are helpful histologic features. Careful clinicopathologic correlation by dermatologists is paramount in the diagnosis in this context and in distinction from specific cutaneous manifestations of hematologic malignancy and hypersensitivity reactions such as true insect bite reactions.

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