Eosinophilic Pustular Folliculitis Associated with Cutaneous Angiosarcoma

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To the Editor: A 64-year-old Chinese man had a 12-month history of nonmetastatic primary scalp angiosarcoma [Figure 1a] that had been confirmed by skin biopsy [Figure 1e]. Immunohistochemical examination showed CD31-, CD34-, and D2-40-positive tumor cells [Figure 1f–1h]. Extensive local excision and placement of a free skin graft was performed in December 2016. The patient declined postsurgical treatments including chemotherapy, radiotherapy, and immunosuppressive drugs. Two months later, he was referred to us with pruritic red eruptions on his cheeks and scalp. The lesions gradually increased in size and number and became pruritic. Physical examination revealed multiple follicular papules and pustules on his scalp and cheeks, forming an annular configuration on the face [Figure 1b]. The total leukocyte count was 6150/μl, with 24.4% lymphocytes (1500/μl) and 22.6% eosinophils (1390/μl). Serological tests were negative for human immunodeficiency virus (HIV), hepatitis B and C, syphilis, serum immunoglobulins, and antinuclear antibodies. The results of brain magnetic resonance imaging and thoraco-abdominal computed tomography were unremarkable. Excisional biopsies from the cheek and scalp showed predominantly intense infiltration of eosinophils, neutrophils, and lymphocytes in the pilosebaceous unit [Figure 1d] and around the blood vessels. Based on these findings, a diagnosis of eosinophilic pustular folliculitis (EPF) was made. He was then treated with oral prednisone at 30 mg/d. The treatment resulted in prompt remission of the EPF [Figure 1c]. The prednisone dosage was then gradually tapered. The skin eruptions on the face recurred when the prednisone dosage was tapered to 5 mg/d. After the dosage was increased to 30 mg/d again, the skin lesions markedly improved. The patient is still undergoing follow-up at our clinic.

EPF, first described by Ofuji, is characterized by the development of follicular papules and sterile pustules associated with eosinophilia.[1] There are three variants: classic EPF, immunosuppression-associated (mostly HIV related) EPF, and infancy-associated EPF.[1] The pathogenesis of EPF remains unknown. Many cases of EPF are associated with immunosuppressive conditions including HIV infection and various hematological malignancies,[2] suggesting that disturbance of immune function is an important causative factor. Angiosarcoma is an aggressive malignant endothelial cell tumor.[3] Several cases of AIDS-related angiosarcoma have been reported, in which the contribution of immunosuppression could be implied in the pathogenesis of angiosarcoma.[4,5] Partly because immune dysfunction may be involved in both diseases, EPF might have been associated with the cutaneous angiosarcoma in our case. To the best of our knowledge, few published reports have described EPF associated with cutaneous angiosarcoma, but it is quite possible that the development of EPF adjacent to the cutaneous angiosarcoma in our patient was coincidental. Nevertheless, this report highlights EPF as an important differential diagnosis to consider in patients with cutaneous angiosarcoma. Our patient was at risk of recurrence of cutaneous angiosarcoma because this malignancy has a poor prognosis with a 5-year survival rate of 10–30%, and the recurrence rate is extremely high.[3] Greater awareness of distinguishing clinical and histological features of the two conditions is required. We hope that our report will alert and encourage reporting of other similar cases.

Declaration of patient consent

We have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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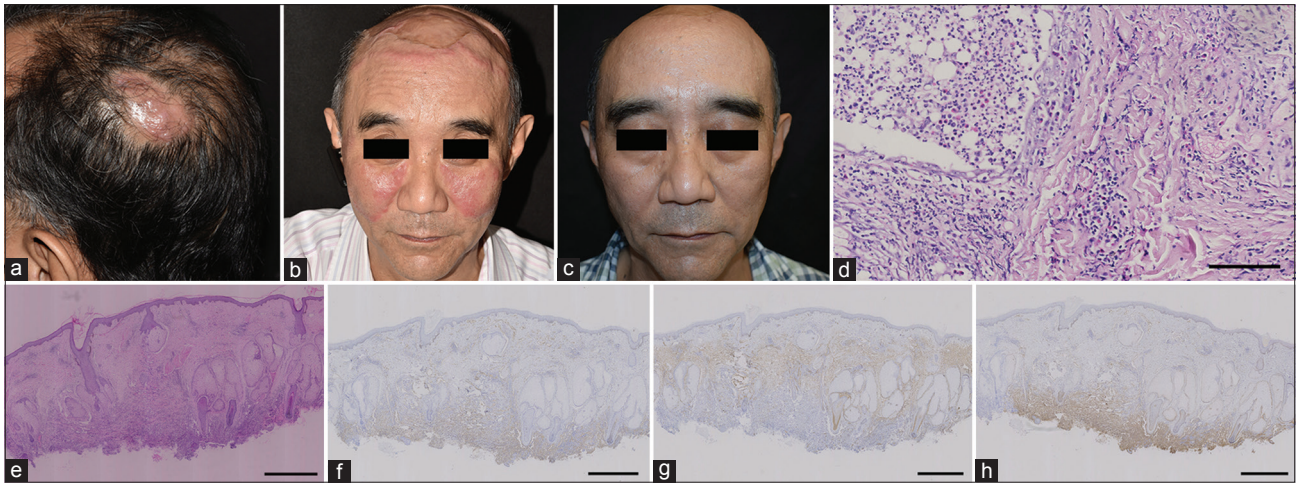

Figure 1: (a) Oval-shaped red nodule with defined borders on the scalp. (b) Multiple follicular papules and pustules on the scalp and cheeks, forming an annular configuration on the face. (c) Two months after prednisone treatment. (d) Infiltrate comprising eosinophils, neutrophils, and lymphocytes within and around the pilosebaceous unit around the hair follicle (H & E staining, bar = 100 μm). (e) Irregular vessels with conspicuously large endothelial cells exhibiting nuclear atypia in the dermis (H & E staining, bar = 1 mm). (f) CD31-positive expression in the tumor cells (bar = 1 mm). (g) CD34-positive expression in the tumor cells (bar = 1 mm). (h) D2-40-positive expression in the tumor cells (bar = 1 mm).

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