1. Eosinophilic pustular folliculitis

Eosinophilic pustular folliculitis (EPF), also termed Ofuji disease, was first described by Ise and Ofuji in 1965 in a 42-year-old Japanese woman who presented with folliculocentric pustules on her face, arms, and trunk. This condition has been reported primarily in Japan, but any race can be affected. It is a distinct skin disorder characterized by noninfectious eosinophilic infiltration of the hair follicles. Men are affected more frequently than women. The exact pathogenesis remains unclear, but immune dysfunction and allergic hypersensitivity might play a role.

Today, EPF is known to comprise 3 clinical subtypes. The first is classic EPF, which tends to occur in Japanese patients who present with recurrent crops or clusters of follicular papules and pustules that have a tendency to form annular plaques. Classic EPF predominantly appears in seborrheic areas (face, upper back, extensor surfaces of the upper arms). Up to 20% of affected patients might display palmoplantar involvement. Peak occurrence of classic EPF is during the third or fourth decades of life. The second subtype of EPF is associated with immunosuppression, mainly HIV infection. In rare instances, it might also be associated with other immunosuppressive conditions, such as hematologic or lymphoproliferative diseases. This subtype is quickly becoming the most common variant of EPF. Unlike classic EPF, immunosuppression-associated EPF tends to manifest as extremely itchy, follicular, urticarial papules mainly involving the head, neck, and proximal extremities. The third subtype of EPF occurs in infancy and the neonatal period. The lesions are similar to those of classic EPF in that they comprise sterile papulopustules, but unlike classic EPF they are not grouped in an annular arrangement. They are often located on the scalp, but might occasionally be found on the face and extremities.

Diagnosis of EPF depends on clinical suspicion in conjunction with characteristic histopathologic findings. The most striking histologic feature is the infiltration of eosinophils into hair follicles and perifollicular spaces. The eosinophilic infiltration is sometimes mixed with lymphocytes or neutrophils, and mucin deposition in the hair follicle might occasionally be noted.

Prognosis is usually good for the neonatal variant of EPF. However, classic and immunosuppression-associated EPF often carry poorer prognoses, with a chronic clinical course and recurrent relapses over many years in most patients.

Differential diagnosis

Tinea faciei is a superficial dermatophyte infection, limited to the face, that predominantly affects pediatric populations owing to children’s frequent contact with pets. The clinical presentation can range from typical erythematous and scaly plaques, with or without active borders composed of papulovesicles, to atypical features, such as discrete patches of small, raised bumps. The diagnosis can be confirmed by combining surface scrapings from the border of the lesions with a potassium hydroxide preparation to reveal the presence of fungus. Topical antifungal agents such as terbinafine or ciclopirox are effective treatments.

Annular pustular psoriasis (APP) is a rare and unique clinical variant of pustular psoriasis. It tends to have a chronic, recurrent course, but carries a good prognosis compared with generalized pustular psoriasis. Clinically, its lesions can present with very similar morphology to that in our patient: annular or circinate plaques with relative central clearing and peripheral pustule formation. However, APP often presents with a hyperkeratotic, scaly surface compared with the usual minimal epidermal changes of EPF. A wider area of involvement, such as the trunk and lower limbs, might also be noted. Clinical exacerbations are common after infections, emotional stress, or steroid withdrawal. Skin biopsy with histopathologic examination can readily differentiate APP from EPF. Prominent eosinophilic infiltration in the hair follicles never appears in APP. Most patients with APP have a good response to mild treatment measures, such as topical corticosteroids and compresses, whereas others might require systemic therapy, such as retinoids, dapsone, or methotrexate.

Erythema annulare centrifugum (EAC) is an uncommon gyrate erythema. It is now believed to be caused by hypersensitivity to a long list of possible triggers, including infection, malignancy, drugs, or hormone changes, or to be idiopathic in nature. It often presents initially as discrete erythematous macules or urticarial papules, which gradually enlarge to form circinate, arcuate, or polycyclic figures with central clearing. The edges of the lesions can often advance by several millimetres a day. Unlike EPF, EAC never shows pustule formation. The primary goal of treatment is to search for possible underlying disease, as many cases of EAC are resolved once the underlying causes are treated. Initial management of skin lesions is mainly symptomatic. Topical or even systemic corticosteroids can usually suppress the lesions, but recurrence is common once the medications are withdrawn. Other treatment options that have been reported to have some success include topical calcipotriol, narrow-band UVB phototherapy, psoralen-UVA photochemotherapy, and etanercept.

Subacute cutaneous lupus erythematosus (S克莱), which primarily affects young to middle-aged white women, accounts for 10% to 15% of all cases of cutaneous lupus erythematosus. Two distinct subtypes of S克莱 have been identified: annular S克莱 and papulosquamous S克莱. The former is not easily differentiated from EPF because of morphological overlap. Annular S克莱 lesions are characteristically photosensitive and appear to prefer sun-exposed areas (upper back, shoulders, extensor surfaces of the arms), exposed areas of the neck, and, less commonly, the face). When the face is involved, lesions are often located on the lateral
surface with relative sparing of the central malar areas. The SCLE lesions might undergo vesiculobullous changes at the active annular border owing to severe injury to epidermal basal cells, but pustules rimming the border are never seen. Laboratory investigations are useful adjuncts to confirming diagnosis. Autoantibodies, such as anti-Ro/SSA, anti-La/SSB, and antinuclear antibody, are present in a high percentage of SCLE patients. Characteristic histopathologic features include hyperkeratosis, epidermal atrophy, vacuolar degeneration of basal cells, dermal mucin deposition, and superficial or deep perivascular and periappendageal mononuclear cell infiltration. Most SCLE lesions can be managed with localized treatments such as topical corticosteroids, calcineurin inhibitors, or intralesional steroid injection. In persistent and substantial local disease activity or superimposed systemic activity, systemic agents such as hydroxychloroquine or immunosuppressive drugs are indicated. Long-term follow-up is mandatory for these patients, as a few might eventually develop active SCLE.

Management of EPF

Topical corticosteroids tend to be the first approach for all 3 types of EPF, but it has been suggested that indomethacin, a nonsteroidal anti-inflammatory drug, is the most effective first-line treatment, especially for facial lesions. Phototherapy with UVB or psoralen-UVA might also be beneficial to some patients. Recently, many cases of EPF were successfully treated with topical tacrolimus or pimecrolimus; therefore, these topical immunomodulators are becoming another treatment option, especially for facial lesions. Most important, with patients suspected to have HIV-associated EPF, prompt serologic evaluation to detect occult HIV infection is highly recommended. Antiretroviral treatment often results in clinical improvement of the extremely pruritic urticarial lesions seen in HIV-associated EPF.

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Competing interests
None declared

References

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4. Senile hemangioma

Senile hemangiomas (or angiomata or De Morgan spots) are the most common benign acquired vascular malformations. They occur in nearly all adults older than 30 years, increasing in number with age. Clinically, these hemangiomas are often incidentally noted on physical examination. Senile hemangiomas often appear as numerous red, firm maculopapular spots, less than 5 mm in diameter, mainly on the trunk and arms. Direct trauma can eliciting bleeding.

The cause and pathophysiology of senile hemangiomas are unclear, but they mainly derive from the venous limb of the capillary loop and have a unique collagen composition. They increase in number during pregnancy, suggesting a hormonal influence. They have also been associated with solid-organ transplantation, graft versus host disease, and exposure to various different chemicals; however, a causative relationship has never been established.

Diagnosis is clinical and based on the appearance and characteristics of the lesions, as outlined above. Senile hemangiomas differ in appearance and distribution from other cutaneous acquired vascular malformations. Venous lakes are larger dark blue, dome-shaped lesions, typically found on sun-exposed areas of the skin, most often the vermilion border of the lip, the face, and the ears of elderly people. Angiokeratomas are superficial dermal blood vessels, with hyperkeratosis of the overlying epithelium, most commonly found on the scrotum or vulva. Lymphangioma simplex consists of small translucent vesicles on a red-brown base, usually made up of a mixture of dilated vascular and lymph channels. Kaposi sarcoma is a vascular hemorrhagic neoplasm most often found in patients with AIDS or in men older than 60 years.

Atypical lesions or lesions that have acutely changed should trigger a more thorough investigation, which might include referral to a dermatologist or plastic surgeon.

Senile hemangiomas are clinically insignificant; reasurance is the only treatment required. Their natural course is one of atrophy and fading with age. Treatment can be sought for cosmetic reasons; scissors excision, electrocautery, or laser desiccation and curettage have all been shown to be effective, while cryotherapy has not.

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