Eosinophilic pustular folliculitis of infancy: A series of 15 cases and review of the literature

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Background: Eosinophilic pustular folliculitis (EPF) of infancy is characterized by the presence of pustular lesions containing eosinophils. It is the least well-characterized of the EPF diseases.

Objectives: We sought to define the clinical and histopathologic features of the condition.

Methods: We conducted a retrospective review of the clinical data and histologic findings of 15 patients given the diagnosis of EPF of infancy at the Hospital Niño Jesús, Madrid, Spain, from 1995 to 2011, and of patient data published in MEDLINE with such a diagnosis from the disease description (1984-2011).

Results: A total of 61 cases were collected. The disease was more common in males than females (ratio 4:1), and presented before 14 months of life in 95% of cases (mean 6.1 months; median 5 months). All patients had recurrent outbreaks and scalp involvement, and 65% had lesions on areas of the body other than the scalp. Tissue eosinophilia was present in all cases; however, true follicular involvement was observed only in 62% of cases in which histologic study was available. More than 80% of the patients were cured by 3 years of age (mean 25.3 months; median 18 months). Topical steroids were effective in 90% of cases.

Limitations: This was a retrospective study.

Conclusions: EPF of infancy presents most often in the first 14 months of life and usually resolves by 3 years of age. All patients showed scalp involvement, tissue eosinophilia, and recurrent outbreaks. The condition does not require aggressive treatment, as it is benign and self-limiting. (J Am Acad Dermatol 2013;68:150-5.)

Key words: eosinophil; folliculitis; infancy; pustular.

Infants may present different types of inflammatory lesions of eosinophilic nature that are usually recognizable with ease on clinical grounds. Age of onset, location of lesions, and coexistence of infections or immunodeficiency are diagnostic keys. Eosinophilic pustular folliculitis (EPF) of infancy (EPFI) usually presents in early infancy and localizes on the scalp and other sites on the body surface. The lesions consist of recurrent crops of isolated or grouped papules and pustules that typically lack the annular or polycyclic ring characteristic of adult EPF. The condition is pruriginous, but patients remain in good health and do not have any associated disease. The recognition of EPFI and its distinction from other types of pustular lesions is critical to avoiding unnecessary diagnostic procedures and inappropriate treatments. In this study we report 15 new cases of the disease and review the literature to further characterize the disease.

METHODS

We performed a retrospective study of all cases of EPFI in the Hospital Niño Jesús, Madrid, Spain, between 1995 and 2011. We included only cases in which infectious processes had been ruled out and that showed tissue eosinophilia confirmed by means

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of smear, skin biopsy specimen, or both. We analyzed gender, age of onset, location of lesions, existence of pruritus, histologic findings, recommended treatment, duration and frequency of outbreaks, and age at resolution. We also assessed blood eosinophilia and IgE levels, when available, along with associated diseases. Because follow-up information had been lost in some of our cases, duration of the process and personal history of atopy were assessed by telephone interview. In addition, the same data were reviewed in cases of EPFI published on MEDLINE between 1984 and 2011. The articles had to be originally written in or translated into the English language. We excluded patients older than 14 years, and several patients whose diagnosis was exceedingly atypical or coexisted either with infectious processes or immune deficiency.

CAPSULE SUMMARY

- Eosinophilic pustular folliculitis (EPF) of infancy is the least well-characterized of the EPF diseases.
- Recurrent crops of papules and pustules involving the scalp are typical. Despite its name, it does not always show true folliculitis, and the eosinophilic infiltrate may spare the hair follicle.
- The condition is benign and self-limiting and does not require aggressive treatment.

RESULTS

A total of 61 cases were evaluated, including our series of 15 cases (3 of which had previously been published in the Spanish-language literature[1]) and 46 more published in the literature.[2-19] Twelve of the previously published cases were excluded,[13,20-27] some of which had already been questioned by other authors[28]: 1 atypical case on both clinical and histologic grounds (genital location exclusively, dermal infiltrate rich in neutrophils in 1 biopsy specimen, and in lymphocytes and macrophages in another)[25]; 3 cases associated with immunodeficiency[26,27]; 1 patient who developed sepsis with hepatic and pulmonary involvement in whom immunodeficiency was not ruled out[13]; and 1 exceptional case in which the eruption did not recur, the histologic findings were inconclusive, and that was treated with granulocyte-colony stimulating factor (G-CSF) because of the extraordinarily high eosinophilia.[22] There were 6 more cases that were also excluded because of their atypical manifestations: a 4-year-old child who had recurrent vesicles and pustules along with erythema multiforme–like lesions on the abdomen and lower limbs that might have corresponded to arthropod bites[23]; an 8-year-old boy with mucosal involvement, mixed dermal inflammatory infiltrate with eosinophils and neutrophils, and good response to dapsone in whom linear IgA was not excluded[20]; an 11-year-old girl who presented with an isolated annular plaque on the cheek showing a dense eosinophilic infiltrate and necrotizing vasculitis of the small vessels,[24] which is not a feature of EPFI; a 5-year-old girl with ulcerated lesions on the lower legs that resolved in 2 weeks[21]; and 2 more patients who presented with crops of pustular lesions on the trunk and lower limbs for 3 months' duration who demonstrated hypersensitivity to *Dermatophagoides pteronyssinus* and elevated IgE levels.[10] The authors did not provide clinical photographs of these 2 patients, 1 of whom also had atopic dermatitis, and included a third case of an 11-month-old patient with typical manifestations of EPFI.

The findings observed in our series of 15 patients were similar to those of the 46 cases collected from the literature. According to the global data, the age at presentation ranges from birth to 36 months of life (mean 6.1 months; median 5 months); in 95% of cases, the disease appeared before 14 months of life, and by 6 months in 70% (Fig 1). The condition was much more common in males than females, with a 4:1 ratio. Lesions consisted of isolated or clustered papules, papulopustules, and vesicles with an erythematous base that were located on the scalp in all patients, and on other body areas in 40 cases (65%) (Fig 2). Two patients, including 1 of our series, showed facial edema.[19] Pruritus was reported in 84% of patients in whom these data were recorded. In all cases, tissue eosinophilia was demonstrated either by smear or skin biopsy specimen. When performed, biopsy specimens demonstrated eosinophilic inflammatory infiltrate in all cases; it involved the follicle in 27 cases (Fig 3) and spared it in 16 instances, in which the infiltrate was perifollicular or interfollicular. In 11 nonbiopsied cases, a pustule smear confirmed the massive presence of eosinophils. Flame figures were seen in 3 cases.[5,12] Blood eosinophilia was requested in 42 cases, being elevated in 35 (83%); it was in the upper normal limit in 4 cases,[5,7] and was normal in 3.[5,15] IgE levels were available in only 13 patients[2,3,5,6,10]; they were very high in 1,[10] a patient who showed hypersensitivity to the dust mite *D pteronyssinus*, and only slightly elevated in 2.[5] The age at resolution ranged from 4 months to 9 years (mean 25.2 months; median 18 months). Recurrences occurred in 100% of the children,