A Case of Eosinophilic Pustular Folliculitis (Ofuji's Disease) in a Newborn

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We describe a case of eosinophilic pustular folliculitis in a 1-month-old Korean male infant. The patient was suffering from erythematous papules and pustules affecting the scalp, trunk, and extremities, detected at birth. The flare was accompanied by leukocytosis and eosinophila. Histopathology revealed folliculitis with a predominant eosinophilic infiltrate. Lesions showed partial response to topical steroid and disappeared in four weeks. Three months after the resolution of the lesions, he presented a similar clinical picture as the previous episode. (Ann Dermatol 14(2) 117-120, 2002).

Key Words : Infantile eosinophilic pustular folliculitis

Eosinophilic pustular folliculitis (EPF), or Ofuji's disease is an inflammatory dermatosis which was first described in 1970 by Ofuji et al[1]. The eruption of EPF consists of follicular papules and pustules that tend to form an annular configuration mimicking fungal infections. The histopathologic feature of EPF is characterized by perifollicular inflammatory cellular infiltration featuring numerous eosinophils and some mononuclear cells[1]. Since the first report by Ofuji et al.[1], there have been numerous case reports of EPF worldwide; however, its pathophysiology remains unclear. In 1984, Lucky et al.[2] first described five children with EPF. Those pediatric cases were characterized by recurrent crops of sterile pustules on the scalp, trunk and extremities with the distinctive feature that in children the lesions did not form the characteristic polycyclic rings found in adults. Since then in only a few instances has EPF been described in infants and children. In 1986, Soeprono and Schinella[3] described three cases of EPF in patients with acquired immunodeficiency syndrome. EPF seen in patients with human immunodeficiency virus infection is clinically distinct from classical EPF first described by Ofuji et al., therefore Moritz and Elmets[4] once proposed to classify EPF into classical, HIV-associated, and infantile type. We describe a case of EPF occurring in a neonate.

CASE REPORT

A 1-month-old Korean male infant born after a normal pregnancy was transferred to our outpatient department with papules and pustules on the scalp, trunk, and feet. This eruption was detected at his birth in the form of erythematous papules evolving into follicular pustules. On physical examination, multiple variable sized erythematous papules and follicular pustules were found on the scalp (Fig. 1), retroauricular areas, trunk, and feet. Laboratory tests revealed no abnormal findings except peripheral blood leukocytosis (22,000/mm^3) with prominent eosinophilia (5,971/mm^3). Examination of pustules for fungi and bacteria was nega-

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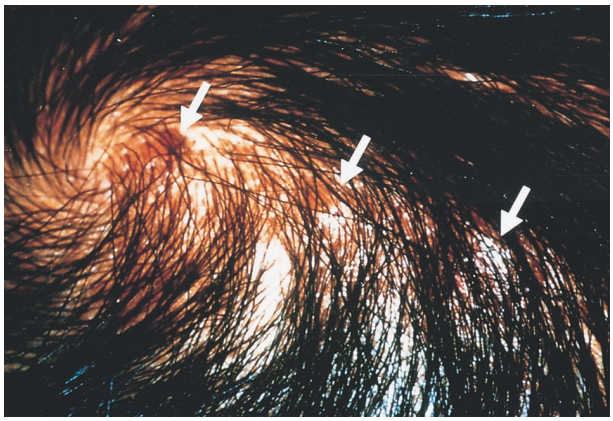

Fig. 1. Multiple variable sized erythematous papules and follicular pustules on the scalp (white arrows).

tive. Parasitic etiology was excluded by the negative result of stool examination. The histopathological study from the scalp lesion (Fig. 2) showed pilosebaceous follicular infiltrates with numerous eosinophils and some mononuclear cells, followed by intrafollicular eosinophilic pustules featuring spongiosis of the follicular outer root sheath. A periodic acid-Schiff (PAS) stain for fungi was negative. Under the diagnosis of EPF, topical corticosteroid therapy was started, and produced some improvement in two weeks of treatment. The skin lesion resolved in the following two weeks. Three months after the resolution of the eruption, the patient suffered from measles and similar papules and pustules were present on the scalp.

DISCUSSION

Lucky et al.[2] described EPF in infancy as a disease characterized by recurrent crops of sterile pustules on the scalp, trunk, and extremities. In children and infants, the lesions tend to be located on the scalp and do not form characteristic annular lesions with centrifugal enlargement, accompanying invariable pruritus and occasional erythematous urticarial, follicular papules. Although there are significant differences in the clinical manifestations between infantile and classical forms, the histopathologic findings of both infantile and classical EPF show no differences. The characteristic histopathologic changes of follicular pustules rich in eosinophils confirmed the diagnosis of EPF in our case. Since the first report of EPF in infancy by Lucky et al.[2], relatively small numbers of pediatric cases have been reported in English literature. In Korea, there have

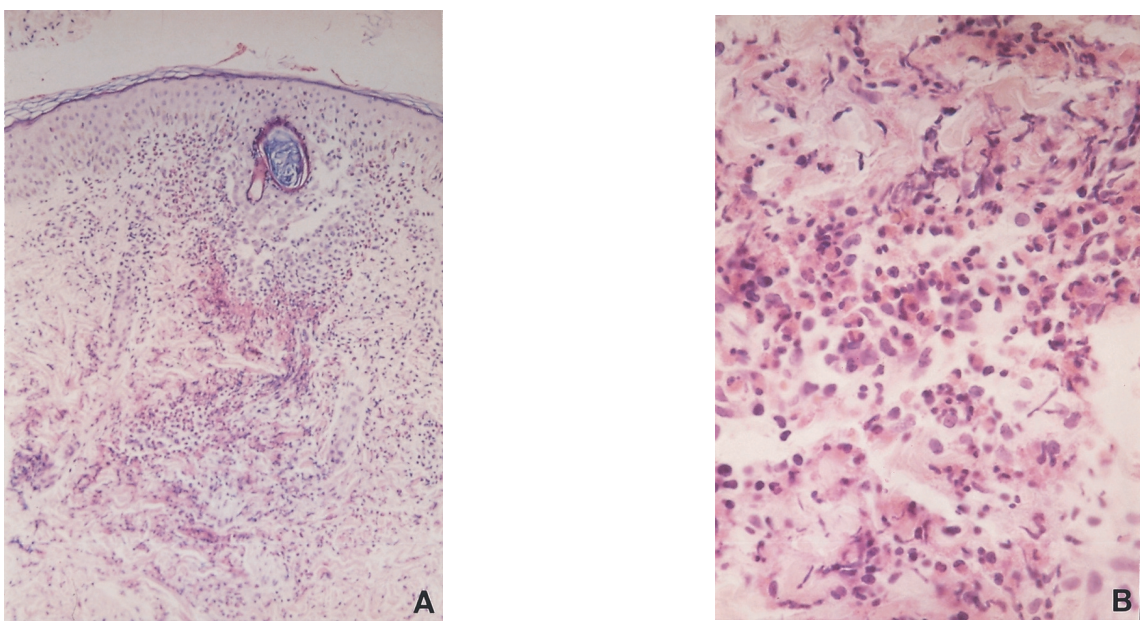

Fig. 2. A) Massive perifollicular and intrafollicular inflammatory infiltrates combined with spongiosis of the follicular outer root sheath (H &E, × 40). B) Follicular inflammatory infiltrate consisting of lymphocytes, neutrophils, and numerous eosinophils (H &E, × 200).

Table 1. Clinical data of patients with infantile EPF in published articles in Korea

Sex/Age (month)	Duration	Distribution	WBC(/mm3)	Eosinophils	Treatment	Recurrence	Reference
F/63	months	Scalp	ND	ND	Antihistamine, Topical steroid	ND	Hong et al.[5]
F/3	20 days	Scalp	7,900	6.3(%)	Antihistamine, Topical steroid	+	Kim et al.[6]
F/7	6 months	Scalp	ND	ND	Antihistamine, Topical steroid	+	Kim et al.[6]
M/8	3 months	Scalp	ND	ND	Topical steroid	ND	Jang et al.[7]
M/6	1 month	Scalp	10,100	25(%)	Antihistamine, Topical steroid	-	Choi et al.[8]
M/1 (present case)	1 month	Scalp, Trunk, Feet	22,000	5,971(/mm3)	Topical steroid	+	-

Abbreviations: M, male; F, female; ND, not described

been no more than five reported cases of infantile EPF[5-8] at all. Table 1 summarizes the clinical data of the patients with infantile EPF in published articles in Korea. As far as we know, this is the first Korean report of a patient with EPF occurring at birth even though there have been a few cases of neonatal EPF[2,9-11] worldwide. All the six Korean cases presented only the scalp lesions. However, our patient showed typical lesions of infantile EPF not only on the scalp but also on trunk and extremities. Three of the six Korean cases revealed that the skin lesions can recur after complete remission; therefore careful follow-up of the patients with infantile EPF is recommended.

Even though the sebaceous gland activity has been postulated to be an etiologic factor[12], the etiology of EPF still remains obscure, especially in the infantile form. However, Boone et al.[13] described three cases of EPF in atopic children with hypersensitivity to *Dermatophagoides pteronyssinus* and have underlined the important role of hypersensitivity reactions in the pathogenesis of EPF. Teraki et al.[14] demonstrated the increased expression of intercellular adhesion molecule 1 (ICAM-1), vascular cell adhesion molecule 1 (VCAM-1), and endothelial cell adhesion molecule 1 (ELAM-1) around the affected hair follicles in the classical form of EPF.

The differential diagnosis of infantile EPF is extensive and includes erythema toxicum neonatorum, infantile acropustulosis, pustular folliculitis, transient neonatal pustular melanosis, incontinentia pigmenti, dermatitis herpetiformis, insect bite, impetigo, and Kimura's disease[15]. Among them, the most common initial diagnosis of EPF occurring at birth or in the neonatal period would be erythema toxicum neonatorum. Only the combination of the specific clinical, laboratory and histologic features determines the diagnosis of infantile EPF[8]. In our patient, along with the characteristic clinical and laboratory findings, typical histopathologic changes of follicular pustules rich in eosinophils confirmed the diagnosis of infantile EPF.

Many therapeutic agents, mainly topical corticosteroids, have been used in EPF without consistent results. Even though topical corticosteroids produced some clinical improvement in our patient, it is unclear whether the lesions disappeared spontaneously or were treated with corticosteroids. Because the clinical course of EPF appears to be neither severe nor extensive in infants and children most of the systemic modalities may not be needed.

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