Generalized Seborrheic Dermatitis in an Immunodeficient Newborn

Hyun Joo Kim, MD, Seoul, Korea
Yeon Soon Lim, MD, Seoul, Korea
Hae Young Choi, MD, Seoul, Korea
Ki Bum Myung, MD, Seoul, Korea

We report the case of a female infant with failure to thrive, generalized seborrheic dermatitis, and intermittent diarrhea. Results of laboratory investigation revealed low serum immunoglobulin G IgG levels. She failed to gain additional weight and experienced recurrent infection. She died 3 months later.

In 1908, Leiner described a series of symptoms appearing within the first few weeks of life in 43 infants with erythroderma, frequent loose stools, and failure to thrive.[1,2] The condition was subsequently given his name. Since then, a variety of immune defects has been reported in patients with this disease.[3-8]

We examined a patient with generalized seborrheic dermatitis with concomitant low levels of IgG, sepsis, and disseminated intravascular coagulation.

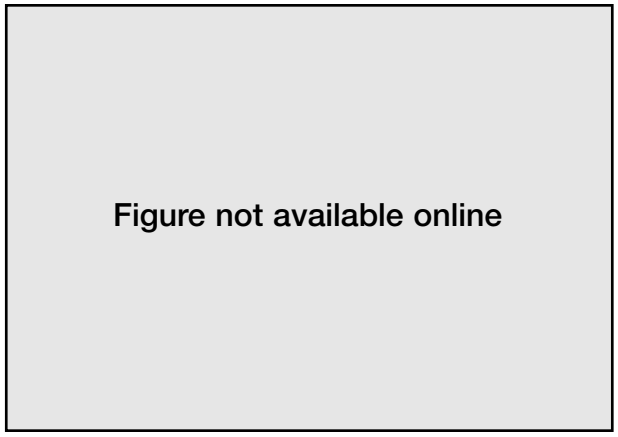

FIGURE 1. A 2-day-old female with generalized fissured and desquamated lesions on an erythematous base.

Case Report

A 2-day-old female presented with generalized fissured and desquamated lesions on an erythematous base that were present at birth (Figure 1). The patient was born by normal, spontaneous vaginal delivery to a gravida IV, para I, 27-year-old mother after 38 weeks of gestation. Birth weight was 3 kg. Apgar scores at 1 and 5 minutes were 8 and 9, respectively.

The patient had normal facial morphologic features, ectropion, abnormal scalp hair, and dystrophic nails. Initial cutaneous examination was remarkable for fissured and desquamated lesions over the whole body. Scale was absent in the flexors of the joints and in the skin folds; Nikolsky's sign was negative.

Results of a complete blood count are shown in Table 1. Other laboratory data were as follows: prothrombin time, 15.3 seconds (69%); partial thromboplastin time, 60.9 seconds; and T_4:T_8 cell ratio, 1.1:1. Fibrinogen degradation product and D-dimer were present. Results of blood culture, cerebrospinal fluid culture, and suprapubic urine culture tests were negative, as were results of serum tests for syphilis, *Treponema pallidum* hemagglutination, and human immunodeficiency virus.

On the fifth hospital day, the infant's laboratory results were as follows: hemoglobin level, 13.2 g/100 mL; hematocrit level, 38.2%; leukocyte count, 14.4×103 mm^3; platelet level, 137,000/mm^3; total bilirubin, 19.5 mg/100 mL; direct bilirubin, 0.6 mg/100 mL; prothrombin time, 13.6 seconds (89%); and partial thromboplastin time, 47.4 seconds.

Examination of a skin biopsy specimen from the right thigh revealed parakeratosis with a few pyknotic neutrophils, moderate acanthosis with elongation of the rete ridges, slight spongiosis, and a mild perivascular lymphoid inflammatory infiltrate in the upper dermis (Figure 2). These findings were consistent with seborrheic dermatitis. Treatment was begun

Drs. Kim, Lim, Choi, and Myung are from the Department of Dermatology, College of Medicine, Ewha Womans University, Seoul, Korea.
Reprints: Ki Bum Myung, MD, Department of Dermatology, Ewha Womans University, Mokdong Hospital, 911-1 Mokdong, YangCheon-Ku, Seoul, Korea 158-710.

Table 1.

Results of a Complete Blood Count and Other Laboratory Data

Measured Level at Day 2	Patient	Normal Level
Hemoglobin	15.1 g/100 mL	13–20 g/100 mL
Hematocrit	46.2%	42%–66%
Leukocyte	26.6×103/mm^3	4.8–10.8×103/mm^3
Platelets	177,000/mm^3	200,000–300,000/mm^3
IgG	498 mg/100 mL	640–1600 mg/100 mL
IgG_1	3.66 mg/100 mL	>320 mg/100 mL
IgG_2	1.69 mg/100 mL	>92 mg/100 mL
IgG_3	0.34 mg/100 mL	>16 mg/100 mL
IgG_4	0.22 mg/100 mL	>1 mg/100 mL
IgA	6.6 mg/100 mL	0–5 mg/100 mL
IgM	12.2 mg/100 mL	6–24 mg/100 mL
IgE	4 mg/100 mL	0–4 mg/100 mL
T_4 cells	22%	41%–64%
T_8 cells	19.6%	16%–35%
C_3	77.1 mg/100 mL	53–175 mg/100 mL
C_4	27.9 mg/100 mL	7–42 mg/100 mL
C_5	7.5 mg/100 mL	2.3–6.3 mg/100 mL

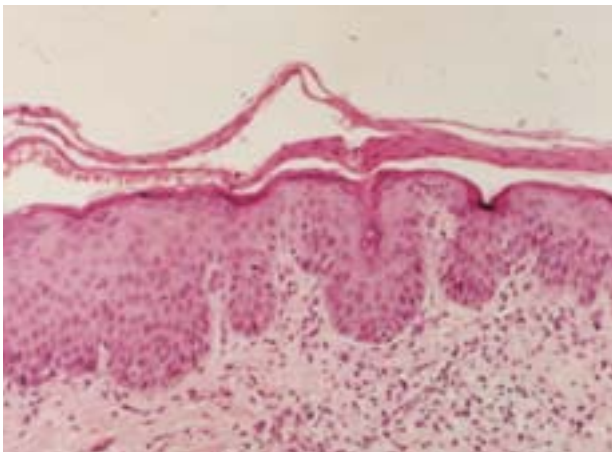

FIGURE 2. A skin biopsy specimen from the right thigh. The epidermis shows parakeratosis with a few pyknotic neutrophils, moderate acanthosis with elongation of the rete ridges, and slight spongiosis (H&E, original magnification ×100).

with topical low-potency steroids, moisturizers, and keratolytics. The infant received a transfusion of plasma for treatment of her low platelet and serum immunoglobulin levels and was given ceftriaxone and vancomycin. Phototherapy was undertaken to treat hyperbilirubinemia.

A few days later, the lesions became greasy, with white-to-yellow crusts, and gradually dried. The patient was discharged from the hospital on day 17. She was readmitted 3 days later because of lethargy, fever, poor feeding, and loose stools. The infant had no more weight gain and experienced recurrent infections, such as external otitis and sepsis. She died 3 months later because of sepsis.

Comment

The combination of generalized erythroderma, failure to thrive, and diarrhea in infancy appears to have been relatively common in Vienna when Leiner[1]

described his cases in 1908. He referred to this condition as a distinctive universal dermatitis in breast-fed children and concluded that nutrition factors may be responsible. Leiner also suggested that the disease was caused by some deficiency in the mother's milk and considered the presence of severe intestinal disturbance as pathognomonic for this disease. Intestinal manifestations consist of frequent, loose, greenish, mucoid stools and occasional vomiting, which may be projectile.[5] Our patient had only intermittent diarrhea. Glover et al[3] preferred to abandon the term *Leiner's disease* because none of their patients were breast-fed. Also, the low incidence of generalized erythroderma, failure to thrive, and diarrhea in infancy reported during the past 20 years markedly contrasts with the relatively high incidence implied by Leiner.

Various immunologic defects have been described in this illness, including a deficiency of C_5,[4-6] a yeast opsonization defect, immunoglobulin deficiency,[7] and diminished C_3.[8] It is now recognized that Leiner's disease represents a heterogeneous group of disorders. Our patient had increased C_5 and reduced IgG with all of the 4 IgG subclasses (IgG_1, IgG_2, IgG_3, IgG_4) lower than normal. C_3 and C_4 levels were normal. Further laboratory studies for measurement of functional C_5 deficiency, such as yeast opsonization defect, could not be performed on our patient.

Although Glover et al[3] believed that generalized erythroderma, failure to thrive, and diarrhea are clinical findings of immunodeficiency, it does not appear that the underlying immunodeficiency is a specific one. There is no consistent pattern of immune dysfunction in these infants and, indeed, minor deviations from normal childhood values are common in otherwise healthy infants.[9]

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