Acquired ichthyosiform erythroderma and sarcoidosis

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We describe a patient with ichthyosiform erythroderma as a manifestation of sarcoidosis. This is the first report of the simultaneous occurrence of erythroderma and ichthyosis in sarcoidosis. (J Am Acad Dermatol 1996;35:826-8.)

Cutaneous manifestations occur in 10% to 35% of the patients with systemic sarcoidosis. The most common of these manifestations are papular, nodular, plaque lesions and involvement of scars. Rare cutaneous expressions of sarcoidosis are erythroderma and acquired ichthyosis. Erythroderma with a fine, silvery and lamellar scaling has also been described.

CASE REPORT

A 60-year-old man had an erythroderma. Two years previously he had complained of malaise and weight loss (10 kg in 1 year). Examination revealed generalized lymphadenopathy and hepatosplenomegaly. His white blood cell count was 17.5 x 10^9/L; differential determinations were neutrophils, 38%; lymphocytes, 24%; monocytes, 3%; and eosinophils, 35%. Examination of liver and lymph node biopsy specimens revealed epithelioid cell granulomas with extensive eosinophilia. Low-grade lymphoma, the hypereosinophilic syndrome, and an unspecified viral infection were considered as diagnoses. Two years later hypercalcemia, increased urinary calcium excretion, progressive hepatosplenomegaly, and a non-pruritic generalized eruption developed.

Examination revealed erythroderma on the extensor surfaces of the extremities (Fig. 1). On diascopy, apple jelly lesions were observed. The extensor aspects of the extremities were covered with large lamellar scales (Fig. 2). The flexures, palms and soles, were relatively free of scales. The patient also had enlarged, painless lymph nodes.

Fig. 1. Generalized erythroderma.
nodes of the head and neck and axillary and inguinal regions. The liver and spleen had a normal consistency and were palpated 12 cm and 6 cm, respectively, below the costal margin.

Several biopsy specimens revealed multiple noncaseating epithelioid cell granulomas surrounded by a mixed infiltrate of lymphocytes and eosinophils (Fig. 3). The epidermis was slightly acanthotic, without a granular layer and with hyperkeratosis, focal acanthosis, and focal parakeratosis.1

Laboratory studies revealed a white blood cell count of 17.5 × 10^9/L, 35% eosinophils, a platelet count of 106 × 10^9/L, and serum levels of creatinine, 200 μmol/L; calcium, 3.88 mmol/L; alkaline phosphatase, 315 U/L; and angiotensin-converting enzyme, 29 U/ml (normal, less than 20 U/L). Serum levels of parathyroid hormone and vitamin D₃ were normal. Results of serum protein electrophoresis were normal. No abnormalities of the lungs or eyes were found. A gallium scan revealed no skeletal abnormalities, and the results of the Mantoux test were negative.

The results of clinical, histologic, and laboratory investigations indicated that the patient had systemic sarcoidosis with an acquired ichthyosiform erythroderma.

The patient was treated with oral prednisone and topical betamethasone valerate cream. After 6 months of treatment the hepatomegaly, splenomegaly, and lymphadenopathy had improved markedly, and erythroderma had faded. Apart from a generalized fine, silvery scaling, the skin had a normal appearance.

**DISCUSSION**

Acquired ichthyosis and erythroderma are rare cutaneous manifestations of sarcoidosis.4-8 Acquired ichthyosis usually occurs as diffuse, marked scaling
overlying erythematous papules and plaques on the legs.\textsuperscript{5,11} In one patient a generalized hyperpigmented ichthyosiform scaling was present.\textsuperscript{6} Although in most patients the distribution and the histologic picture are compatible with ichthyosis vulgaris, it should be emphasized that in all cases the ichthyosis was acquired.\textsuperscript{11} However, the absence of the stratum granulosum in the sarcoid ichthyosis, as in the report of Griffith et al.\textsuperscript{5} and in the present case, is inconsistent with ichthyosis vulgaris. In the present case, parakeratosis was present. Lamellar ichthyosis is usually characterized by hyperkeratosis,\textsuperscript{12} but parakeratosis has been described in some forms of lamellar ichthyosis.\textsuperscript{13}

In the previously reported cases, both erythroderma and acquired ichthyosis were associated with systemic involvement.\textsuperscript{14-16} In our patient the eosinophilia and lack of pulmonary involvement were unusual findings.

REFERENCES