

Erythema Gyratum Repens Accompanied by Essential Thrombocythemia, Followed by a Blastic Crisis

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Accepted November 18, 2002.

Sir,

Erythema gyratum repens (EGR) was first described by Gammel (1) in 1952 in a patient with a breast carcinoma. Since then, EGR has been reported in conjunction with underlying malignancies, lung carcinoma being the most common (2, 3). In 1978, Barber et al. described the first case unassociated with malignancy (4) in a patient with pulmonary tuberculosis. Furthermore, there have also been several cases associated with benign breast hypertrophy (5), CRST syndrome (6), ichthyosis (7), palmoplantar hyperkeratosis (7), nail dystrophy (7) and hypereosinophilic syndrome (8). In addition, a few cases of EGR have been described in healthy persons (9-11). Neither essential thrombocythemia nor hematopoietic malignancies have been reported among patients with EGR.

CASE REPORT

A 78-year-old woman had suffered from EGR for about 3 years. Annular or serpiginous red bands resembling wood grain appeared and vanished successively



Fig. 1. Annular or serpiginous red bands on the back.

on the back and upper extremities (Fig. 1). The main treatment was a topical corticosteroid ointment. In October 1996, she presented with bilateral submandibular lymphadenopathy and indurated erythemas on the extremities, accompanied by high fever and general fatigue.

Laboratory examination revealed elevated CRP and thrombocytosis (platelet count, $54.8 \times 10^4/\text{mm}^3$). The patient had a 10-year history of hydroxyurea administration owing to presumed essential thrombocythemia. Bone marrow aspiration and biopsy could not demonstrate the latency of hematopoietic malignancies. A CT scan of the chest and abdomen, magnetic resonance imaging of the brain, gallium scintigraphy and gastrointestinal examination did not reveal any evidence of malignancy, but a tuberculin test was positive.

Histopathologically, annular or serpiginous bands revealed superficial perivascular lymphohistiocytic infiltrate with mild acanthosis. Spongiosis was slight. These findings were compatible with EGR. The involved lymph nodes were entirely occupied by tuberculous nodules, which were composed of caseation necrosis with a peripheral rim of epithelioid cells and giant cells, including Langhans' type. The granulomas were surrounded by a wall of mononuclear cells. There were no atypical cells, but the characteristic tubercles. Acid-fast bacilli were not demonstrated by means of Ziehl-Nielsen stain and culture. Polymerase chain reaction (PCR) detection of mycobacterial DNA was positive. The indurated erythemas revealed a lobular panniculitis, which consisted of lymphocytes, epithelioid cells, foam cells, and a few eosinophils. Foreign body and Langhans' type giant cells were present. Leukocytoclastic vasculitis or caseation necrosis could not be seen in the skin. We could not make a definitive diagnosis other than a non-specific lobular panniculitis.

The patient received tuberculostatic therapy consisting of isoniazid and rifampicin from December 21, 1996. Both lymphadenopathy and indurated erythemas soon vanished. Although the serpiginous bands have not appeared since then, annular erythemas continued to appear. After about 2 years, the treatment ended. The patient was free of lymphadenopathy and indurated erythemas, but not the annular erythemas. In September 1999, she developed high fever, general fatigue, and arthralgia. The laboratory findings demonstrated the development of a blastic crisis from essential thrombocythemia. White blood cell count was $115 \times 10^3/\text{mm}^3$. Myelocytes, promyelocytes, and myeloblasts were innumerable increased in the peripheral blood. The bone marrow examination could not be done. CRP and lactic dehydrogenase were high. The patient died within one month despite combination chemotherapy.

DISCUSSION

The clinical appearance corresponded to EGR. Histopathological examination revealed a non-specific,

superficial, perivascular dermatitis, compatible with EGR. At first, no other internal diseases than presumed thrombocythemia could be detected, despite a detailed search. The swollen lymph nodes were occupied by characteristic tubercles including typical caseation necrosis and granulomas. Although Ziehl-Nielsen stain and culture were negative, mycobacterial DNA could be detected by the PCR method. Indurated erythemas revealed a lobular panniculitis including epithelioid cells and Langhans' type giant cells. Although it was not consistent with any kinds of tuberculids, it was thought to be a non-specific panniculitis owing to tuberculosis.

The essential thrombocythemia can transform into either myelofibrosis with myeloid metaplasia or acute myeloid leukemia. It is a clonal hemopathy and belongs to chronic myeloproliferative disorders. The definitive diagnosis of it should be made after the exclusion of another chronic myeloid disorder that mimic its presentation.

At first, we regarded this case as EGR associated with tuberculosis and not any underlying malignancies, because tuberculosis has been reported among patients with EGR. The thrombocytosis was not definitive in this case. Finally, the patient developed a blastic crisis from thrombocytosis about 6 years after the development of EGR when we could make a definitive diagnosis.

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