A 25-year-old Man with Acute Maculo-Papular Rash and Target Lesions

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A 25-year-old student had an erythematous eruption on his face, trunk and upper extremity. Initial maculo-papules evolved to target lesions with a vesicular center surrounded by alternating pale and dark rings. The eruption appeared at the dorsum of the hand and progressed proximally, four days after a flu-like episode, without use of drugs or medicines for a minimum of six-months. The changes were restricted to the skin, painless and mildly itching. More conspicuous lesions appeared on his right scapula, left shoulder, and the dorsum of the left hand (Figure 1). Previously, he was in good health, without a history of allergy or skin or mucosal disturbances. He denied alcoholism, tobacco smoking, use of illicit drugs, and vaccination. His girlfriend had fever, fatigue, pharyngitis and lymphadenopathy shortly before the onset of his actual disease. Physical examination revealed the aforementioned skin changes, in addition to discrete bilateral posterior cervical lymphadenopathy, and a moderate non-tender liver and spleen enlargement. The patient was eutrophic and afebrile, with no ocular, oral, nasal, genital or anal lesions. In addition to routine laboratory tests, he underwent a skin biopsy aiming to clear the diagnosis. Blood determinations revealed leukocytosis, lymphocytosis and 12% reactive lymphocytes. Except for a transient elevation of the aminotransferase levels, biochemical data were normal. Hepatosplenomegaly and lymph node enlargement rapidly regressed to normal, as well as the lymphocyte count. The skin lesions improved gradually and healed in about two weeks.

What is your diagnosis?

A. Behçet’s disease
B. Erythema elevatum diutinum
C. Erythema gyratum repens
D. Erythema multiforme
E. Sweet syndrome

Figure 1
ANSWER to PHOTO QUIZ

**Erythema multiforme minor (EMM)**

**Discussion**

Erythema multiforme is an acute or recurrent eruption due to a hypersensitivity reaction. This condition can be idiopathic, but infections and drugs are the main precipitating factors. EMM was suspected, based on edematous papules and typical target lesions; involvement of less than 10% of body surface area; lack of lesions in mucous membranes; and absence of epidermal detachment. Findings from the skin biopsy were also indicative of EMM (apoptotic keratinocytes, hydropic degeneration of the basal layer of the epidermis, intercellular edema, dermal perivascular lymphocytic infiltrate, and absence of vasculitis). Microorganisms were not detected by cultures or histopathology studies. These laboratory and histopathology data contributed to rule out alternative hypotheses. Differential diagnosis of EMM includes diverse other entities such as dermatitis herpetiformis, necrotizing vasculitis, pemphigoid, pemphigus, serum sickness, systemic lupus erythematosus, and urticaria. As observed in this patient, EMM can heal spontaneously in two to three weeks, but supportive or symptomatic care may be necessary, such as antihistamines and corticosteroids for itching lesions. Clinical and laboratory data led to suspicion of mononucleosis, but the heterophile antibody test and Epstein-Barr virus specific tests were not confirmatory. Serologic tests for cytomegalovirus, herpes simplex, adenovirus, HIV, hepatitis A, influenza A and B, mycoplasma, syphilis and toxoplasma were negative. Although clinical data, aminotransferase and lymphocyte changes strongly suggested mononucleosis, idiopathic EMM was not discarded. Behçet’s disease (BD) is a multisystem inflammatory chronic condition characterized by vasculitis and vascular thrombosis, with conspicuous cutaneous and mucosal changes. BD is related to geographical and genetic factors, in addition to immunological factors. Most cases occur in men (20-40 years) from Turkey, geographical and genetic factors, in addition to immunological factors. The histopathologic data characteristic for the diagnosis of EED are dermal neutrophilic infiltrates and leukocytoclastic vasculitis. Dapsone constitutes the treatment of choice for EED.

Erythema gyratum repens (EGR), is a rare condition characterized by progressively migratory concentric and elevated erythematous pruritic skin lesions, spreading as serpiginous bands with scaly borders over the trunk and extremities. Palmoplantar keratoderma and blood eosinophilia may be observed in some patients. The histopathologic findings are nonspecific and the pathogenesis remains unclear. In the vast majority of cases, EGR occurs associated with pulmonary, esophageal, pancreatic and breast cancers, or mycosis fungoides. This paraneoplastic disorder usually regresses after control of the underlying malignancy.

Sweet syndrome (SS) or acute febrile neutrophilic dermatosis is an acute condition characterized by fever, neutrophilia, and painful erythematous, solid, and bullous lesions affecting the skin and mucous membranes. SS can be drug-induced, as well as associated with Behçet disease, rheumatoid arthritis, erythema nodosum, sarcoidosis, and thyroid diseases. Of note, is the association of SS with hematologic malignancies and carcinomas (genitourinary, breast, gastro-intestinal). Histopathology features of SS include epidermal spongiosis, subepidermal edema and vesicles, dense and diffuse neutrophilic infiltrate in the upper dermis, and absence of vasculitis.

**References**