Asymptomatic, Reddish Papules and Pustules with Hemorrhagic Crust on the Trunk of a Girl: Answer of the Last Issue's Case Question

Bir Kız Çocuğunun Gövdesinde Hemorajik Kurutlu, Kırmızımsı Papül ve Püstüller

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Pityriasis lichenoides et varioliformis acuta (PLEVA) is a cutaneous disorder of unknown etiology and characterized by a generalized eruption of acute onset, consisting of papular lesions that undergo central vesiculation which may ulcerate and resolve with hemorrhagic crusts.[1,2] Although descriptions of this disorder had been made many times previously, term "PLEVA" was first described in 1925 by Habermann.[3]

PLEVA occurs predominantly in the second or third decades of life and it is uncommon in childhood. There is a male predominance.[4] The disease starts on the trunk with erythematous papules that develop crusts, vesicles, pustules or erosions, then spontaneously regresses within a few weeks.[3,4] Varioliform scars and postinflammatory hyper-or hypopigmentation may result.[3] The eruption may be asymptomatic or sometimes accompanied by itching, fever and malaise.[1]

There are three major pathogenic theories of PLEVA: an inflammatory reaction triggered by infectious agents; an inflammatory response secondary to a T- cell dyscrasia and an immune complex-mediated hypersensitivity vasculitis.[3] A number of physicians suspect an infectious etiology such as *Epstein-Barr virus, Toxoplasma gondii* and *Human immunodeficiency virus* since the disease appears rapidly and favors younger patients.[2,3] Several reports have demonstrated clonal T-cell receptor rearrangements from patient specimens, suggesting that PLEVA is a lymphoproliferative disorder involving T-cell subtypes, despite its clinically benign course and the absence of morphologically atypical cells in the skin lesions.[4]

PLEVA is the prototypic disease featuring lymphocytic vasculitis, except that fibrin deposition is not seen. Lymphoid atypia is not a standard feature of PLEVA. All cases of PLEVA show interface dermatitis. There is exocytosis, parakeratosis and extravasation of erythrocytes. Epidermal damage range from intercellular and extracellular edema, to extensive keratinocyte necrosis, vesicles, pustules and ulcers. Immunohistochemical stainings are mostly positive for CD8 and negative for CD4 and CD30.[3,4]

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The diagnosis of PLEVA is made by the correlation of clinical features and lesional histopathology. The most challenging disease is lymphomatoid papulosis. However, it can be easily differentiated by its clinical findings (papules can develop into nodules, tumors, plaques), duration of disease (many years) and histological findings (atypical nonlymphoid cell, CD30 positive) from PLEVA.[3] The differential diagnosis of PLEVA also includes chickenpox, Gianotti-Crosti Syndrome (papular acrodermatitis of childhood), lichen planus, pityriasis rosea, guttate psoriasis, arthropod bite, scabies, disseminated Herpes zoster, drug eruption, primary HIV infection, secondary syphilis, vasculitis and viral exanthems (Table 1).

There are several different therapeutic modalities that have been used in PLEVA. When pruritus is severe, topical corticosteroids and antihistamines may provide symptomatic relief without changing the disease course. In adult patients, administration of methotrexate and oral tetracycline has led to good results; but, these medications are inappropriate for the first-line treatment of children. Erythromycin seems to be the preferred initial treatment for children.[3] However, the data about the response to oral erythromycin in the children with PLEVA is controversial and the response rates varies between 25-73%.[5] In many of these cases, the rash returned if the medicine was tapered too rapidly. Tetracycline has been reported to be of value as well.[3,5] Another oral medication successfully used in the treatment of PLEVA is azithromycin. There are a few reports with good clinical results with the use of oral azithromycin 500 mg/day for three consecutive days every week.[2] In our patient, oral erythromycin 4 x 250 mg/day and topical steroids were administered. After four weeks, no clinical response was experienced by the patient. As a consequence, a new therapy with oral azithromycin (500 mg/day for three consecutive days every week) was administered. After three courses of azithromycin, the eruptions rapidly improved. After eight weeks, the patient completely recovered and the treatment stopped. After two months of follow-up, there was no recurrence (Figure 1). In addition, a number of reports indicate the benefits of ultraviolet (UV) therapy for PLEVA. PUVA and UVB have been used with varying degrees of success.[3]

PLEVA has a variable clinical course characterized by recurrent crops of lesions that spontoneously resolve. Usually, new crops may cease to

TABLE 1: The differential diagnosis of PLEVA with some important diseases.

Disorder	Lesion characteristics	Duration	Histological findings
Lymphomatoid papulosis	Papules can develop into nodules, tumors, and large plaques	Years	Large, atypical non-lymphoid cells, CD30 positive cells
Arthopod bite	Grouped or disseminated 1- to 4-mm urticarial erythematous papules that are markedly pruritic	1-2 weeks	High prevalence of eosinophils in dermal infiltrate, common presence of capillary proliferation
Varicella	Vesicles surrounded by narrow red halos involvement of mucous membranes and face	7-10 days	Tzanck-positive clear vesicles, presence of balloon cells and multinucleated giant cells
Erythema multiforme	Target lesions, predominantly acral distribution	2-3 weeks	Apoptosis of individual keratinocytes, spongiosis and focal vacuolar degeneration, T lymphocytes with exocytosis into the epidermis
Gianotti-Crosti Syndrome	Skin-colored, symmetrically edematous papules, predominantly acral distribution, trunk is usually spared	3-4 weeks	Epidermal acanthosis, hyperkeratosis and a lymphohistiocytic perivascular infiltrate in the dermis with dilation of dermal capillaries

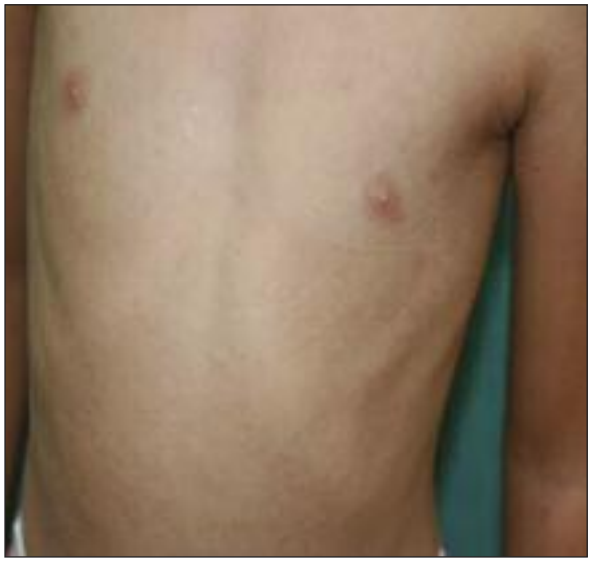

FIGURE 1: The eruptions had almost disappeared after the therapy.

develop after a few weeks, and many cases clear within six months. In general, the immediate prognosis is said to be better when the onset is acute and the lesions in successive crops.[3] Although the conclusion was not confirmed by a subsequent investigation, a smaller study comparing adults and children found that the disease tended to run a longer course in children, with greater extent of lesions, more pigmentation, and poor response to conventional treatment.[5] We report a child with PLEVA unresponsive to erythromycin, and rapidly resolved with three courses of azithromycin. The response of our patient to azithromycin can be explained by improved systemic absorption, enhanced penetration into tissues, and the longer half-life of azithromycin over erythromycin.[2] Of course, the possibility of spontaneous resolution must be entertained. However, the timing of resolution after starting azithromycin treatment after persistence of disease with other treatment is suggestive of the benefit of azithromycin randomized double-blind placebo controlled trials are needed to fully elucidate the benefit of this antibiotic in this skin disease.

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