AUTOIMMUNE CONNECTIVE TISSUE DISEASES

SYNDROMA WELLS, URTICARIAL-VESICULLOUS TYPE WITH POSITIVE LUPUS-LIKE DIRECT IMMUNOFLUORESCENCE TEST

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Background: Wells syndrome is an uncommon inflammatory skin disease of unknown etiology defined by various clinical presentatition (plaque type, annular granuloma-like, urticaria-like, papulovesicular, bullous, papulonodular and fixed drug eruption-like) and granulomatous eosinophilic infiltrates in the dermis.

Observation: A 58-year-old female presented with a one-year history of recurrent episodes, of pruritic red spots and vesicles. Clinical examination revealed large erythemo-edematous plaques, mainly located on the thighs. Annular and targetoid smaller plaques/papules, some with vesicules and hemorrhagic crusts, were mainly presented on the abdomen and flexor side of forearms. On the gingival mucosa, vesicles and erythema were present. Examinations of other systems were normal. Laboratory examination revealed mild Eosinophilia and elevated Reuma Factor. In stool analysis, a small number of Candida species was found. Levels of immunoglobulins, including IgE, were normal. Immunological test, and tumor markers, were within normal limits and there were no laboratory evidence for viral, bacterial and parasitic infection.
Incisional biopsy performed and pathohistological findings comprised of epidermal light acantosis and spongiosis with focal basal vacuolar degeneration, without of intra/subepidermal cleft. In the dermis, perivascular, interstitial infiltration of lymphocytes and eosinophils was present. Direct immunoflurescent tests from lesional and perilesional skin were positive, with discontinuous fine granular deposits of IgG and IgA autoantibody along the basal membrane zone. Lupus band test from fotoexposed skin was negative. In Tzanck test normal keratinocytes were present.
Under treatment with oral Prednisolone 0.5mg/kg daily combined with Hydroxichlorochine 200mg twice daily combined with a topical steroid, patient showed a dramatic improvement of her skin lesions.

Key message: We reported the new clinical presentation of Wells syndroma, urticarial-vesicullous type with lupus-like positive direct immunofluorescent test, which has not been documented yet. In addition, our case suggest that combined therapy with Hydroxichlorochine may be successfully used to prevent relapse after discontinuation of corticosteroid treatment.