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RESEARCH ARTICLE

PEMPHIGUS HERPETIFORMIS TREATED EFFECTIVELY WITH DERMOCORTICOIDS ALONE

El Bakalielkhalil, Keby Edgar, Kerrouch Hasna, Anouar Ilias, Hanafi Tarik, El Azhari Jawad, Amraoui Mohamed, Zemmez Youssef, Frikh Rachid and Hjira Naoufal
Dermatology Department, Mohammed V Military Hospital, Mohammed V University, Rabat.

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Abstract

Introduction: Pemphigus herpetiformis is a rare form of pemphigus.
Case Report: A patient presented with a well-limited erythematous-squamous and crusty dermatosis with a ring-like arrangement, with vesiculo-bullous lesions with a herpetiform grouping of the trunk and 4 limbs. Histological and direct immunofluorescence findings were consistent with pemphigus. The diagnosis of pemphigus herpetiformis (PH) was made. The patient was treated with dermocorticoids with lasting remission.
Discussion: Pemphigus herpetiformis is a particular form of pemphigus characterised by a clinical picture resembling that of dermatitis herpetiformis and an immunopathological appearance similar to that of pemphigus. It generally has a good prognosis. Treatment is based on dapsone and/or general corticosteroid therapy.

Introduction:-

Pemphigus herpetiformisis a particular and rare form of pemphigus, first describedin 1975 by Jablonska et al [1]. It ischaracterised by clinicalsigns of dermatitisherpetiformis and immunohistologicalfeatures of pemphigus. Consequently, the presence of diagnostic difficulties for thisentity.

Case Report :

A 101-year-old womanwith a history of cataractsurgery and valve diseaseundertreatmentconsulted for erythematosquamous and crustylesionsthathad been developing for 5 months, associatedwith a pruriticbullous rash thathadappeared 3 monthspreviously. Clinicalexaminationrevealedwell-limitederythematous-squamous and crusty plaques with an annular pattern, associatedwitherosivelesionswith a vesiculobullous rash with a herpetiformgrouping (Figures 1, 2 and 3), located on the trunk and 4 limbs. Nikolsky'ssignwasnegative and therewas no mucosalinvolvement.

Pathologicalexaminationrevealedacantholysiswithoutnecrosis or cleavage. The dermiswas the site of a polymorphicperivascular and interstitialinfiltrate, essentiallylympho-histiocyticwith the presence of numerouseosinophilicpolymorphs (Figure 4). Direct immunofluorescence (DIF) showed inter-keratinocytedeposits of IgG and C3 in a "meshwork" pattern, predominantly on the lower 2/3 of the epidermis. The diagnosis of pemphigus herpetiformiswas made on the basis of all thesefindings.

The patient was put on verystrong class dermocorticoids. The evolutionwasfavourablewith a lasting remissionafter 9 months.

Corresponding Author:-El Bakalielkhalil
Address:- Dermatology Department, Mohammed V Military Hospital, Mohammed V University, Rabat.

Discussion:-

Pemphigus herpetiformisis a non-classical and rare entity, accounting for 6 to 7.3% of all pemphigus[2]. It canoccuratanyage, with an averageageatdiagnosis of 53 years[3], and can affect both sexes withoutgenderpredilection[2,4,5,6] or with a slightfemalepredominance[3].

Clinically, pemphigus herpetiformisismostfrequently (82% of cases) manifested as erythematousurticarial plaques withvesicles and bullaearound the periphery, whichoftentake on a herpetiformappearance[2,3], sometimesonly as annularerythematousurticarial plaques (8% of cases) or as vesiculobullouslesionswith a herpetiformappearance (9% of cases)[3]. In some cases, otherlesionsmaybeassociated: eczematous plaques [7] or erosions. Pemphigus herpetiformismainly affects the trunk and extremities, rarely the neck, scalp or face. Pruritusisfrequentlyobserved, and issometimessevere. Mucous membranes are rarelyaffected, withinvolvement of the buccal mucosa or labia minora[3]. Nikolsky'ssignisinconsistent.

Biologically, hypereosinophiliaissometimesnoted [3,6].

Histologically, eosinophilicspongiosisisthe mostcharacteristicfeature of pemphigus herpetiformis. Otherfeaturesmaybepresent: spongiosiswitheosinophilic or neutrophilic infiltration, or both - eosinophilic and neutrophilic - subcorneal pustules, or intraepidermalvesicles. Acantholysisisusuallydiscreet, but rarely (4% of cases) canbesevere[3]. An inflammatoryinfiltratedominated by neutrophils or eosinophilsisoftenobserved in the dermis[8]. Direct immunofluorescence (DIF) examination of a peri-lesionalbiopsy shows intra-epidermal inter-keratinocytedeposition of IgG, associatedwith C3 depositionin 40% of cases[3]. IgAdepositsmayalsobeobserved, posingproblems of differentialdiagnosiswithIgA pemphigus, especiallywhenintercellularIgAdepositsalone are present [9,10,11]. The profile of circulatingantibodiestargetingepidermalproteins, mostreported cases of pemphigus herpetiformis are positive for anti-desmoglein 1 but rarely for anti-desmoglein 3 [3].

The cause of pemphigus herpetiformis has not been elucidated. It israrelyassociatedwithotherdiseases, but itsmostfrequentlyreported association iswithanotherautoimmunebullousdermatosis (other types of pemphigus, linearIgAbullousdermatosis and bullouspemphigoid). Otherdiseasesmaybeassociatedwith PH, such as rheumatoidarthritis, psoriasis, dysthyroidism, systemic lupus erythematosus, HIV infection, sarcoidosis, myasthenia and autoimmunehaemolyticanaemia. While rare cases of association withneoplasia have been reported in the literature, in patients withlung cancer, oesophageal cancer and prostate cancer [3]. There have been reports of drug-inducedpemphigus herpetiformisinduced by erdosteine, D-penicillamine, bucillamine[3] and tislelizumab [12].

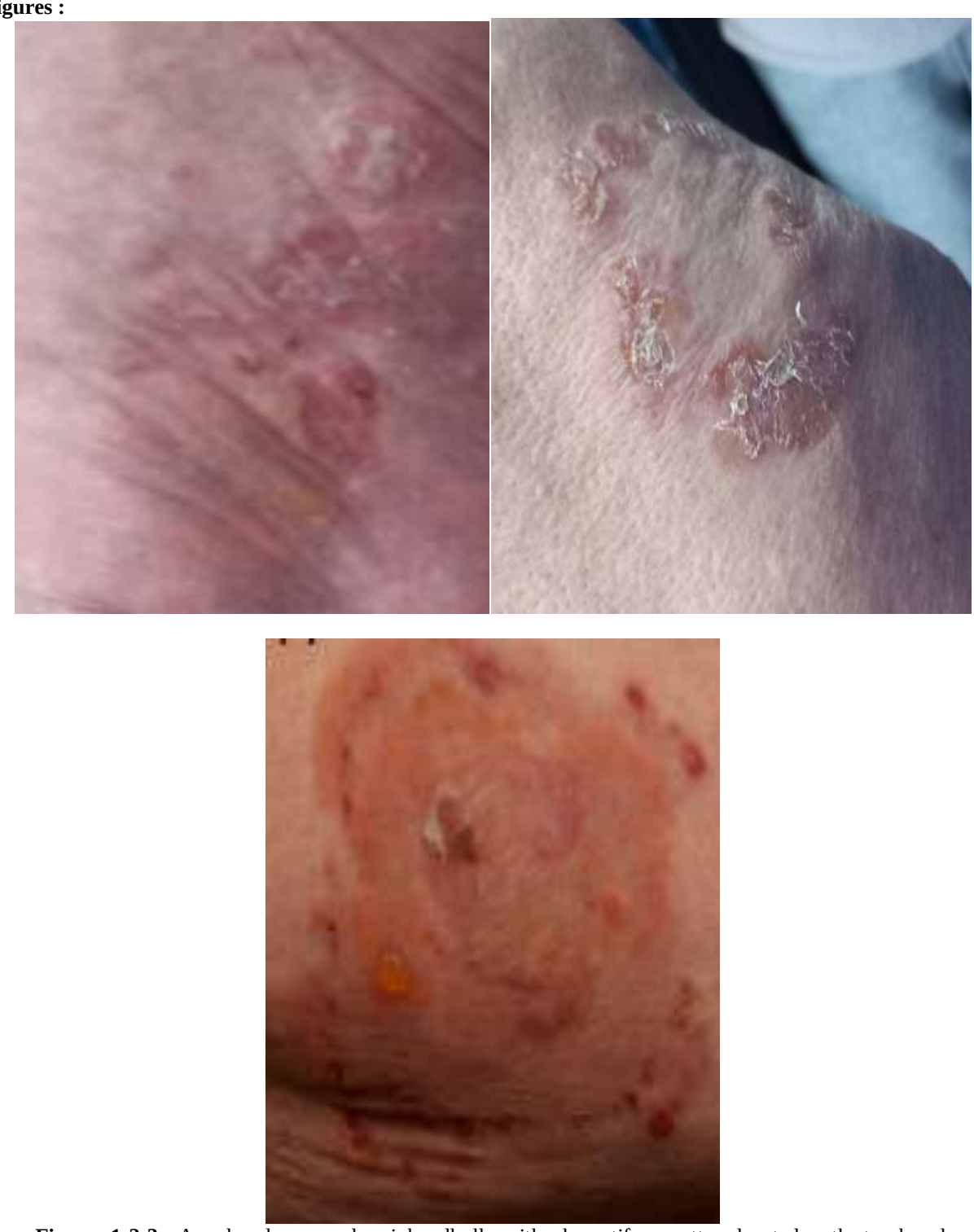
According to a review of the literature byCosta et al[3], pemphigus herpetiformis has generally been successfullytreatedwithdapsone, oral corticosteroids or a combination of the two. Dapsone, because of itsefficacy in reducingneutrophil migration, isconsidered by manyauthors to be the first-line treatment. The doses of dailycorticosteroidsrequired for completeremission are muchlowerthanthoserequired in other types of pemphigus [13]. More intensive treatmentmodalities have been proposed to control the disease in recalcitrant cases, includingazathioprine, cyclophosphamide, intravenousimmunoglobulin, methotrexate, mycophenolatemofetil, cyclosporine, sulfapyridine, minocycline, nicotinamide, doxycycline and leflunomide, rituximab and plasmapheresis [3,13]. In our case, remarkableclinicalimprovementwasobservedwithdermocorticoidsalone. Theywerealsoreported to be effective in another case [14], with a good clinicalresponseat one yearfollow-up.

Pemphigus herpetiformisgenerally has a good prognosis and a rapidlyfavourable course withtreatment, with rare cases (3% of cases) reportingspontaneousremissionwithouttreatment [3]. However, some cases mayprogress to pemphigus foliaceae and rarely to pemphigus vulgaris [6].

Conclusion:-

Pemphigus herpetiformisis a rare form of pemphigus with a good prognosis. Itsclinicalappearanceismisleading, initiallysuggestingdermatitisherpetiformis and making the diagnosisdifficult to establish. DFI is essential to confirm the diagnosis. We report a rare case of pemphigus herpetiformistreatedeffectivelywithdermocorticoidsalone.

Figures :



Figures 1-2-3:- Annular plaques and peripheralbullaewith a herpetiform pattern located on the trunk and extremities.

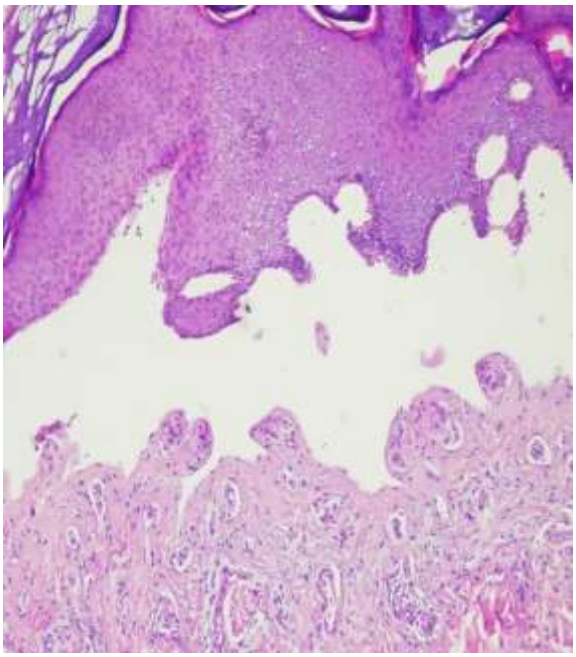


Figure 4:- The histologicalappearance shows spongiosiswith intra-epidermalacantholysis.

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