References

1. Yohn JJ, Weston WL. Topical glucocorticosteroids. Current problems in dermatology 1990; vol.II, 2: 47-9.
2. Bedi TR. Periungual hypopigmentation from intralesional triamcinolone. Ind J Dermatol Venereol Leprol 1977; 43: 270-71.
3. Sharma RC, Sharma NL, Gupta N. Steroid induced perivenous depigmented atrophy of skin. Ind J Dermatol Venerol Leprol 1982; 48: 362-4.
4. Kikuchi I, Horikawa S. Perilymphatic atrophy of skin. Arch Dermatol 1975; 111: 795-6.

CHILDHOOD HERPETIFORM PEMPHIGUS RESPONSIVE TO CO-TRIMOXAZOLE

To the Editor,

Jablonska et al[1] in 1975 suggested the term herpetiform pemphigus for a variant of pemphigus which exhibited the features of dermatitis herpetiformis and of pemphigus. Ingber et al[2] (1986), reported their own cases and reviewed the literature. We report a patient in whom the diagnosis of herpetiform pemphigus was made on the basis of the clinical pattern of an extensive, itchy, blistering, sulfone-responsive disorder suggestive of dermatitis herpetiformis, and histopathology showing intraepidermal bulla and acantholytic cells.

A 6-year-old boy presented with itchy blistering skin lesions since the age of 13 months. A clinical diagnosis of dermatitis herpetiformis had been made elsewhere, and the child started on dapsone to which he showed a moderate response. Dapsone had been withdrawn on several occasions due to fall in haemoglobin. Examination revealed a pale emaciated child with many crusted lesions on the scalp, trunk, gluteal area and limbs. Papulovesicles and a few flaccid vesicles arising on normal skin were interspersed. Flexural lesions were few. Nikolsky's sign was negative. Grouping of lesions was not marked. Genitalia and mucosa were unaffected.

Laboratory investigations indicated a hypochromic microcytic anaemia with a haemoglobin of 10 gm/ml. ESR was 98 mm/hour. Total WBC was 6000/mm^3 : polymorphs 64% and lymphocytes 36%.

The histopathology of a vesicular lesion (biopsied twice) was reported as consistent with herpetiform pemphigus. It showed intraepidermal bullae with eosinophils, neutrophils, lymphocytes and acantholytic cells. A few sections showed the vesicle in a subepidermal location. The dermis had a small perivascular collection of eosinophils and lymphocytes. Immunopathology studies and jejunal biopsy could not be done due to lack of resources.

The child was given co-trimoxazole (single strength) 1 bd to combat the secondary infection, while awaiting biopsy. Three days later the parents reported that the itching had settled dramatically. He was then also started on dapsone, 25 mg/day, and advised a gluten-free diet. There was rapid healing of lesions after which co-trimoxazole was dropped. One week later the patient returned with many active lesions. Blood investigations revealed a mild eosinophilia (TC 10,550; E 11%). Co-trimoxazole was restarted and again the lesions settled rapidly.

It was apparent at this stage that co-trimoxazole was playing a marked role in controlling the disease activity. The dose of dapsone could not be raised beyond 2mg/kg due to his tendency to anaemia. Further attempts to taper and withdraw either drug resulted in a recurrence of lesions within 2 to 3 weeks. Steroid therapy was considered but in his low socio-economic environment its inherent risks were considered greater that the benefits.

This child has now been on a regimen of co-trimoxazole and dapsone for the past 5 years. He continues to develop occasional crusted lesions which heal rapidly. He has had no mucosal lesions while under review. It remains to be seen whether the disease will burn itself out or enter a more malignant phase necessitating aggressive therapy.

The majority of cases of herpetiform pemphigus have shown a closer relationship to pemphigus foliaceus than to pemphigus vulgaris. Most patients have been managed on sulphones with low to absent doses of steroids. One-third of a reported series required high dose immunosuppressive therapy as in pemphigus.[2]

The beneficial role of co-trimoxazole in controlling disease activity was noted incidentally. Probably the sulphamethoxazole part of the combination acts in a way similar to the well-know sulfapyridine. The synergisitc role of trimethoprim cannot be completely ruled out.

Sarojini et al[4] have reported a patient with dermatitis herpetiformis who was maintained on co-trimoxazole and gluten-free diet, when drug-induced peripheral neuropathy forced the withdrawal of dapsone.

Co-trimoxazole could probably be used as an adjunct in sulphone responsive dermatoses where an increase of sulphone dosage is limited by side effects.This, to our knowledge, is the youngest reported case of herpetiform pemphigus.

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References

1. Jablonska S, Chorzelski TP, Beutner EH, et al. Herpetiform pemphigus : A variable pattern of pemphigus. Int J Dermatol 1975; 14: 353-9.
2. Ingber A, Feuerman EJ. Pemphigus with characteristics of dermatitis herpetiformis - A long term follow-up of 5 patients. Int J Dermatol 1986; 25: 575-9.
3. Maciejowska E, Jablonska S, Chorzelski T. Is pemphigus herpetiformis an entity ? Int J Dermatol 1987; 26: 571-7.
4. Sarojini PA, Abraham S, Nair BKH. Dapsone-induced peripheral neuropathy. Ind J Dermatol Venereol Leprol 1988; 54: 207-8.

SCLEREDEMA ADULTORUM : REPORT OF TWO CASES

To the Editor,

Scleredema adultorum is a chronic skin disorder of unknown aetiology which manifests clinically as an acquired non-pitting symmetrical induration of the skin with a peculiar wooden-like consistency. Few cases have been reported in Indian literature.[1,2] We report our experience with two such cases.

Two male children aged 9 years and 11 months respectively presented with 10 and 15 days history of hardening of the skin starting from neck and extending to back, shoulders and arms with a preceding history of febrile episode. The skin was hard, woody and non-pitting without any overlying erythema, atrophic changes, or areas of hypo and hyperpigmentation. Systemic examination did not reveal any abnormality. Haemogram and blood biochemistry were normal. Skin biopsy revealed normal epidermis with markedly thickened collagen bundles in the dermis separated by clear spaces suggesting oedema. Both children were given low dose oral steroids for 2 weeks. The former remained unchanged whereas the latter had complete recovery at 2 months.

Scleredema is characterized b