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EOSINOPHILIC SPONGIOSIS PRECED BY A PHASE OF CLASSICAL PEMPHIGUS VULGARIS

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A 32-year-old female had classical pemphigus vulgaris for 2 years. After a treatment-free remission period of 5 months, she developed itchy, gyrate, erythematous plaques with central clearing. These were occasionally studded with vesicles. Histopathology of such lesions revealed eosinophilic spongiosis.

Key words : Eosinophilic spongiosis, Pemphigus vulgaris.

The term eosinophilic spongiosis (ES) was first coined in 1968[1] for a distinctive histopathologic reaction characterised by spongiosis and intra-epidermal cleft formation in the epidermis and intra-epidermal accumulation of eosinophils which usually precedes classical pemphigus vulgaris and occasionally foliaceus. Similar observations had been made earlier by Perry[2] and Sneddon and Church.[3] It is now believed that it is a harbinger of pemphigus.[4-6] On the basis of positive immunofluorescence for intercellular autoantibodies, cases of ES lacking classical lesions of pemphigus have been considered to be cases of pemphigus.[5,7,8]

Histopathology similar to ES is also classically seen in the vesicular phase of incontinentia pigmenti, pemphigoid and herpes gestationis and, rarely, in the intra-epidermal vesicles of allergic contact dermatitis and arthropod reactions.[9] Recently, Crotty et al[10] reported that of the 71 cases having classical histopathologic features of ES, 36 were bullous pemphigoid, 13 were pemphigus, while the remaining included acute dermatitis, milker's nodule, arthropod reactions, drug reactions, incontinentia pigmenti, cicatricial pemphigoid, herpes gestationis, subcorneal pustular dermatosis, eosinophilic cellulitis and mixed bullous disease. The precise diagnosis of a case having histopathologic features of ES is therefore established on the basis of additional clinical and immunofluorescent findings.[10] However, there may still be some cases having some unclassifiable clinical entity.[7,11]

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Case Report

A 32-year-old woman had clinically classical pemphigus vulgaris for 2 years. Tzanck test was positive. Disease had initially started in the oral mucosa and had subsequently involved the skin. She had had 3-4 exacerbations and remissions during the first year. In the following year it was controlled with 30 mg oral prednisolone per day. This was eventually withdrawn completely over a period of 6 months. After remaining in remission for another 6 months, the patient developed erosions in the oral cavity, followed 2 days later, by multiple, itchy, spreading, gyrate and annular, 1-20 cm sized, oedematous and erythematous plaques with well-defined margins and central clearing but with mild hyperpigmentation. Tense, well-defined, 0.5 to 2 cm sized, vesicles and bullae studded the plaques, especially at their periphery (Fig. 1). The bulla spread sign and Tzanck test were negative. These bullae remained intact for 2-3 days. Some of these lesions were reminiscent of erythema multiforme and some of dermatitis herpetiformis. After one month, the lesions morphologically resembled those of erythema annulare centrifugum. There was no systemic involvement.

Hemoglobin, total and differential leukocyte counts, erythrocyte sedimentation rate, blood sugar, blood urea, serum electrolytes, urine and stools examinations and X-ray chest were within

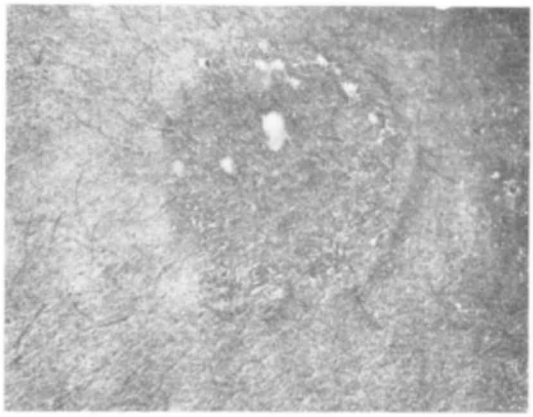

Fig. 1. Annular, erythematous plaque with central clearing and a few tense vesicles.

normal limits. Four biopsies from the lesions clinically resembling DH and erythema annulare centrifugum were essentially similar (Figs. 2 and 3). Spongiotic vesicles and clefts containing eosinophils and occasional acantholytic cells were present below the granular cell layer. There was marked oedema of the upper dermis, and occasional sub-epidermal clefts at the tip of dermal papillae contained eosinophils. The upper dermis in addition had a moderately intense infiltration of eosinophils and histiocytes. In the lower dermis, the infiltrate was perivascular. Direct immunofluorescence revealed

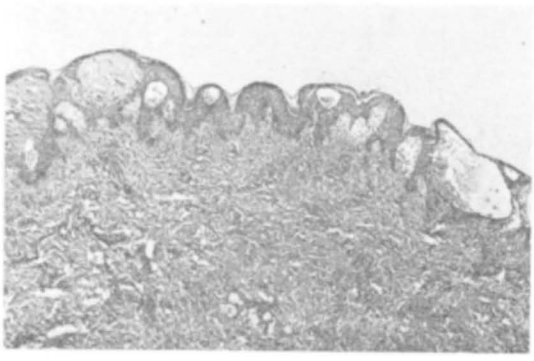

Fig. 2. Intra-epidermal and sub-epidermal clefts in the same section. Upper dermal and perivascular infiltrate (H & E X 10).

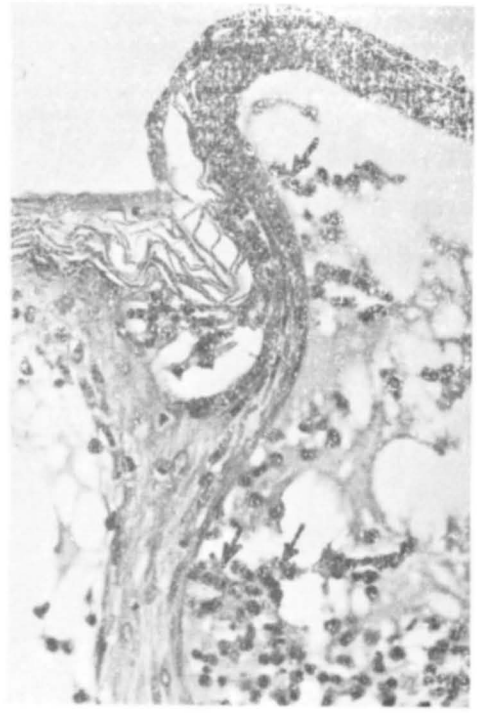

Fig. 3. Intra-epidermal cleft containing eosinophils and occasional acantholytic cells (H & E X 100).

deposition of IgG antibody in the intercellular region of epidermis.

Administration of oral diamino-diphenyl-sulphone (DDS), 300 mg/day gave partial relief. Complete control was achieved after addition of 30 mg prednisolone per day orally.

Comments

Clinically, the lesions of ES have been recorded to resemble DH,[1,3,12,13] bullous pemphigoid, erythema multiforme,[7,8] and erythema annulare centrifugum.[14] In two cases, DH like lesions followed pityriasis rosea like lesions[3,4] before classical lesions of pemphigus appeared. In the present case, two morphologically different types of lesions of ES occurred sequentially. Initially, the lesions were reminiscent of DH and erythema multiforme and subsequently these resembled erythema annulare centrifugum.

Occurrence of lesions of eosinophilic spongiosis in an established case of pemphigus vulgaris is unusual. Usually it precedes overt pemphigus.[4-6] Uncommonly, it has occurred concurently with lesions of pemphigus[1] and rarely, it has followed classical pemphigus.[7]

Histopathologically, epidermal and subepidermal vesicles were seen in the same section. This has apparently not been observed earlier. Sub-epidermal clefts have been observed only in eosinophilic spongiosis associated with bullous pemphigoid.[10] The diagnosis of pemphigus in our case was made on the basis of positive immunofluorescent test. Only one case of pemphigus vulgaris presenting as DH has been recorded so far in the Indian literature.[13] But this case did not reveal eosinophilic spongiosis. Our case appears to be the first report from India.

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