EOSINOPHILIC SPONGIOSIS IN VESICULO-BULLOUS DISEASES
(Reassessment in 93 cases)

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A retrospective review of the skin biopsies of 93 patients having various vesiculo-bullous diseases seen between 1983 and 1985 revealed eosinophilic spongiosis (ES) in 31 cases. Of these, 19 (36.5%) out of 52 patients having pemphigus vulgaris, 2 (20%) of 10 patients having pemphigus foliaceous, one patient having pemphigus vegetans, 3 (15%) of 20 patients with bullous pemphigoid, and 3 (50%) of 6 patients with dermatitis herpetiformis had ES. The severity of eosinophilic spongiosis, graded on an arbitrary scale, was 3+ in 2 cases of pemphigus vulgaris, 2+ in 7 cases of pemphigus vulgaris, 1 case of pemphigus vegetans and 2 cases of dermatitis herpetiformis, while in the remaining it was only 1+. Only 3 patients of pemphigus vulgaris showed distinctive atypical clinical features in the form of erythema multiforme like lesions in 2 cases and erythema annulare centrifugum like lesions in 1 case. In the remaining cases the clinical presentation was not different from those who did not have ES.

Key words : Eosinophilic spongiosis, Pemphigus, Bullous pemphigoid, Vesiculo-bullous diseases.

The term eosinophilic spongiosis (ES) was first coined by Emmerson and Wilson-Jones in 1968,[1] for a distinct histopathologic reaction characterized by epidermal spongiosis associated with focal or diffuse infiltration of eosinophils predominantly in the stratum granulosum.[2] It was earlier considered to be a harbinger of pemphigus,[3-5] but later on it has been seen in several other diseases as well such as dermatitis herpetiformis (DH), pemphigoid, allergic contact dermatitis, arthropod reactions and herpes gestationis.[1,6] The only case of eosinophilic spongiosis reported from India,[7] had pemphigus vulgaris with the clinical features suggestive of erythema annulare centrifugum. To assess the significance of ES, we have re-examined the biopsy specimens received between 1983 and 1985, for the presence of ES.

Materials and Methods

Skin biopsies from patients having pemphigus vulgaris, pemphigus foliaceous, pemphigus erythematosus, pemphigus vegetans, bullous pemphigoid, dermatitis herpetiformis, chronic benign familial pemphigus, subcorneal pustular dermatosis and bullous impetigo were included for review. The minimal criteria to diagnose ES were distinct spongiosis and eosinophils present among the spongiotic cells. The severity of eosinophilic spongiosis was subjectively graded on an arbitrary scale, the grade being 1+ when only a few eosinophils were seen, 3+ when there were a considerable number of eosinophils, and 2+ when it was in between these two grades. All cases having eosinophilic spongiosis were reviewed again for the clinical findings, to look for any atypical clinical features. Peripheral eosinophilic counts in the peripheral blood were noted in the case of in-patients.

Results

Of the 93 cases reviewed, 31 (33.3%) cases (22 males and 9 females, ranging in age from 9 to 54 years) had eosinophilic spongiosis. The disease-wise frequency and intensity of ES are shown in table I. Peripheral eosinophilia was not seen in any of the 12 in-patients studied.

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Table I. The disease-wise frequency and severity of eosinophilic spongiosis (ES).

Disease	Total number of patients		Severity of ES		
	Examined	With ES(%)	1+	2+	3+
Pemphigus vulgaris	52	19 (36.5%)	10	7	2
Pemphigus foliaceous	10	2 (20%)	2	—	—
Pemphigus vegetans	1	1	—	1	—
Bullous pemphigoid	20	3 (15%)	3	—	—
Dermatitis herpetiformis	6	3 (50%)	1	2	—
Chronic benign familial pemphigus	1	1	1	—	—
Subcorneal pustular dermatosis	2	1	1	—	—
Bullous impetigo	1	1	1	—	—
Total	93	31 (33.3%)	19	10	2

Clinically atypical lesions were seen in only 3 cases, all having pemphigus vulgaris. Two of these had erythema multiforme like lesions and the degree of ES was 2+ and 3+ respectively, while the third had erythema annulare centrifugum like lesions, and the degree of ES was 2+. In the remaining cases, the clinical presentation was not different from those who did not have ES.

Comments

Eosinophilic spongiosis is a non-specific but distinctive, and predominantly epidermal histopathologic reaction, seen in several vesiculobullous as well as other diseases. The experience so far indicates that ES is not specific to pemphigus alone as believed earlier, but it has also been demonstrated in bullous pemphigoid, contact dermatitis, arthropod reactions, subcorneal pustular dermatosis, herpes gestationis, eosinophilic cellulitis and milker's nodule.[1] Clinically atypical features however have been seen only in pemphigus and these included lesions similar to DH,[1,5] pityriasis rosca,[3,8] erythema annulare centrifugum,[7,9] and erythema multiforme.[10,11] The clinically atypical phase preceded the classical lesions of pemphigus and the diagnosis of pemphigus at that stage was possible only because of the presence of acantholysis and intercellular immunofluorescent staining.

Crotty et al[1] found ES to be most frequent in bullous pemphigoid (36 out of 71), whereas we have seen the largest number of cases in association with pemphigus. Clinically atypical lesions were seen in 3 cases of pemphigus, 2 had erythema multiforme like lesions while the third had erythema annulare centrifugum like lesions. Though the most severe forms of ES were seen in pemphigus vulgaris and DH, there is no correlation between the atypical features and the severity of ES, because moderate to severe ES has also been seen in patients with classical features of pemphigus.

Peripheral eosinophilia recorded earlier in 22 (61%) of 36 patients with bullous pemphigoid and 6 (46%) of the 13 pemphigus patients having ES,[1] was not seen in any of our 12 patients studied for eosinophilia.

Seah et al[12] and Knight et al[10] have reported good response to dapsone in 1 and 3 patients with ES respectively. Among our patients, out of 12 cases, 4 cases all of pemphigus, responded to 100 mg dapsone twice daily, when 80-120 mg prednisolone a day could not control the disease effectively.

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