CLINICAL PATTERN OF BULLOUS DISORDERS IN EASTERN LIBYA

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A retrospective clinical analysis of 66 patients with various bullous disorders seen over a period of 5 years in Benghazi, Libya showed that pemphigus vulgaris was the commonest disorder, followed by bullous pemphigoid. Other bullous dermatoses were rare. The clinical picture and treatment schedule with follow-up of some patients is presented.

Key words : Bullous dermatoses, Clinical analysis, Libya.

Bullous disorders in which the aetiology is unknown or probably autoimmune constitute only 0.3% of all the skin disorders among patients attending the dermatology clinic in Benghazi, Libya.[1] The present study is a retrospective analysis of 66 patients with bullous disorders seen over a period of 5 years at the Al-Jamahiriya hospital, Benghazi, Libya which is the main referal centre catering to the dermatological needs of patients from the eastern part of Libya.

Materials and Methods

Clinical case records of all the patients admitted with the diagnosis of pemphigus (and its variants), bullous pemphigoid, dermatitis herpetiformis and benign chronic bullous dermatosis of childhood were reviewed. Details of age, sex, duration of disease, extent of skin and mucous membrane involvement, associated disorders, investigations done and treatment administered were noted. In some patients, follow-up records could also be traced.

Results

A total of 66 patients were admitted in the skin wards during this period. There were 32 males and 34 females (Table I). In all cases the clinical diagnosis was confirmed on histopathology. Facilities for immunofluorescence were not available.

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Out of 29 cases of pemphigus seen, 24 had pemphigus vulgaris, 3 had pemphigus foliaceus and 2 pemphigus erythematosus. No case of pemphigus vegetans was seen. Patients with pemphigus vulgaris had classical lesions. Mucous membrane was involved in all the patients. In 4 patients, it was the presenting clinical manifestation. The youngest patient with pemphigus vulgaris was 35-year-old and the eldest 96 years in whom the disease had begun only one year back.

Of 3 patients with pemphigus foliaceus, 1 was a female. The cutaneous involvement in all the 3 was quite extensive and the clinical picture resembled exfoliative dermatitis. However, the history of vesiculation, positive Nikolsky's sign and histopathology confirmed the diagnosis. None of the patients had mucous membrane involvement.

Table I. Frequency of various bullous disorders.

Disease	Number of patients		Total
	Males	Females	
Pemphigus vulgaris	16	8	24
Pemphigus foliaceus	2	1	3
Pemphigus erythematosus	1	1	2
Bullous pemphigoid	7	14	21
Dermatitis herpetiformis	3	5	8
Benign chronic bullous dermatosis of childhood	2	4	6
Subcorneal pustular dermatosis	1	1	2
Total	32	34	66

Pemphigus erythematosus was seen only in 2 cases. Small flaccid bullae with scaling and crusting predominated on the face, the lesions were erythematous and scaly in a butterfly distribution on the face. There was no mucous membrane involvement. Laboratory investigations for lupus erythematosus were negative.

Of 24 patients with pemphigus vulgaris, the disease terminated fatally in 7, the cause of death being peripheral circulatory failure. Other patients have been on low doses of corticosteroids as maintenance.

Bullous pemphigoid was observed in 21 patients. Most of the patients were elderly, over the age of 60 years, the oldest being a 90-year-old male. Characteristic tense bullae on an erythematous urticarial base were seen on the trunk and extremities. The bullae turned haemorrhagic in 7 patients and were arranged in an annular configuration in six. Mucous membrane involvement was observed in 7 patients. In one 67-year-old female, the lesions acquired a vegetative appearance in the axillae and groins, the so called vegetating pemphigoid.[3] No case of cicatricial pemphigoid was seen.

Dermatitis herpetiformis was diagnosed in 8 cases; 5 females and 3 males. Grouped papulo-vesicular lesions with marked pruritus were clinically sufficient to suggest the diagnosis of dermatitis herpetiformis which was confirmed on histopathology. Face and mucous membranes were not involved in any patient.

Benign chronic bullous dermatosis of childhood was seen in 6 children. Of these, 4 were females and 2 males. In all patients, tense bullae with clear fluid were predominantly localized on the lower abdomen and extremities. Face was not involved in any of the patients. Bullae were seen to arise from the previously normal skin. Only 1 patient had involvement of oral mucous membranes. Pruritus of mild degree was observed in two. However, secondary bacterial infection was quite frequent. This was associated with fever and lymphadenopathy. Skin biopsy revealed subepidermal bullae in all cases. Other routine laboratory investigations were within normal limits except for 6% eosinophilia in 1 patient.

Comments

Of various bullous disorders, pemphigus and its variants were the most common. Pemphigus vulgaris was the commonest type and most of the patients were in the age group 45 to 60 years. This is in contrast to a recent report on pemphigus from India[2] where 62% of the patients have the disease by the age of 40 years. However, like the Indian patients, the response to treatment was good in our patients. Systemic corticosteroids in doses ranging from 40 to 80 mg prednisolone had been used initially and on subsequent relapses depending upon the extent and severity of the lesions. After the initial control, the steroids were slowly reduced to a maintenance dose. All our patients with pemphigus vulgaris required higher doses of systemic corticosteroids to control the acute phase. However, in patients with pemphigus foliaceus and pemphigus erythematosus, even the initial dose for control was lower. We have so far not used immunosuppressives or other adjuvents for pemphigus.

Almost all the patients with bullous pemphigoid in the present study were elderly individuals which is in agreement with reports from Western countries.[3] In none of the patients, an association with internal malignancy was observed, though an association with malignant neoplasm has often been described in some cases of bullous pemphigoid.[3] Though pemphigoid patients usually require low doses of corticosteroids for its control, in our patients the dose was as high as 80 mg prednisolone a day along with 100 mg of dapsone. Follow-up has been available in 15 patients; at present they are in remission and off therapy.

Dermatitis herpetiformis appears to be less common in the eastern part of Libya as only 8

cases were observed during a period of 5 years. In a study on dermatitis herpetiformis in Tripoli, which is in the western part of Libya, Shafi et al[4] observed 20 cases in just over a period of 2 years. In 8 of their patients, the lesions were also present on the face and all the patients responded to either dapsone or sulphapyridine. However, in none of our patients, face was affected and no patient responded to dapsone alone. All patients required low doses of systemic corticosteroids in addition, to control pruritus and the eruption. The dramatic response of pruritus to dapsone which is considered as a diagnostic test for dermatitis herpetiformis was also not observed in any patient.

Benign chronic bullous dermatosis of childhood is rather uncommon in this part of the world, only 6 cases were seen. It is impossible to differentiate it clinically from bullous pemphigoid. However, annular erythematous patches with bullae arranged at the periphery of the patches like a string of pearls is considered by some authors to be diagnostic for it.[5] This was helpful to us as a clinical diagnostic sign in only 3 patients. Blood eosinophilia over 5% which is also considered to be diagnostic for benign chronic bullous dermatosis of childhood was observed only in one case. Dapsone was our first drug of choice for this group and in 5 patients, the response was excellent. One patient required low doses of systemic corticosteroids in addition. A 3-year follow-up has been available in 4 patients; they have continued to show fresh blisters though the severity and intensity of the disease has become less.

Only 2 patients of subcorneal pustular dermatosis were observed; these are being reported separately.[6]

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