Autoimmune blistering diseases in Sri Lanka – a review

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The occurrence of blistering diseases in Sri Lanka has been originally documented in 1908. However, any documentation of the incidence and the pattern of autoimmune blistering diseases has not been published yet.

This article is based mostly on the data collected by the author during a period of five years. This includes a two year period at the General Hospital, Kandy (1987-1989) and a three year period at the General Hospital, Colombo South (1992-1995). To avoid misrepresentation, relevant data obtained from the General Hospital, Colombo have also been included.

After making the diagnosis clinically, histological examination of the blisters were done in all the patients. Except in a few patients, diagnosis was confirmed by direct and indirect immunofluorescent studies.

Table 1. Relative incidence of autoimmune blistering diseases

Total No. of Patients	*51*	*%*
Pemphigus	29	59%
Pemphigoid	10	19%
Epidermolysis Bullosa Acquisita	6	11%
Chronic Bullous Dermatosis of childhood	5	9.5%
Herpes Gestationis	1	

Pemphigus is the most common autoimmune blistering disease in Sri Lankan Adults, accounting for 59% of such patients. Annual incidence of Pemphigus is approximately 0.3/100,000 population. There is a clear female prepondarance with a M:F ratio of 9:20. In the majority of the patients, the illness has started in the 5th decade. The youngest with the disease was 08 years old, and the oldest was 70 years of age. No particular ethnic predilection was noted.

Pemphigus vulgaris is the most common variety of Pemphigus, accounting for 86%. Pemphigus foliaceous, erythematosus and vegetans are less common. Endemic, neonatal or drug induced forms of disease were not observed among Sri Lankan patients.

Oral mucosal lesions were found in 75% of the patients with Pemphigus vulgaris. In 25% of the patients, oral lesions preceded the skin lesions by a variable time interval of 1-16 weeks. Ocular lesions in the form of erosions on palpebral conjunctiva were noted in one patient. Two patients had genital erosions. By the time of presentation to a hospital, the skin lesions were widespread in most of the patients.

In the acute stage, prednisolone 60-100 mg/day was necessary for the control of the disease. Complete remissions were observed only in two patients. Most of the patients needed long term maintenance steroids, side effects of which were very common. 38% of the patients developed steroid induced diabetes. 17% became hypertensive after steroid therapy. Severe intercurrent infections were noted in 31%. Incidence of steroid induced myopathy was 14%. These side effects were much less in patients started on intermittent high dose dexamethasone cyclophosphamide therapy.[2]

During the period of follow up 3 deaths occured, two patients died of severe phenmonia. One patient died due to severe attack of varicella.

Bullous Pemphigoid is less common compared to Pemphigus. The relative incidence is 19%. This is in contrast to the incidence in U.K., where Pemphigoid is twice as common as Pemphigus[3]. It affects both sexes equally. In most of the patients the illness has started in the seventh decade. Youngest affected was of 02 years of age. In addition to the typical skin lesions, 30% of the patients had oral mucosal lesions. In the

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acute stage, the illness could be controlled with 30-40 mg of prednisolone/day. Dapsone used on two juvenile patients was not effective. Permanent remissions were noted after a variable period (2 months - 3 years). The clinical course was less alarming when compared to Pemphigus. Perhaps this may be the reason for the less regular clinic attendance noted among these patients. Two deaths observed were due to unrelated causes. Malignancies were not noted in any of the patients.

Cicatricial Pemphigoid is rare. A 60 year old female with the disease has been reported from the General Hospital, Colombo[4]. She has presented with scarring of the eyes and blistering localised to the neck. Skin lesions have responded well to steroids, but the eye damage was permanent

Two patients with Herpes Gestationis have been reported in 1987[5,6]. Both patients were in early thirties, and the disease started in the third pregnancy. Post partum exacerbations were noted in both. No foetal complications were noted. Response to steroids was satisfactory.

Epidermolysis Bullosa Acquisita comprised 11% of all the patients with autoimmune blistering diseases. Most patients presented in the inflammatory phase of the disease, which resembled severe bullous Pemphigoid. Like in the other parts of the world, it's course is markedly variable. It is poorly responsive to steroids. One of the patients developed a pulmonary neoplasm after the onset of the illness.

Out of the IgA Dermatoses, Chronic Bullous Dermatosis of the childhood is the most common is Sri Lanka. It accounts for 85% of the autoimmune blistering diseases in childhood[7]. The average age of onset was 4½ years. Sex incidence was equal. Initial episodes were characterized by tense blisters, which were predominently distributed in perioral and perineal areas, lower abdomen and thighs. During the recurrences and in children whose illness started at an older age the lesions were atypical and mostly papulovesicular. The disease was dapsone responsive.

Adult linear IgA disease and Dermatitis Herpetiformis are found to be rare.

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