

Pemphigus in younger age group in Bangladeshi population

Abdul Wahab¹, MD, Lubna Khondker¹, MD, Jamal Uddin¹, MD, Ishrat Bhuiyan², MD
Shirajul Islam Khan³, MD, Zafrul Islam¹, MD, Rahmat Ali¹, MD, Wahida Afroz¹, MD
Hasibur Rahman⁴, MD

¹Department of Dermatology & Venereology, Banghabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

²Department of Dermatology & Venereology, Shaheed Sohrwardi Medical College Hospital, Dhaka, Bangladesh

³Department of Dermatology & Venereology, Combined Military Hospital, Dhaka, Bangladesh

⁴Department of Dermatology & Venereology, Community Based Medical College, Bangladesh

ABSTRACT

Background: Pemphigus is an uncommonly encountered bullous dermatoses among younger age group. The disease is characterized by autoimmune blistering disorder with remission and exacerbation. Bangladeshi population and highlight any differences and similarities with adult type.

Patients and methods: A cross sectional study was conducted in the department of Dermatology and Venereology in 3 different hospitals in Bangladesh from 2006 to 2010. 40 patients of age group 15-35 years. The patients, suffering from pemphigus diagnosed clinically and confirmed by histopathological examination of skin and DIF. The samples were selected purposively.

Result: In this study, among the 40 cases, pemphigus vulgaris was found in 18 (60%) followed by pemphigus foliaceus 8 (26.67%), pemphigus erythematosus 2 (6.67%) and 2 (6.67%) paraneoplastic pemphigus.

Conclusion: The study showed that among the thirty cases of the younger age group, the most common clinical variant is pemphigus vulgaris, mucosal involvement is the initial presentation and complications are mainly due to steroid therapy and all findings are similar with adult type.

KEY WORDS: Pemphigus, pemphigus in younger age, pemphigus in Bangladeshi population

INTRODUCTION

Pemphigus vulgaris (PV) is an autoimmune disease characterized by acantholysis in the epithelium.¹ Pemphigus vulgaris (PV) frequently begins with oral lesions and progresses to skin lesions.² PV is often difficult to diagnose in the early stages, since the oral lesions are relatively nonspecific.¹ A patient is described who developed skin lesions during follow-up and whose only initial symptom was desquamative gingivitis (DG).² Pemphigus vulgaris is uncommon in adolescence and only ten well documented cases in this age group were found in a recent review.³

Pemphigus is a group of rare autoimmune blistering disorder of the skin and mucous membrane, clinically characterized by flaccid, easily ruptured bullae on normal appearing skin or erythematous base.⁴ Autoimmune bullous skin disorders are rare, potentially fatal disorders of skin and mucous membranes which are associated with IgG or IgA auto antibodies against distinct adhesion molecules of the epidermis and dermal epidermal basement membrane zone, respectively. These autoantibodies lead to a loss of skin adhesion which shows up clinically as the formation of blisters or erosions. In pemphigus, loss of adhesion occurs

Correspondence: Dr. Abdul Wahab, Associate Professor, Department of Dermatology & Venereology, Banghabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh. E mail: wahab_shin@yahoo.com

within the epidermis while in the pemphigoids, linear IgA dermatosis, epidermolysis bullosa acquisita and dermatitis herpetiformis, loss of adhesion takes place within or underneath the basement membrane zone. The autoantigens of these disorders are largely identified and characterized. The diagnosis of autoimmune bullous skin diseases is based on histology and direct immunofluorescence of perilesional skin and the serological detection of auto antibodies by indirect immunofluorescence and recombinant autoantigens.⁵ Two major variants of pemphigus are identified by the level of cleavage within the epidermis by different clinical pattern and by the auto antibodies. Suprabasal clefting is seen in pemphigus vulgaris (PV) and its variants pemphigus vegetans (PVge). Most superficial subcorneal bullae are formed in pemphigus foliaceus (PF) and pemphigus erythematous (PE) and presented as superficial blisters on the seborrheic areas of the body. In addition there may be paraneoplastic pemphigus, drug induced pemphigus, IgA pemphigus. In para-

neoplastic pemphigus, the patient with underlying neoplasms presented with painful mucosal ulceration and polymorphous skin lesions. Although the onset usually occurs in middle aged and older adults. PV and PF also occurs in young adults and children.⁶

PATIENTS AND METHODS

A cross-sectional study was done in the department of Dermatology and Venereology in three different hospitals in Bangladesh for a duration of 2006 to 2010. Thirty patients of 15-35 years both male and female suffering from pemphigus diagnosed clinically and confirmed by histopathologically and immunologically. The patients were selected purposively. (Table 1)

RESULTS

Among the patients, 18(60%) had pemphigus vulgaris, 8(26.67%) had pemphigus foliaceus, 2(6.67%) had pemphigus erythematous and 2(6.67%) had paraneoplastic pemphigus. Among

Table 1 Clinical varieties of pemphigus with sex distribution (N=30)

Clinical Variants	Sex distribution		
	Male	Female	Total
Pemphigus Vulgaris	10 (33.33%)	8 (26.67%)	18 (60%)
Pemphigus Foliaceous	5 (16.67%)	3 (10%)	8 (26.67%)
Pemphigus Erythromatosus	2 (6.67%)	0	2 (6.67%)
Paraneoplastic Pemphigus	2 (6.67%)	0	2 (6.67%)
Total	19 (63.33%)	0	30 (100%)

Table 2 Duration of the disease

Duration	Pemphigus Vulgaris	Pemphigus Foliaceous	Pemphigus Erythromatosus	Paraneoplastic Pemphigus	Total
1-2 month	14 (46.67%)	4 (13.33%)	2 (6.67%)	-	20 (66.67%)
7 months - 1 year	2 (6.67%)	2 (6.67%)	-	1 (3.33%)	5 (16.67%)
1-2 years	1 (3.33%)	1 (3.33%)	-	1 (3.33%)	3 (10%)
Over 2 years	1 (3.33%)	1 (3.33%)	-	-	2 (6.67%)

the different varieties of pemphigus, 66.67% had the duration 1 to 6 months, 16.67% had the duration 7 months to 1 year, 10% had the duration 1 to 2 years and 6.67% had the duration more than 2 years. (Table 2)

Among the patients of pemphigus vulgaris, 10% patients were in the age group of 15-21 years, 20% patients were in the age group of 22-28 years, 30% patients were in the age group of 29-35 years. Among the patients of pemphigus foliaceus, 3.33% patients were in the age group of 15-21 years, 10% patients were in the age group of 22-28 years, 13.33% patients were in the age group of 29-35 years. Among the patients of pemphigus erythematosus, 6.67% patients were in the age group of 29-35 years. Among the patients of paraneoplastic pemphigus, 3.33% patients were in the age group of 22-28 years, 3.33% patients were in the age group of 29-35 years. (Table 3)

Among the patients of pemphigus vulgaris, male-female ratio was 2:1 in the age group of 15-21 years, male- female ratio was 4:2 in the age group of 22-28 years and 4:5 in the age group of 29-

35 years. Among the patients of pemphigus foliaceus, male- female ratio was 2:1 in the age group of 22-28 years and 2:2 in the age group of 29-35 years. And only male patients were in pemphigus erythematosus and paraneoplastic pemphigus. (Table 4)

Fig. 1 showed distribution of the patients by initial presentation of the disease. In 56.8% of patients, the initial presentation of the disease was mucosal involvement and in the remaining 43.2% of the disease initially presented on the skin. Fig. 2 showed distribution of the patients by complica-

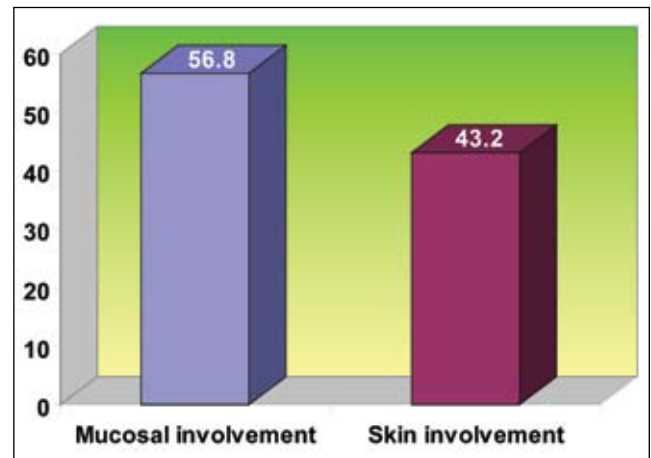


Fig. 1 Distribution of patients by initial presentation of the disease.

Table 3 Age distribution

Age	Pemphigus Vulgaris	Pemphigus Foliaceous	Pemphigus Erythromatosus	Paraneoplastic Pemphigus	Total
15-21 years	3 (10%)	1 (3.33%)	-	-	4 (13.33%)
22-28 years	6 (20%)	3 (10%)	-	1 (3.33%)	10 (33.33%)
29-35 years	9 (30%)	4 (13.33%)	2 (6.67%)	1 (3.33%)	16 (53.33%)

Table 4 Distribution of pemphigus among male and female according to age (N=30)

Age	Pemphigus Vulgaris (N=18)	Pemphigus Foliaceous (N=8)	Pemphigus Erythromatosus (N=2)	Paraneoplastic Pemphigus (N=2)
15-21 years	M : 2 (6.67%) F : 1 (3.33%)	M : 1 (3.33%) F : 0 (0%)	-	-
22-28 years	M : 4 (13.33%) F : 2 (6.67%)	M : 2 (6.67%) F : 1 (3.33%)	-	M : 1 (3.33%)
29-35 years	M : 4 (13.33%) F : 5 (16.67%)	M : 2 (6.67%) F : 2 (6.67%)	M : 2 (6.67%)	M : 1 (3.33%)

tions of the disease. Complications of the pemphigus were due to the steroid therapy and were seen in 16% cases. Among them, most common complication was hyperglycemia, which was seen in 76.19% cases, sepsis was 14% and hypertension was 9.52%.

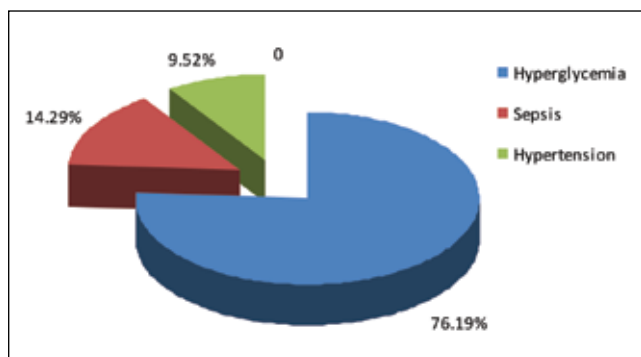


Fig. 2 Distribution of patients by the complications.

DISCUSSION

Among the patients, 18(60%) had pemphigus vulgaris, 8(26.67%) had pemphigus foliaceus, 2(6.67%) had pemphigus erythematosus and 2(6.67%) had paraneoplastic pemphigus. Yazdanfar who conducted a study with two hundred new patients of pemphigus during the 10 year period in Iran, also observed pemphigus as most common form in his study.⁷

Among the patients of pemphigus vulgaris, 10% patients were in the age group of 15-21 years, 20% patients were in the age group of 22-28 years, 30% patients were in the age group of 29-35 years with the mean age of onset was 32 ± 2 years. This finding is not similar to the study finding of Asilian *et al.* they conducted a study of 188 patients of pemphigus vulgaris in Alzahra University Hospital, Iran for duration of 10 year period. Their mean age of onset was 41.1 ± 13.7 years.⁸ This finding is not similar to the study finding of Esmaili *et al.* they conducted a study of 140 patients of pemphigus vulgaris in Razi Hospital, Iran . Their mean



Fig. 3 A 34 year old lady with pemphigus vulgaris showing flaccid bulla and denuded area in right upper extremity and abdomen.

age of onset was 41.5 ± 15.7 years.⁹

Among the patients of pemphigus vulgaris, male-female ratio was 2:1 in the age group of 15-21 years, male- female ratio was 4:2 in the age group of 22-28 years and 4:5 in the age group of 29-35 years. This finding is not similar to the study finding of Asilian *et al* and Esmaili *et al.* Asilian *et al* conducted a study of 188 patients with a 1.6:1 ratio of women compared with men of pemphigus vulgaris in Iran⁸. Esmaili *et al* conducted a study of 140 patients with a 1.59:1 ratio of women compared with men of pemphigus vulgaris in Iran.⁹



Fig. 4 A 30 year old man with pemphigus foliaceus presented with crusting and denuded areas all over the body.



Fig. 5 A 29 year old young lady with pemphigus foliaceus presented with erythematous scaly, crusted lesions on face, trunk & arms.



Fig. 6 A 35 year old woman with pemphigus erythematosus presented with erythematous scaly, crusted lesions on face and upper chest.

Among the patients of pemphigus foliaceus, male-female ratio was 2:1 in the age group of 22-28 years and 2:2 in the age group of 29-35 years. And only male patients were in pemphigus erythematosus and paraneoplastic pemphigus group.

In 56.8% of patients, the initial presentation of the disease was mucosal involvement and in the remaining 43.2% of the disease initially presented on the skin. Asilian et al found mucosal involvement in 74% of patients as initial presentation of the disease and in the remaining 26%, the disease initially presented on the skin.⁸ Esmaili et al ob-

served oral cavity as initial localization in 77.5% cases.⁹ Yazdanfar also noticed mucosal involvement as first manifestation in 60% cases among the total mucosal involvement of 84.5% cases.⁷ Complications of the therapy were seen in 16% cases. Among them, most common complication was hyperglycemia, which was seen in 76.19% cases, sepsis was 14% and hypertension was 9.52%. These findings have a similarity with Yazdanfar who conducted a study with two hundred new patients of pemphigus during the 10 year period in Iran. Yazdanfar showed complications in pemphigus as a consequence of steroid therapy in 6.5% cases. The most common complication was increased blood sugar level in 3.5% cases and mortality was 1% due to sepsis.⁷



Fig. 7 A 31 year old man with paraneoplastic pemphigus having erosive and crusted lesions on lips.

CONCLUSION

The study showed that among the thirty cases of the younger age group, the most common clinical variant is pemphigus vulgaris, mucosal involvement is the initial presentation and complications are mainly due to steroid therapy and all findings are similar with adult type.

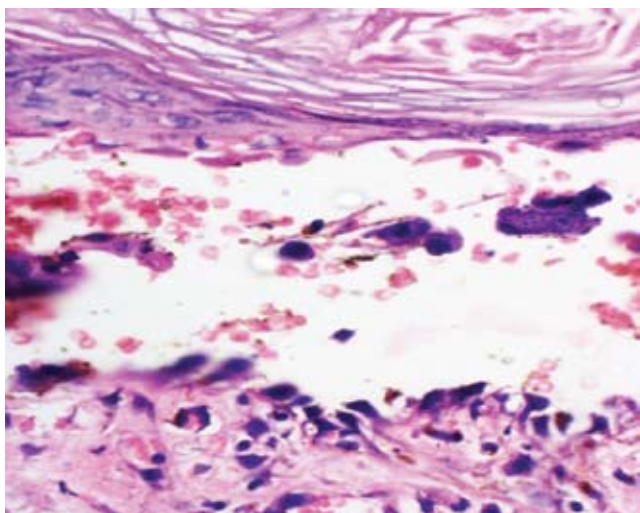


Fig. 8 Histopathological image of pemphigus vulgaris (H&E,x100) showing acantholytic cells and intraepidermal cleft.

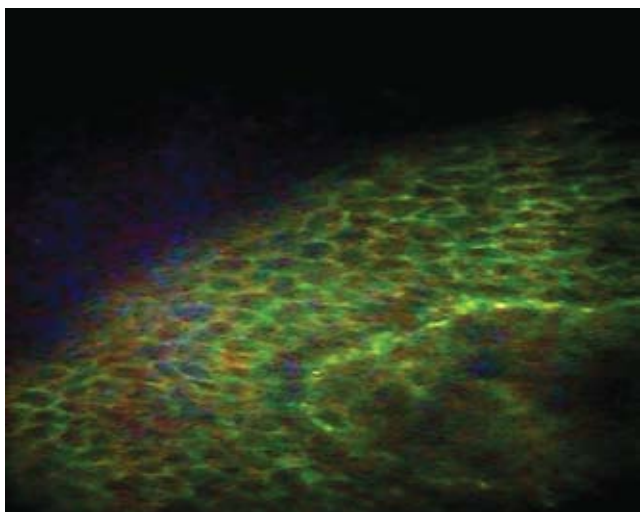


Fig. 9 IgG deposition in chicken wire pattern in DIF of pemphigus vulgaris.

REFERENCES

1. Endo H, Rees TD, Matsue M, Kuyama K, Nakadai M, Yamamoto H. Early detection and successful management of oral pemphigus vulgaris: a case report. *J Periodontol.* 2005 Jan; 76(1):154-60.
2. Disease progression from mucosal to mucocutaneous involvement in a patient with desquamative gingivitis associated with pemphigus vulgaris. *J Periodontol.* 2008 Feb; 79(2):369-75.
3. Harrington I, Sneddon IB, Walker AE. Pemphigus vulgaris in a 15-year-old girl. *Acta Derm Venereol.* 1978; 58(3):277-9.
4. James WD, Berger TG and Elston DM. *Andrew's Disease of the skin-Clinical Dermatology.* 10th edition. USA: Saunders Elsevier; 2006. p. 459-65.
5. Hertl M, Niedermeier A, Borradori L. Autoimmune bullous skin disorders. *Ther Umsch.* 2010 Sep; 67(9):465-82.
6. Stanley JR. Pemphigus. Wolff K, Goldsmith LA, Katz SI, Gilchrest BA, Paller AS and Leffell DJ. *Fitzpatrick's Dermatology in General Medicine.* New York: The McGraw- Hill Companies; 2008. p.459-74.
7. Yazdanfar A. Epidemiology of pemphigus in Hamedan: A 10 year retrospective study(1995-2004). *J Am Acad Dermatol* 2005 May; 52(5): 839-45.
8. Asilian A, Yoosefi A, Faghini G. Pemphigus vulgaris in Iran: epidemiology and clinical profile. *Int J Dermatol.* 2007 Nov; 46(11):1166-70.
9. Esmaili et al. Pemphigus vulgaris in Iran: a clinical study of 140 cases. *BMJ* 2008 Jul. 337.
10. Smolle J, Kerl H. Pitfalls in the diagnosis of pemphigus vulgaris (early pemphigus vulgaris with features of bullous pemphigoid). *Am J Dermatopathol.* 1984 Oct; 6(5):429-35.
11. Mihai S, Sitaru C. Immunopathology and molecular diagnosis of autoimmune bullous diseases. *J Cell Mol Med.* 2007 May-Jun; 11(3):462-81.