Pemphigus in younger age group in Bangladeshi population

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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 foliaceous 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...
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.\textsuperscript{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 it’s variants pemphigus vegetans (PVge). Most superficial subcorneal bullae are formed in pemphigus foliaceus (PF) and pemphigus erythematosus (PF) and presented as superficial blisters on the seborrhic areas of the body. In addition there may be paraneoplastic pemphigus, drug induced pemphigus, IgA pemphigus. In paraneoplastic 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.\textsuperscript{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 foliaceous, 2(6.67\%) had pemphigus erythematosus and 2(6.67\%) had paraneoplastic pemphigus. Among

<table>
<thead>
<tr>
<th>Clinical Varients</th>
<th>Sex distribution</th>
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<tbody>
<tr>
<td></td>
<td>Male</td>
<td>Female</td>
</tr>
<tr>
<td>Pemphigus Vulgaris</td>
<td>10 (33.33%)</td>
<td>8 (26.67%)</td>
</tr>
<tr>
<td>Pemphigus Foliaceous</td>
<td>5 (16.67%)</td>
<td>3 (10%)</td>
</tr>
<tr>
<td>Pemphigus Erythromatosus</td>
<td>2 (6.67%)</td>
<td>0</td>
</tr>
<tr>
<td>Paraneoplastic Pemphigus</td>
<td>2 (6.67%)</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>19 (63.33%)</td>
<td>0</td>
</tr>
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<table>
<thead>
<tr>
<th>Duration</th>
<th>Pemphigus Vulgaris</th>
<th>Pemphigus Foliaceous</th>
<th>Pemphigus Erythromatosus</th>
<th>Paraneoplastic Pemphigus</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-2 month</td>
<td>14 (46.67%)</td>
<td>4 (13.33%)</td>
<td>2 (6.67%)</td>
<td></td>
<td>20 (66.67%)</td>
</tr>
<tr>
<td>7 months - 1 year</td>
<td>2 (6.67%)</td>
<td>2 (6.67%)</td>
<td>-</td>
<td>1 (3.33%)</td>
<td>5 (16.67%)</td>
</tr>
<tr>
<td>1-2 years</td>
<td>1 (3.33%)</td>
<td>1 (3.33%)</td>
<td>-</td>
<td>1 (3.33%)</td>
<td>3 (10%)</td>
</tr>
<tr>
<td>Over 2 years</td>
<td>1 (3.33%)</td>
<td>1 (3.33%)</td>
<td>-</td>
<td>-</td>
<td>2 (6.67%)</td>
</tr>
</tbody>
</table>
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 complication.

Table 3 Age distribution

<table>
<thead>
<tr>
<th>Age</th>
<th>Pemphigus Vulgaris (N=18)</th>
<th>Pemphigus Foliaceous (N=8)</th>
<th>Pemphigus Erythromatosus (N=2)</th>
<th>Paraneoplastic Pemphigus (N=2)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>15-21 years</td>
<td>M : 3 (10%)</td>
<td>F : 1 (3.33%)</td>
<td>-</td>
<td>-</td>
<td>4 (13.33%)</td>
</tr>
<tr>
<td>22-28 years</td>
<td>M : 6 (20%)</td>
<td>F : 3 (10%)</td>
<td>-</td>
<td>M : 1 (3.33%)</td>
<td>10 (33.33%)</td>
</tr>
<tr>
<td>29-35 years</td>
<td>M : 9 (30%)</td>
<td>F : 4 (13.33%)</td>
<td>2 (6.67%)</td>
<td>M : 1 (3.33%)</td>
<td>16 (53.33%)</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Age</th>
<th>Pemphigus Vulgaris (N=18)</th>
<th>Pemphigus Foliaceous (N=8)</th>
<th>Pemphigus Erythromatosus (N=2)</th>
<th>Paraneoplastic Pemphigus (N=2)</th>
</tr>
</thead>
<tbody>
<tr>
<td>15-21 years</td>
<td>M : 2 (6.67%)</td>
<td>F : 1 (3.33%)</td>
<td>M : 1 (3.33%)</td>
<td>-</td>
</tr>
<tr>
<td>22-28 years</td>
<td>M : 4 (13.33%)</td>
<td>F : 2 (6.67%)</td>
<td>M : 2 (6.67%)</td>
<td>-</td>
</tr>
<tr>
<td>29-35 years</td>
<td>M : 4 (13.33%)</td>
<td>F : 5 (16.67%)</td>
<td>M : 2 (6.67%)</td>
<td>M : 1 (3.33%)</td>
</tr>
</tbody>
</table>
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 foliaceous, 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. 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. 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 age of onset was 41.5 ±15.7 years.

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 Iran. 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.

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

Fig. 4 A 30 year old man with pemphigus foliaceous 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.

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

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. Esmaili et al observed oral cavity as initial localization in 77.5% cases. Yazdanfar also noticed mucosal involvement as first manifestation in 60% cases among the total mucosal involvement of 84.5% cases. 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.

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