Parade of Roses at the EADV Poster Presentation - Clinico-epidemiological profile of Pemphigus vulgaris patient in a tertiary care hospital of Bangladesh

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Abstract

Background: Pemphigus vulgaris is a life threatening rare blistering disorder and has the greatest impact on the quality of life of patients. Epidemiological studies of autoimmune blistering diseases rather rare in Bangladesh.

Aim: To evaluate the epidemiology and clinical profile of Pemphigus vulgaris patients.

Materials and methods: This is a descriptive cross-sectional study carried out in the department of Dermatology and Venereology, Chittagong Medical College Hospital, Chittagong, Bangladesh between the period September, 2017 to May, 2019. Seventy six patients of Pemphigus vulgaris diagnosed clinically and confirmed by histopathology and immunology were included in this study.

Results: Among the total 89 patients of autoimmune blistering diseases, 76 (85.4%) patients were of Pemphigus vulgaris, followed by 11 (12.3%) Pemphigus foliaceus, 15 (12.1%) pemphigus erythematosus and 11 (12.1%) Paraneoplastic pemphigus. Among the patients of Pemphigus vulgaris were male and 41 (43.9%) were female. Mean age at disease presentation was 3 months. 83 (93.25%) patients presented with oral erosion. 22 (2.4%) patients presented with flaccid bulla and 2 (2.2%) with erosion. Nikolsky sign was positive in 87 (97.75%) patients.

Conclusion: The study reveals mainly female predominance and oral mucosal involvement in Pemphigus vulgaris. In proper diagnosis and evaluation of the clinical presentation is necessary for management.

Introduction

The word “Pemphigus” derived from the Greek pemphix meaning blister or bubble. Pemphigus vulgaris (PV) is an autoimmune disease in which immunoglobulins (antibodies) target desmocollin proteins to produce intraepithelial mucocutaneous blistering. Desmosome 3 is the major antigen. PV has a worldwide distribution. It is most common in the fourth and fifth decade of life. Usually, PV appears first in the oral cavity, other common sites include groin, scalp, face, axilla and genitalia. Several drugs such as penicillamine, penicillin, captoril, enalapril, rifampicin, nfedipine, probamic and thiopronine has been reported to induce pemphigus.

Methods and Materials

This cross-sectional study was done in the Department of Dermatology and Venereology of Chittagong Medical College Hospital, Chittagong, Bangladesh from September, 2017 to May, 2019. Eighty nine patients more than 8 years old, both male and female, suffering from pemphigus diagnosed clinically and confirmed by histopathology and immunofluorescence (IF) were included in the study. The sampling was purposive. Data were collected through face to face interview by using a semi-structured questionnaire.

Results

Among the total 89 patients of autoimmune blistering diseases, 76 (85.4%) were of Pemphigus vulgaris, followed by 11 (12.3%) Pemphigus foliaceus, 15 (12.1%) pemphigus erythematosus and 11 (12.1%) Paraneoplastic pemphigus. Among the patients of Pemphigus vulgaris were male and 41 (43.9%) were female. Mean age at disease presentation was 3 months. 83 (93.25%) patients presented with oral erosion. 22 (2.4%) patients presented with flaccid bulla and 2 (2.2%) with erosion. Nikolsky sign was positive in 87 (97.75%) patients.

Discussion

Among the 89 patients, PV was 76 (85.4%), PF was 11 (12.3%) and paraneoplastic pemphigus was 1 (1.2%). Among the patients of PV male and female patients were 35(46.1%) and 41(53.9%) respectively and male to female ratio was 1:1.7. In case of PF male and female patients were 3(27.3%) and 8(72.7%) respectively and male to female ratio was 1:2.7. 16.85% patients presented with only mucosal involvement, 9% of the disease initially presented by skin only and 74% showed both mucosal and skin involvement. Among the different varieties of pemphigus, 67.4% had the duration 1 to 6 months and 11.2% had the duration 7 to 12 months and 1.1% had the duration 13 to 18 months and 5.6% had the duration more than 18 months.

Conclusions

The study reveals that 93.25% of Pemphigus vulgaris were presented with oral mucosal involvement. Many of them were misdiagnosed and treated with multivitamin. Proper diagnosis and evaluation of the clinical presentation is necessary for management. Moreover, this study will provide a baseline data for future research and will contribute to proper planning and establishment of blistering disorder patients friendly healthcare services.

Table 1. Distribution of patients by types of pemphigus:

<table>
<thead>
<tr>
<th>Types of Pemphigus</th>
<th>No. of Patients</th>
<th>Percentage</th>
<th>M:F</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pemphigus vulgaris</td>
<td>76</td>
<td>85.6%</td>
<td>1:1.7</td>
</tr>
<tr>
<td>Pemphigus foliaceus</td>
<td>11</td>
<td>12.3%</td>
<td>2.7:1</td>
</tr>
<tr>
<td>Pemphigus erythematosus</td>
<td>1</td>
<td>1.1%</td>
<td>-</td>
</tr>
<tr>
<td>Paraneoplastic pemphigus</td>
<td>1</td>
<td>1.1%</td>
<td>-</td>
</tr>
<tr>
<td>Total</td>
<td>89</td>
<td>100%</td>
<td></td>
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References