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A study of Epidemiology of Pemphigus at a tertiary care centre in North India.

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Abstract:-
Background:*Pemphigus is the commonest variant of autoimmune bullous disorders faced in practice of Dermatology. The basis of disease is autoantibodies targeted against intercellular adhesion molecule of epidermis and it manifests with mucosal and skin involvement. The aim of the study was to assess the profile of pemphigus patients attending our department.*
Materials and Methods:*It was a cross sectional observational study done among patients admitted in department of Dermatology, Venereology and Leprosy. The study was done for a period of 18 months from August 2018 to January 2020. After clinical and histopathological correlation, patients being diagnosed for the first time with pemphigus were included in our study. Follow up cases were left out.*
Results*: A total of 44 patients were included in our study, out of which 38 patients were of Pemphigus Vulgaris and 6 patients of Pemphigus Foliaceus. Majority of patients were male in P.Vulgaris while P.Foliaceus had predominant female cases. 20-40 yrs of age group had most no of cases-22 followed by 18 cases in 40-60 yrs age group. In 77.3% of cases, diffuse type of lesions were found involving more than 10% of body area. 16 patient had comorbidities at time of diagnosis with type 2 diabetes mellitus being most common.36 patients attained remission after treatment whereas 8 patients (18.2%) could not survive. Relapse was seen in 50% of recovered patients.*
Conclusion: *Pemphigus as a disease has a poor prognosis and potential fatal outcome due to commonly associated complicating factors.*
Keywords:- *Autoimmune bullous disorders, Pemphigus Vulgaris, Pemphigus Foliaceus, Autoantibodies, Intercellular adhesion molecule, Histopathological,*

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I. Introduction

Autoimmune vesiculobullous disorders are a heterogeneous group of disorders in which autoantibodies target structure essential for integrity of skin and mucous membrane leading to blister formation.[1] Depending on location of split, these disorders can be classified as intraepidermal and subepidermal.[2]

The word pemphigus comes from Greek word "Pemphix" which means blister. Autoantibodies directed against Desmoglein (Dsgs), which are intercellular adhesion molecules present in the epidermis, leads to pemphigus group of disorder (Dsg 3&1 in P. Vulgaris; Dsg 1 in P. Foliaceus).[3] It is a life threatening autoimmune bullous disease that involves the squamous epithelia and mucous membranes, manifesting as loose blisters and painful erosions.[4]

Pemphigus can be further classified into Pemphigus Vulgaris, its variant Pemphigus Vegetans and Pemphigus Foliaceus, its variant Pemphigus Erythematosus.[5] Pemphigus Vulgaris is the most common type accounting for approximately 80% of cases worldwide.[4]

Although pemphigus forms a small proportion of cases among those attending department of Dermatology; but it has a serious impact on those affected. The morbidity and mortality associated with the disease is troubling.

The study was done to assess the clinical and epidemiological profile of pemphigus patients and to analyse the association of co-morbidities.

II. Materials And Methods

Our study was a cross sectional observational study conducted from August 2018 to January 2020 for a period of 1.5 years. It was carried out in In-patient Department of Dermatology, Venereology & Leprosy of our tertiary care centre.

Irrespective of age and sex, all patients admitted with a diagnosis of pemphigus group of disorders after clinico-histopathological correlation were included in our study. Only patients diagnosed for the first time were considered. Follow up cases of previously diagnosed pemphigus were excluded from the study. Informed consent was taken from patients before inclusion in study. Routine blood investigations like complete blood count, blood sugar level, liver function tests, renal function test, serum electrolytes & total protein were done for each patient.

The study population was studied for age, gender, site and extent of lesions. Patients with more than 10% involvement were taken as diffuse pemphigus and those with 10% or less involvement were categorised as limited pemphigus.[4] Cases which were complicated with co-morbidities were analysed. Course of disease in terms of remission, relapse and death was documented.

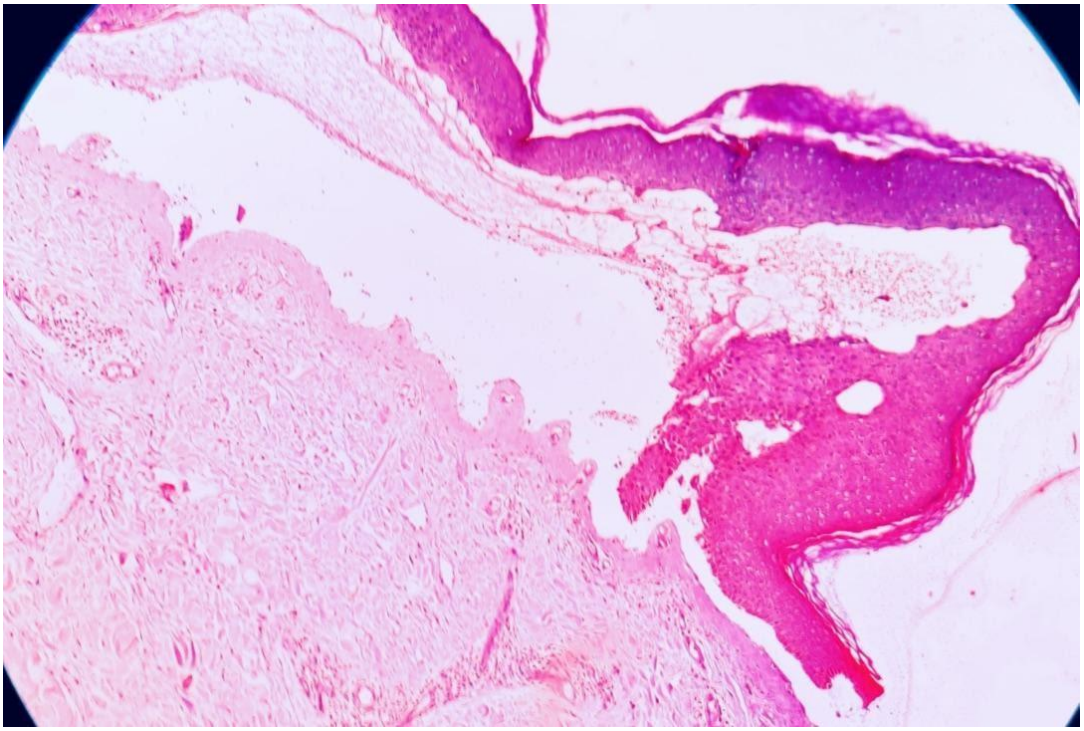


Figure 1-Photomicrograph showing suprabasal split in case of Pemphigus Vulgaris (H&E Stain).

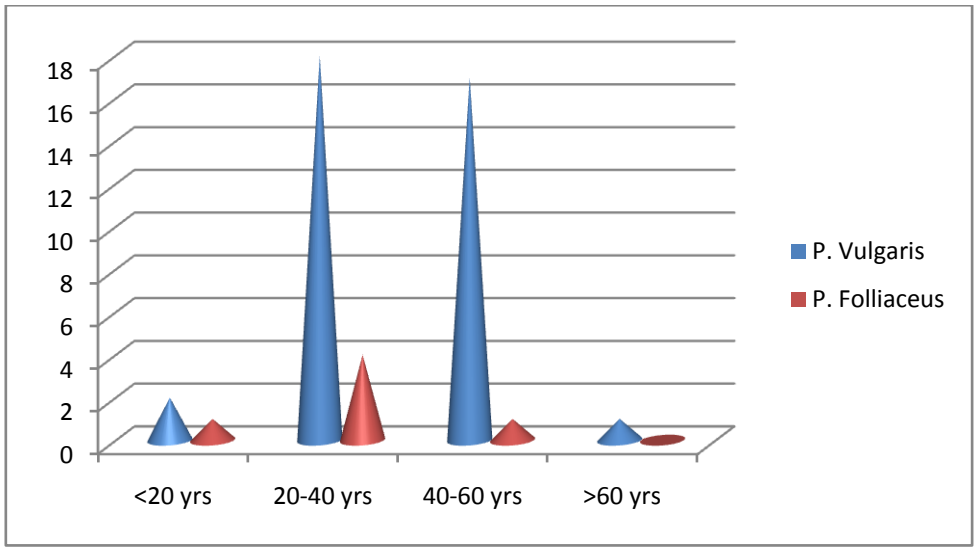
III. Results

Our study comprised of 44 cases, out of which 38 cases were of Pemphigus Vulgaris and 6 cases of Pemphigus Foliaceus. Among 38 cases of Pemphigus Vulgaris majority were male – 21 (55.3%) & 17 females (44.7%). Pemphigus Foliaceus had female predominance – 4 cases (66.66%) and only 2 male cases. In total the predominant gender found was males- 23 (52.3%) and 21 females (47.7%). M:F ratio was 1.1:1.

	P. Vulgaris	*P.Foliaceus*	*Total*
MALE	21	2	23
FEMALE	17	4	21

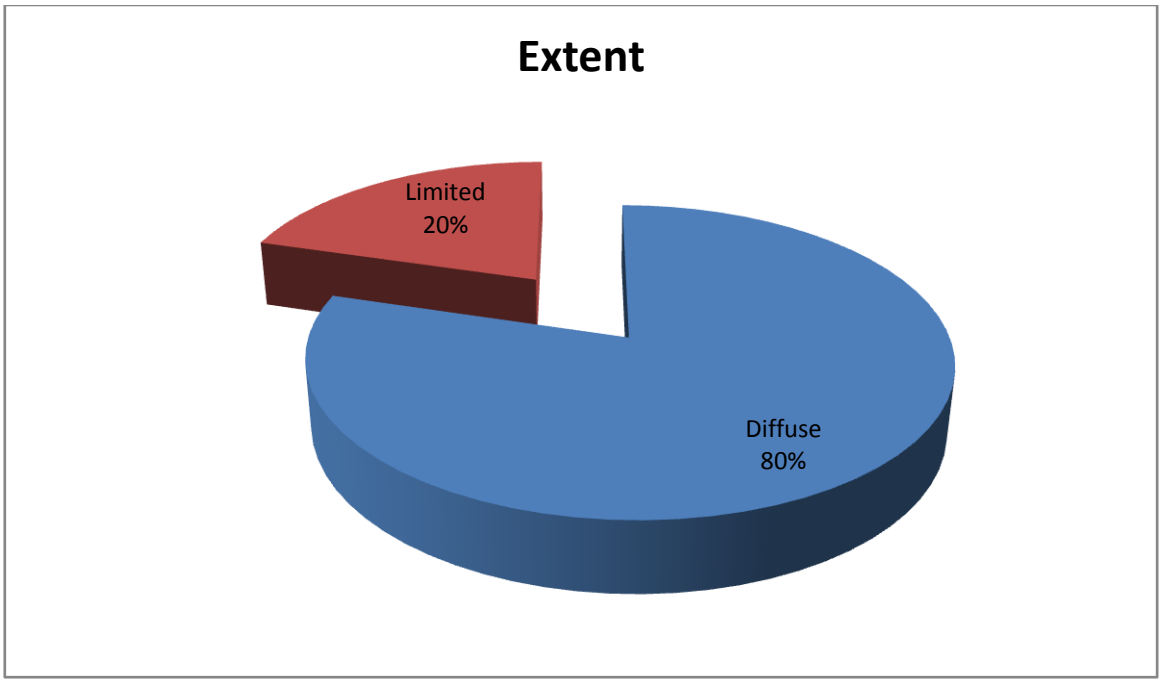
Table 1- Gender distribution among pemphigus patients.

Most of the patients of pemphigus belonged to 20-40 yrs age group – 22 (50%) followed by 18 patients in 40-60 yrs age group (40.9%). 3 patients were less than 20 yrs of age while one patient was more than 60 years of age. The mean age of onset in our study was 35.31 yrs.



Bar 1- Number of cases of pemphigus in different age group.

Based on extent of body area involved, patients were divided into limited and diffuse pemphigus. 35 patients (79.6%) had diffuse pemphigus while 9 patients had 10% or less body area involvement.



Pie 1-Extent of involvement seen among cases.

Oral lesions were seen in all cases in our study. In majority of cases, mean interval between mucosal and dermal involvement was 1-2 months.

Figure 2-A patient of pemphigus vulgaris with significant morbidity due to oral involvement

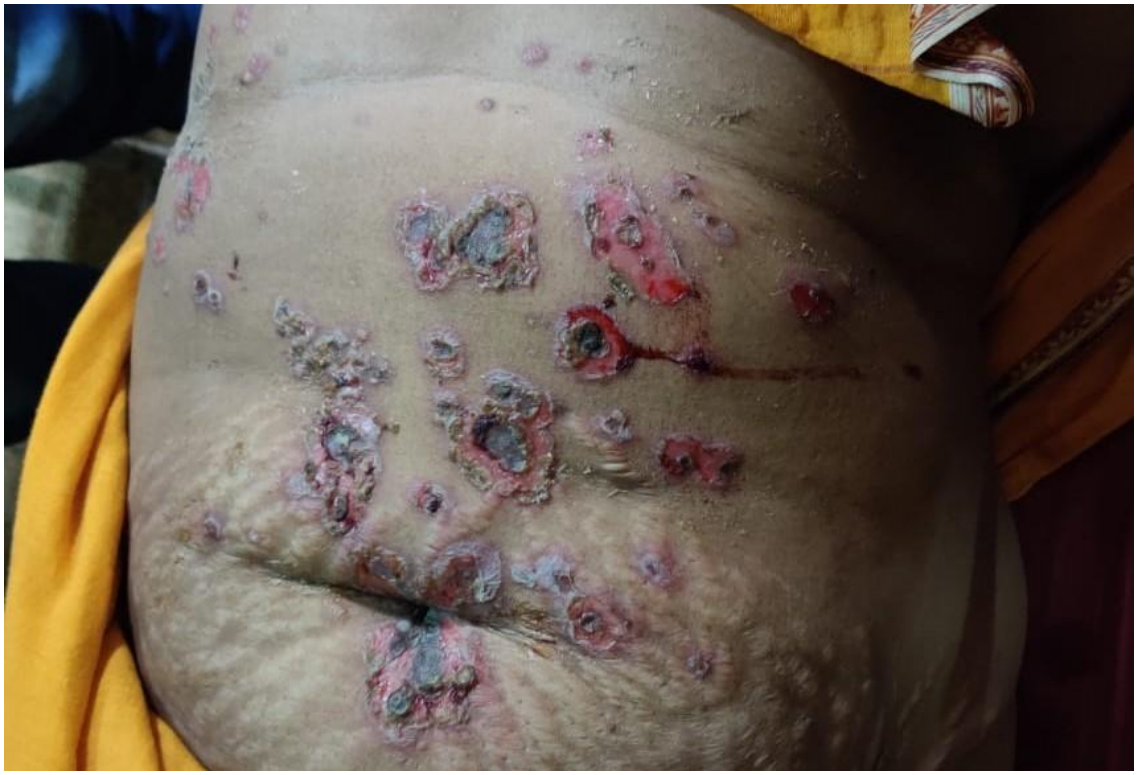


Figure 3- A pemphigus vulgaris patient with multiple erosions over trunk,associated with crusting.

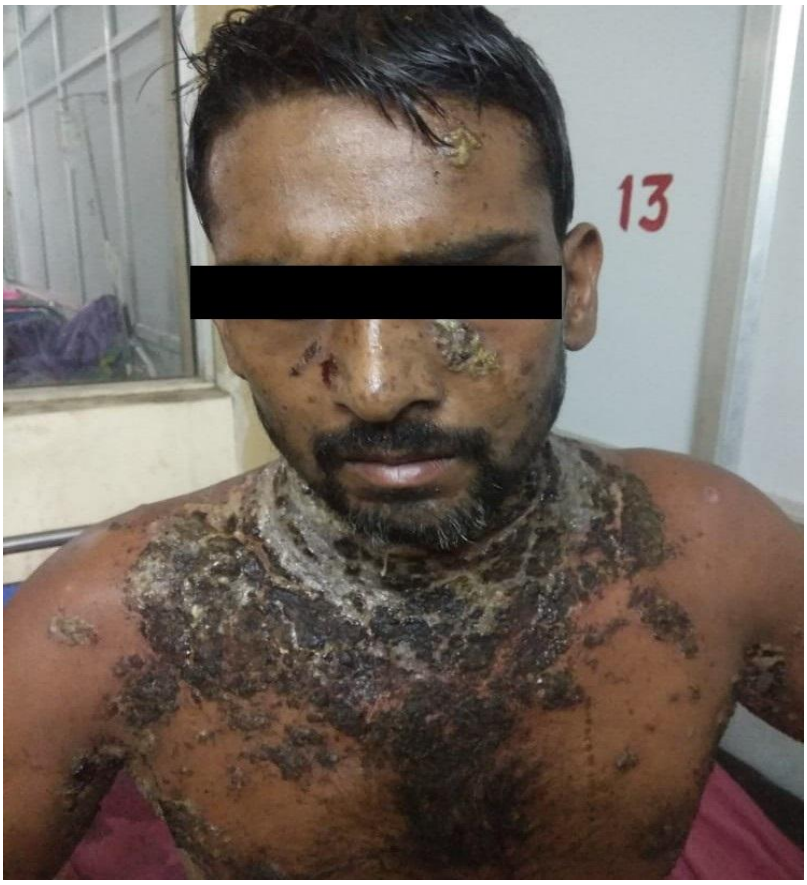
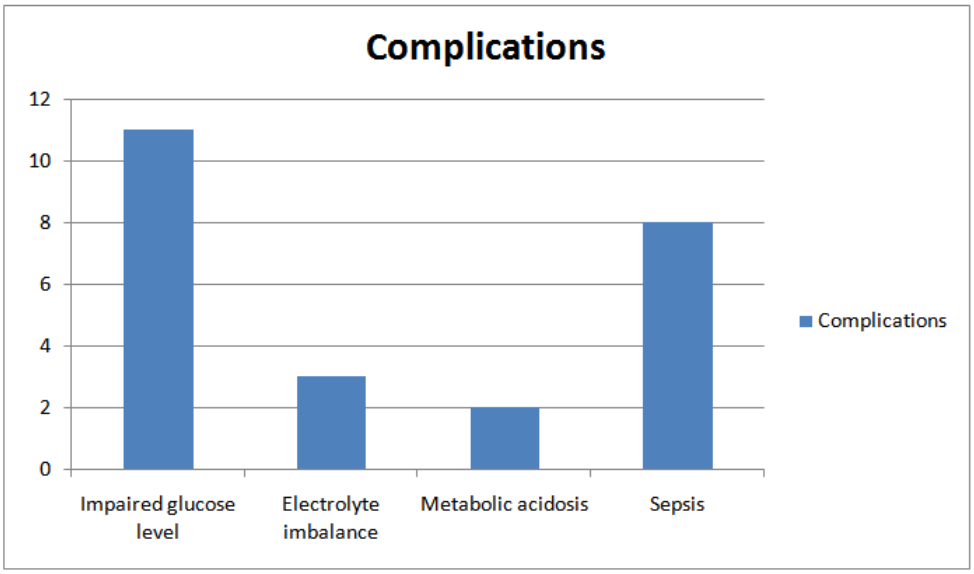


Figure 4- Crusted lesion in typical distribution in a case of Pemphigus Foliaceus.

Around 16 patients (36.4%) in our study had co morbidities at time of diagnosis. Type 2 diabetes mellitus was most commonly encountered in 13 patients (11 patients had diabetes alone while 2 patients had both diabetes and hypertension), while 5 patients had hypertension (3 patients had only hypertension and 2 patients had both diabetes and hypertension). All 13 patients were on medications for type 2 diabetes mellitus but only 8 patients had satisfactory glycaemic control (HbA1c <6.5).

During the course of disease, various patients developed complications. 11 patients with normal blood sugar level previously started showing impaired glucose control. Around 8 patients developed sepsis. Electrolyte imbalance was seen in 3 patients while 2 patients developed Metabolic Acidosis.



Bar 2-Complications encountered during the course of the disease.

Among our study population, 36 patients attained remission (30 cases of P. Vulgaris and 6 cases of P. Foliaceus). However relapse was seen in 18 of these cases (17 cases of P. Vulgaris and 1 case of P. Foliaceus). Almost all these patients relapsed within 2 months. 8 patients succumbed to the disease. 6 of these 8 patients had some complicating factors or comorbidities.

IV. Discussion

Two major types of pemphigus were recorded in our study – Pemphigus Vulgaris & Pemphigus Foliaceus. A total of 44 patients were included in our study of which majority were Pemphigus Vulgaris (86.4%).

In literatures incidence of pemphigus is reported between 0.5-1.62/10^5/year.[6,7] A high incidence of pemphigus is seen in some ethnic groups namely Ashkenazi Jews, Mediterranean and Japanese.[8] A study by Chams et al showed an incidence of 1 over whole of Iran.[9]

M:F ratio among pemphigus patients in our study was 1.1:1.This was similar to finding of Arya et al who also showed a higher male subjects.[10]Our study finding was also supported by finding of study done in Saudi Arabia where the ratio was 2.2:1.[11] However a study done in India showed a female predominance (1:1.16).[12] Similarly Amin et al also found higher no. of females.[13]

Majority of cases in our study belonged to 20-40 yrs age group (50%). A study by Kambil et al showed 77.78% of pemphigus patients belonging to 21-50 yrs age group.[14] The mean age of onset in our study was 35.31 yrs which was lower than previous studies done in Africa (46 yrs) ; Saudi Arabia (43 yrs).[6]

34 patients (77.3%) had diffuse involvement (>10% area involvement) . This draws attention to the ignorance prevalent in the community regarding the disease. It was seen that many cases, due to superstitions and cultural norms, were first taken for some local remedies before bringing to specialised centres. This resulted in significant loss of time in proper diagnosis and initiation of treatment.

According to studies, the interval between mucosal and dermal involvement as 6-9 month in India and 5-12 months in Croatia.[7] However in our study, in majority cases the mean interval was 1-2 months.

Systemic corticosteroids has been the mainstay of treatment despite advent of many newer treatment modalities. During the course of this disease, the treating doctor is often faced with several complications either as a result of acute skin failure or due to adverse effects of treatment. In our study complications were encountered in 54.5% cases.

Relapse was seen in 18 cases (50% of successfully treated cases). The main reason for relapse was poor compliance of patients, since treatment offered are long term and requires regular follow up. Mortality was seen in 8 cases, all of which were of Pemphigus Vulgaris. Interestingly, 6 out of these 8 patients had associated co morbidities like impaired glucose control seen in 3 cases (2 – known diabetic & 1 developed during course of treatment), metabolic acidosis seen in 2 cases & septic shock in 1 case.

LIMITATIONS

The study, being a hospital based study, may or may not be a true reflection of the disease in the community. Immunofluorescent studies could not be done for our cases due to limited resources.

V. Conclusion

Pemphigus Vulgaris is the most common variant encountered. The disease, affecting both skin and mucosa, is very troublesome for the patients. Poor outcome associated with the disease is further complicated by associated complications both disease & treatment related. With better awareness among general public, early diagnosis and treatment may help us push the prognosis towards a more favourable side and reduce relapses.

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