

A CLINICAL STUDY OF 85 CASES OF PEMPHIGUS

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Summary

A clinical study of 85 cases of Pemphigus observed over a span of 12 years from 1961 to 1972 at Rajendra Hospital and Medical College, Patiala, Punjab is reported. 49 cases were of age group of 11-45 years. The youngest case seen was of 13 years of age.

Pemphigus vulgaris, constituting the single largest group of 83 cases, was the commonest type observed in the present study. Majority of the cases had moderate severity and were asymptomatic.

Histopathological observations were made in 32 cases; 31 of Pemphigus foliaceus and 1 of Pemphigus erythematosus.

Pregnancy in cases of Pemphigus vulgaris seems to cause a mild flare up of the lesions without having much adverse effect on the foetus.

Causes of relapses in the disease are briefly discussed. ✓

Pemphigus is a rare bullous disorder characterised by vesiculo-bullous eruptions in skin and mucous membrane. The condition remains fatal inspite of best treatment. In 1808, Robert Willian¹ described this condition under the title "Pompholyx Diutinus" in his essay on skin diseases. Civatte² and Tzanck³ established the histopathological criteria for diagnosis of this disease. Clinical studies on Pemphigus have been reported by various authors namely Gellis and Glass⁴ Combs and Canizares,⁵ Director⁶ Saunders et al⁷ Lever and White⁸. From India the notable studies are relatively few. A study of 34 patients of Pemphigus was reported by Kandhari and Pasricha⁹. Ambady¹⁰ reported on 21 cases of Pemphigus vulgaris; Fernandez et al¹¹ have reported on 100 cases of pemphigus. The present study is aimed at giving our experience of this disease in 85 cases of Pemphigus.

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Material and Methods

The present study is based on clinical evaluation of 85 cases of Pemphigus seen in Rajendra Hospital and Medical College, Patiala, Punjab over a period of 12 years from 1961 to 1972. 83 cases of Pemphigus vulgaris, 1 case of Pemphigus foliaceus and 1 of Pemphigus erythematosus were observed during this period.

Observations and Discussion

The incidence of sex, age and occupation is tabulated in Table 1 and 2. There were 83 cases of Pemphigus vulgaris, 1 case of Pemphigus foliaceus and 1 case of Pemphigus erythematosus. No case of Pemphigus vegetans has been seen by us. Both cases of Pemphigus foliaceus and Pemphigus erythematosus presented with bullous lesions resembling Pemphigus vulgaris. In pemphigus foliaceus case mucous membrane of the mouth was involved besides skin. In Pemphigus erythematosus lesions were present on the face only without involvement of mucous membrane.

TABLE 1

Showing sex incidence in 85 cases of Pemphigus

Type of Disease	Male	Female
Pemphigus vulgaris	49	34
Pemphigus foliaceus	—	1
Pemphigus erythematous	—	1

TABLE 2

Showing Age incidence in 85 cases of Pemphigus

Age in years	Type of Pemphigus & Number of Cases		
	Pemphigus vulgaris	Pemphigus foliaceus	Pemphigus erythematous
0—10	—	—	—
11—15	3	—	—
16—20	3	—	1
21—25	3	—	—
26—30	4	—	—
31—35	11	—	—
36—40	15	—	—
41—45	8	1	—
46—50	14	—	—
51—55	13	—	—
56—60	5	—	—
61—65	3	—	—
66—70	1	—	—

Majority of the cases both male and female belonged to lower-socio economic group. The female cases consisted of 33 housewives coming from poor socio-economic and one from upper socio-economic status. The occupation of majority of the males (26 cases) was farming. Other cases had their occupation as shopkeepers 13, masons 2, police constable 1, students 2. Only one male and one female belonged to upper socio-economic group. Since cases coming to the hospital belonged to Punjab, being an Agricultural State, hence the occupation of the majority of cases was farming.

There are 49 males and 34 females cases of Pemphigus vulgaris, 1 female case of Pemphigus foliaceus and 1 female of Pemphigus erythematous.

The male to female ratio in the present series is 1.4:1. The disease seems to affect male and female equally although in the present study there was a slight preponderance of males over females. This could be explained on the preponderance of male population over the female population in Punjab State as such and also on the larger number of males attending the hospital. Our findings are in agreement with those of Desai et al.¹¹

The age incidence has varied from 13 years to 68 years, the age indicated for each patient being the age at which the disease manifested itself first. The maximum number of cases were seen between the age groups of 36—40 and 46 to 50 years. 3 cases were seen between the age group of 11-15 years and another 3 cases in the age group of 16-20 years. The youngest patient was seen at the age of 13 years. Our findings are in agreement with other Indian observers like Kandhari and Pasricha⁹ and Fernandez et al¹¹. Unlike western countries Pemphigus vulgaris is seen in a younger age group of Indian patients.

TABLE 3

Showing occupation of cases of Pemphigus,

Status	Occupation of patients	Type of Pemphigus & No. of Cases		
		Pemphigus vulgaris	Pemphigus foliaceus	Pemphigus erythematous
Lower socio-economic	House-wife	33	1	1
	Farmers	26	—	—
	Shopkeepers	13	—	—
	Masons	2	—	—
	Police constable	1	—	—
	Washerman	1	—	—
	Clerks	2	—	—
	Factory worker	1	—	—
	Students	2	—	—
	Upper socio-economic	House wife	1	—
Business man		1	—	—

Symptomatology of Pemphigus

Majority of the cases of Pemphigus vulgaris presented are with involvement of skin and mucous membrane especially of the mouth. Lesions were observed mostly on the scalp, face, trunk, axillae and groins. Most of the cases were asymptomatic and presented with blister like eruption on an apparently normal skin or multiple ulcerated areas, which were crusted in some. One case presented with maggot infection of the scalp. Bullous lesions appeared elsewhere on the skin during the subsequent course of illness. Distribution of lesions at the time of presentation is shown in Table No. 4. 69 cases presented with

TABLE 4
Showing the distribution of lesions at the time of presentation.

Site of Lesions	Type of Pemphigus & No. of Cases		
	Pemphigus vulgaris	Pemphigus foliaceus	Pemphigus erythematosus
Involvement of mucous membrane	2	—	—
Involvement of skin only	12	—	1
Involvement of both skin and mucous membrane	69	1	—

involvement of both skin and mucous membrane, 12 cases presented with skin lesions alone, only in 2 cases mucosal lesion were the first to bring the patient to the hospital. Out of cases with mucosal involvement one case presented with conjunctival and oral lesions, 2nd case presented with involvement of the mucous membrane of the mouth alone. None of the cases showed any lesion on the palms and soles. Larynx was not involved in any of the cases of present study. Mucous membrane involved in order of frequency were oral mucous membrane,

nasal mucous membrane, genital mucous membrane and conjunctival. Conjunctival involvement seems to be very rare in our experience and it was observed in only 1 case. Cases with involvement of the mucous membrane of the mouth complained of difficulty in mastication and dysphagia. No intact bullae were observed in lesions of the mouth. Cases presenting with longer duration of the disease showed marked loss of weight and anaemia. Fever and toxæmia were seen in severe cases of Pemphigus vulgaris. Patients with Pemphigus vulgaris developed pyrexia due to complications like massive secondary infection, pulmonary tuberculosis or pleural effusion during the course of illness. Symptomatology of our cases is in agreement with the observations of Kandhari and Pasricha* and those of Fernandez et al¹¹. However, we did not notice any itching or burning sensation in our cases.

A solitary case of Pemphigus foliaceus was seen in a woman of 45 years. Clinically the case resembled Pemphigus vulgaris, but histopathological examination revealed Pemphigus foliaceus. In case of Pemphigus erythematosus the lesions were confined to the face only. Nikolsky's sign was found to be positive in the majority of the cases during the active phase of the disease. In one case this sign was so conspicuous that the patient herself complained that her skin peeled off with the slightest friction. 10 cases of pemphigus vulgaris presented with severe involvement and the remaining 73 with moderate involvement. The case of Pemphigus foliaceus was moderate in severity whereas Pemphigus erythematosus was relatively mild in intensity.

Duration of the disease is tabulated in Table No. 5. Majority of the cases presented themselves within 1 to 12 months of the onset of the disease; 43 cases within 1 to 6 months and 30 cases

within 7-12 months. 6 cases presented within 13 to 24 months and 4 cases within 25 to 36 months of the onset of the disease.

TABLE 5
Showing the duration of the disease at the time of presentation.

Time	Type of Pemphigus and Number of Cases		
Duration of disease in months	Pemphigus vulgaris	Pemphigus foliaceus	Pemphigus erythematosus
1-6 months	43	—	1
7-12 months	30	1	—
13-24 months	6	—	—
25-36 months	4	—	—

Duration of stay in the hospital of admitted cases varied with the intensity of the disease. Longest stay was of 180 days and shortest stay was of 28 days. The average stay in the hospital before a patient could be sent home on maintenance dose of prednisolone was 56 days.

Pregnancy and Marital Status in Pemphigus Cases

Out of 36 female cases, 1 case of Pemphigus erythematosus and 2 cases of Pemphigus vulgaris were unmarried. Rest of the females consisting of 32 cases of Pemphigus vulgaris and 1 case of Pemphigus foliaceus were married at the time of presentation with the disease. 3 cases of Pemphigus vulgaris who were on a maintenance dose of Prednisolone ranging from 12-20 mg., became pregnant and were admitted in the hospital with a mild relapse of the disease. Their pregnancies continued uneventfully till delivery. Two gave birth to full term healthy males in the hospital. One male child survived and is living; 2nd male child had to be put on corticosteroid but died on the 4th day. Third woman gave birth to a healthy female child at home and the child is alive and well. Fourth woman developed Pemphigus vulgaris after

becoming pregnant. She had a fulminating course of the disease. She aborted a 4 month old foetus and died of hyperpyrexia and shock a few days later. In none of our cases, there was any transformation of one type of Pemphigus to another. This observation of the pregnant woman with Pemphigus vulgaris leading to full term pregnancy has been hitherto unreported, and is difficult to explain on the physiological grounds knowing the atrophy of the foetal adrenal glands in mothers receiving prednisolone.

Histopathology in Pemphigus Cases

Histopathological examination could be undertaken in 32 cases of Pemphigus. The observations are tabulated in Table No. 6. In 31 cases, the histopathological examination was consistent with the diagnosis of Pemphigus vulgaris and in 1 case with that of Pemphigus foliaceus. Wherever necessary, the histopathological examination was supplemented with demonstration of acantholytic cells in the smears made from the lesions. In all cases of Pemphigus vulgaris the split was suprabasal and the cavity contained acantholytic cells. Histopathological study of skin lesions and demonstration of Tzanck cells should always be done in cases where the clinical diagnosis is doubtful.

TABLE 6
Showing the histopathological findings in cases of Pemphigus.

Type of Pemphigus	No. of cases	Histopathological Findings
Pemphigus vulgaris	31	Intraepidermal suprabasal cavitation, some acantholytic cells seen. Polymorphonuclear leucocytes & an occasional eosinophil were seen in some cases.
Pemphigus foliaceus	1	Intraepidermal subcorneal split was seen. Cavity contained few acantholytic cells.

Management of Cases of Pemphigus

The management consisted of (a) administration of systemic corticosteroids (b) treatment of secondary infection with suitable antibiotic (c) local treatment of the lesions and (d) general supportive measures.

Most of the cases were put on oral prednisolone. The dose varied from 30 to 40 mg. daily, maximum dose being 80 mg. in severe cases. Adjuvant therapy consisted of anabolic hormones like Durabolin and Electrolyte therapy with potassium chloride and antibiotic therapy in cases with secondary infection. Locally an antiseptic dye like 1% gentian violet or brilliant green was given. When all the lesions cleared up and no fresh lesions appeared for two weeks, the dose of prednisolone was lowered 5 to 10 mg. per week till the patient could carry on with a maintenance dose of 12-16 mg. daily. Patients were instructed to take prednisolone without stopping it. We had to use A. C. T. H. in some brittle cases of Pemphigus vulgaris while stabilising them on maintenance dose of prednisolone. The skin lesions healed without scarring. Residual lesions like pigmented macules took several weeks to clear up. Mucosal lesions cleared later than the skin disease. We have no experience in sudden withdrawal of prednisolone. It has been noticed that cases discharged on a higher maintenance

dose of prednisolone relapsed earlier and more frequently. To our mind the factors responsible for relapse are (1) reversion to increased physical activity, the body demanding a higher dose of prednisolone (2) non-availability of the drug and (3) sudden stoppage of the drug due to intercurrent disease (4) massive secondary infection of the lesions demanding a higher dose of the corticosteroids for stabilising the lesions. None of our cases was given any blood transfusion. Features of iatrogenic Cushing syndrome were seen invariably in all cases of the present study. Steroid diabetes was observed in six cases only. It tended to improve on lowering the dose of prednisolone and cases were managed on the lines of diabetes mellitus. Other complications observed during the management were pulmonary tuberculosis 1 case; pleural effusion 2 cases, pulmonary embolism with pneumothorax-1 case. Out of 85 cases 8 cases (4 males and 4 females) are dead. These cases died of pulmonary embolism, hyperpyrexia, and circulatory failure. The survival period of the dead cases after the development of the disease varied from 4 months to 5 years. 10 cases are known to be alive and well. The longest surviving case is alive and well for the last 10 years with a maintenance dose of 5 mg. of prednisolone. The fate of remaining 67 cases is unknown as they would not communicate with us.

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TRUE or FALSE ?

Kidney transplantation in cases of Fabry's disease has proved to be palliative in some and detrimental in others suffering from this condition.

(Answer page No. 115)