PEMPHIGUS

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Pemphigus is one of the major bullous diseases of the skin characterised by the presence of multiple bullae on skin and mucous membrane.

The earliest description of 'Pemphigus' as a separate clinical entity was made by Robert Willian (1), who in his classic "On Cutaneous Diseases" (1808) described it under the title "Pompholyx Diutinus". He described the condition as "an eruption of bullae, without any inflammation round them, and without fever", and added that it "is a tedius and painful disorder". He pointed out that both skin and mucous membranes may be involved and commented on the uncertainty of the etiology.

The following is a brief review of the cases of Pemphigus treated as inpatients at Skin and V. D. Department of the Medical College Hospital, Trivandrum during the three years from 1962 to 1964. There was a total of 21 cases, all belonging to the group of Pemphigus Vulgaris. This year we had under our care one case of Pemphigus foliaceous. This case presented first as Pemphigus Vulgaris and gradually changed to florid generalised pemphigus foliaceous. The diagnosis was made on history and clinical features, and in all cases was confirmed by histopathological studies.

Clinical features : The presenting features were bullae or denuded areas from ruptured bullae. The lesions were generalised without any specific localisation, or regional distribution. The bullae tended to occur more frequently at areas of pressure or trauma. The lesions were asymmetrical in distribution.

Only ten cases (less than 50%) showed mucosal lesions (Table I). These were mainly in the oral, ocular and genital regions.

The primary lesions were monomorphous, unilocular bullae of varying size and shape. Most of the bullae appeared as such on normal skin without any signs of inflammation around. The earlier lesions were tense and contained clear fluid. The older ones were more flaccid often with purulent contents. Occasional bullae were frankly haemorrhagic. The bullae showed characteristic peripheral extension on pressure. Nikolsky's sign i. e. detachment of epidermis on lateral tension–was positive in all cases. After rupture, the bullae left raw oozing surfaces with tendency to spread and no tendency to spontaneous healing. The lesions on healing left variable scarring and pigmentary changes, mostly hyperpigmentation.

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TABLE I — SUMMARY OF CASES

Sl. No.	Name	Age	Sex	Blood group	Mucosal involvement	Duration of stay in Hospital in weeks	Date of admission	Remarks
1.	L	38	M	A	–	2	7—1–62	
2.	A	24	F	B	–	2	17—1–62	
3.	B	38	F	O	–	2	19—5–62	
4.	M	39	F	B	–	3	2—9–62	
5.	C	28	M	B	–	1	26—9–62*	
6.	T	46	M	A	–	10	12–10–62**	
7.	B	37	F	A	–	3	12–11–62	
8.	V	53	M	O	–	7	10—3–63	Discharged at request
9.	K	22	M	A	–	3	20—9–63	
10.	S	14	M	O	–	2	4—3–63	
11.	D	50	F	A	–	4	31—1–63	
12.	G	58	M	B	–	7	7–11–64	discharged at request
13.	W	40	M	C	–	2	11–12–63*	
14.	G	40	M	B	–	1	9—1–64	
15.	S	35	F	B	–	2	4—2–64	
16.	B	6	M	B	–	2	28—2–64	
17.	S	33	M	B	–	3	5—5–64	
18.	B	55	M	O	–	2	20—5–64	
19.	S	18	M	B	–	3	5—6–64	
20.	A	36	M	B	–	1	6–10–64	
21.	O	5	F	B	–	4	19—9–64	

* Admitted with severe toxaemia from " Native " treatment.

** Died during 2nd hospital stay on 6–2–63.

Onset in most cases was gradual and insideous, the bullae tending to appear in crops. Rarely bullae appeared singly.

In the majority of cases symptoms were remarkably mild. Some complained of pain, mainly from extensive raw areas and secondary infection. Itching was not a major symptom in any. The general health was unimpaired initially, but became rapidly worse in cases with extensive exudative surface and secondary infection. Cases with mucosal lesions in oropharynx often developed moderate to severe gingivitis and stomatitis. Restricted intake of food and fluids in these cases resulted in rapid deterioration of general health.

Histopathology. Histopathological diagnosis was made by biopsy and exfoliative cytology (Tzanck Test). Intra epidermal location of bullae, acantholysis and relatively little inflammatory reaction in the dermis were the main features noted. Some of the

bullae showed a large number of eosinophils. Very early lesions showed a rather characteristic acute angle at the junction of the roof and floor of the bullae, with acantholytic cells floating out into the cavity.

The Tzanck Test was considered positive when scrapings from the floor of the bullae showed characteristic acantholytic cells with relatively large hyperchromatic nucleus and basophilic cytoplasm showing peripheral condensation. This was found to be a useful supplementary test.

Age. The youngest case in this series was 5 years and oldest 58 years (Table I). The highest incidence of the disease was in the 4th decade (Fig. I). This is in variance with the usual reported incidence. According to Pillsbury etal (2) Pemphigus "is not seen in childhood and is rare during adolescence". We had two cases below the age of 10, and one in the 2nd decade. This year we had another case of pemphigus vulgaris in a boy aged 14 years.

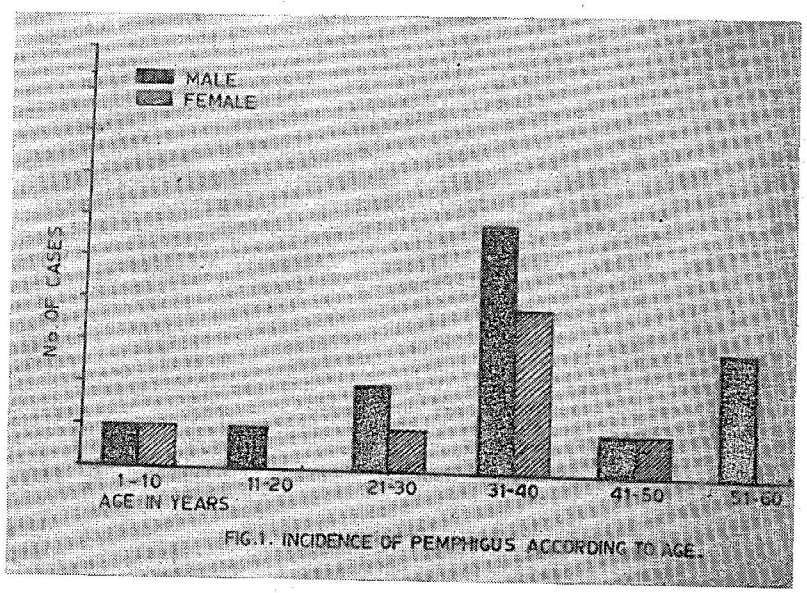

FIG.1. INCIDENCE OF PEMPHIGUS ACCORDING TO AGE.

Sex The M:F. Ratio in this series is 2:1. The reported incidence varies a lot. While Pillsbury etal (3) say that "there is no sex predisposition to pemphigus", Borrie quotes Rook that "Males are affected three times as often as females". Incidence in this series falls half way between these two.

Blood groups. An interesting feature in this series is the unusually high incidence of the disease in persons of blood Group B. (Table III). The percentage incidence of blood groups in a consecutive series of over 10,500 donors to Blood Bank, Medical College, Trivandrum, is given for comparison.

TABLE II

Blood Group	A.	B.	O.	AB.
Pemphigus cases	6	11	4	Nil
% incidence in pemphigus cases	29	52	19	Nil
% incidence of Blood groups in Blood donors.	25	25	46	4

Management. The general principles of treatment consisted of:

(i) Corticosteroid therapy
(ii) Blood Transfusions
(iii) Control of Sepsis
(iv) Local treatment and
(v) General supportive treatment.

Corticosteriods. It is true that Corticosteriods have markedly improved the prognosis in pemphigus. The prohibitive cost of the drug is still a problem in the management of the average case. We have found that starting treatment with large doses would bring the disease under control in a short time. We have been using as a routine prednisolone or triamcinalone, with the addition of ACTH on occasions. The usual dose of prednisolone required to produce a remission was 30 to 40 mg. per day given in divided doses. Once a remission was achieved the dose was rapidly 'tailed–off' and either stopped completely or continued on a small maintenance dose. During 'tailing off' ACTH was given with the hope of stimulating endogenous production of steroids.

Corticosteroid therapy was embarked upon after taking the usual precautions and a close watch was kept on the general health of the patient with special reference to the well–known complications of steriod therapy. All patients were given potassium supplements in the form of Mist. Pot. Cit. routinely. None of the patients developed severe infections, gastro–intestinal complications, diabetes mellitus or hypertension. Mooning of the face and fluid retention, manifested as weight gain or clinical oedema were noted in a few. These improved on reduction of dose. A few required oral diuretics to relieve the oedema. Mental disturbances were noted in a few, but it is difficult to say whether this was due to corticosteroids or a reaction of the distressing disease.

Blood transfusion. We have found blood transfusions to be of singular value in the management of pemphigus. During the active stage, patients were given weekly or twice weekly tranfusions of blood one bottle (250 ml) at a time. Once the activity of the disease was controlled the patients were discharged home and advised to attend as out–patients for further tranfusions. These were given at monthly intervals. This interval was shortened if patients showed any signs of relapse earlier.

Control of sepsis. Systemic antibiotics were needed to control the secondary infection in cases with widespread raw areas. We have found that therapy solely with Benzylpenicillin injection, B. P., 10 lakhs units 8th hourly, to be most effective in this respect.

Local treatment. This was given according to the local condition of the skin. It varied from cold saline compresses and shake lotions to creams and ointments. Antibiotic creams and ointments with or without corticosteroids were found valuable on denuded areas with secondary infection.

Gnenral treatment. It is of the utmost importance to maintain the morale and general health of the patient with appropriate supportive therapy. All patients were given a nourishing diet with a high protein content. Patients with lesions in oropharyngeal mucosae often needed parenteral therapy with fluids and electrolytes to maintain their nutritional status.

Other measures. Sulphapyridine, dapsone, chloroquin and Suramin were tried in a few cases without any benefit.

On the above lines of treatment, the overall result is encouraging. Case No. 4 and Case No. 12 are considered treatment failures as they were discharged from hospital at their request. We had two deaths (Case No. 6 and 7) on first admission. These two cases were admitted with severe toxaemia from 'native' therapy with drugs containing arsenic and mercury. They also had widespread secondary infection. Case 13, died during the second hospitalisation.

The cases that had responded to treatment needed hospitalisation only for 2-3 weeks (Ref. Table No. I). On discharge from hospital they were advised to attend regularly as out-patients for follow-up and blood transfusions. Only a minority of these cases are on maintenance dose of corticosteroids. Six cases (No. 1, 2, 3, 5, 8 and 10) in this group have been followed-up for more than 2 years without severe relapse.

SUMMARY

Clinical and histopathological features of 21 cases of pemphigus treated as inpatients are reviewed.

An unusually high incidence of pemphigus in the younger age group is noted.

A relatively high preponderance of the illness in persons of Blood Group B in this series is pointed-out.

The line of treatment followed is briefly out-lined. The regime has been found to be relatively less expensive and encouragingly efficatious.

The value of blood transfusions in the management is stressed.

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