ORIGINAL ARTICLE

A CLINICO-PATHOLOGICAL STUDY OF 22 CASES OF PEMPHIGUS

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ABSTRACT

Introduction: Blistering diseases are alarming skin conditions where blister formation occurs in various ways and cannot be differentiated clinically. For confirmation of diagnosis routine histological examination of early lesions is very much helpful. Pemphigus is one of autoimmune blistering disorder involving skin and mucosal surfaces. Tzanck smear may be used as a rapid diagnostic tool.

Material and Method: A clinico-palhological study of 22 cases of pemphigus over a span of two year at the Department of Pathology, Medical College Baroda and S.S.G. Hospital has been carried out to know incidence of pemphigus.

Result: Pemphigus vulgaris constituted the single largest group of 18 cases followed by pemphigus foliaceus (2 cases) and pemphigus erythmatosus (2 cases). Majority of the cases were seen in the age group of 21-60 years, with a slight female predominance. The youngest patient was 20 years while the eldest was aged 73 years. Flaccid bullae were present in 100% cases. Tzank smear found to successful in 75% cases of pemphigus vulgaris. However, it is difficult to take Tzank smear in some patients as patients were presented with predominantly crusted lesions.

Conclusion: Most of the patients have pemphigus vulgaris. Tzank smear is proved very useful tool for rapid diagnosis of pemphigus.

Key Words: Pemphigus, Pemphigus Vulgaris, Skin conditions, blisters

INTRODUCTION

Pemphigus is one of the severe and life threatening autoimmune blistering diseases[1]. The term pemphigus stems from the Greek word pemphix (blister) and describes a groups of chronic blistering skin diseases in which antibodies are directed against the cell surface of keratinocytes. This group of diseases carries bad prognosis. In 1880, Auspitz recorded the histological findings of pemphigus blister and coined the term acantholysis [2] Pemphigus can be divided into five types (a) Pemphigus Vulgaris, (b) Pemphigus Foliaceus, (c) Drug-induced Pemphigus, (d) IgA Pemphigus, (e) Paraneoplastic Pemphigus. Histopathological examination of the lesions shows intraepidermal blister formation, acantholysis and mild superficial dermal inflammatory infiltrate.[3]

The present study of 22 cases intends to highlight various clinico-pathological & cytological aspects of the disease in the patients affected with pemphigus vulgaris.

MATERIAL AND METHOD

This study comprises of 22 cases of pemphigus seen at S.S.G. Hospital & Medical College, Baroda observed over period of two year. A detailed history with particular reference to the mode of onset, characteristics and distribution of the lesions was taken. Punch biopsy of early vesiculo-bullous lesions were carried out in all patients with biopsy punch of 4mm or 5mm diameter.

The selection of the site for the biopsy is very critical step and lesion should be selected very carefully. Lesion should be fully developed and uncomplicated, otherwise histopathological examination may be misleading. Biopsy specimen should include lesion as well as normal surrounding tissue, so that advancing border of the lesion can bevisualized for histopathological examination of vesicles and bullae. It is important that the entire lesion be removed intact to permit the study of location of vesicle, the nature of its roof and floor and type, condition and cells present in the blister. The entire specimen should be fixed in formalin and embedded in paraffin.[4]

Cytological smear (Tzank smear) were carried out where it is possible because most of the patient come with older lesions with crusting so it was not possible to take smear in all patients. To make Tzank smear, intact roof of a blister is opened along one side, folded back and the floor is gently scraped. The material thus obtained is smeared on microscopic slide, air dried, and stained with H & E[5].

RESULTS

Out of the 22 cases of pemphigus studied, pemphigus vulgaris was the predominant type with 18 cases (81.81%) followed by 2 cases (9.09%) of pemphigus foliaceus and 2 cases of (9.09%) of pemphigus erythmatosus in present study.

Table 1: Gender wise distribution of pemphigus

Type of Pemphigus	Male (%)	Female (%)	Total (%)
Pemphigus vulgaris	07(31.81)	11(50)	18(81.82)
Pemphigus erythmatosus	01(4.54)	01(4.54)	02(9.09)
Pemphigus foliaceus	00	02(9.09)	02(9.09)
Total	08(36.35)	14(63.65)	22(100)

Of the 18 cases of pemphigus vulgaris, 8 patients (44.46%) were between 31 and 40 followed by 4 cases from age group of 21 to 30 & 4 cases from age group of 41 to 50 years. Only 1 case (5.55%) was above 70 years. Out of the 2 cases of pemphigus foliaceus, 1 patient (50%) was in the age group of 31-40 years & 1 patient (50%) was in the age group of 31-40 year. Two cases of pemphigus erythmatosus were seen. Out of the 2 cases of pemphigus foliaceus, 1 patient (50%) was in the age group of 21-30 years; & 1 patient (50%) was in the age group of 31-40 year. There was a slight female preponderance in the ratio of 1.6:1 (11females and 7 males). Most commonly affected age group was 31–50 years of age in case of pemphigus vulgaris.

Pemphigus vulgaris showed initial lesions involving mucous membranes in 33.33% cases and lesions including both skin and mucosa in 66.87% cases. Only mucosal involvement not found in any cases. On the other hand, in pemphigus foliaceus, both cases showed involvement of both skin and mucous membrane involvement.

Clinically flaccid bullae were found in all cases. Blisters seen arising on nonerythematous skin was seen in 16 cases (88.88%) of pemphigus vulgaris, which spontaneously ruptured to give rise to erosions in 13 cases (72.22%). Crusted lesions, erythematous plaques, vegetations and pustules were present less frequently. In pemphigus foliaceus, blisters arising on erythematous skin were seen in both cases (100%).

Table 2: Histological findings in pemphigus vulgaris

Histological findings	Cases (%)
Spongiosis with supra basal bulla	02 (11.11)
Suprabasal bulla with acantholytic cells in blister cavity	04 (22.22)
Suprabasal bulla	12 (66.67)
Total	18 (100)

Of the 2 cases of pemphigus foliaceus studied, both cases (100%) showed acantholysis. Subcorneal bulla was seen in bothcases (100%). Dyskeratosis, basal layer budding and pseudoepitheliomatous proliferation were not seen in any of the cases. An inflammatory infiltrate was present in the bulla cavity in 1 case (50%).

Of the 2 cases of pemphigus erythmatosus studied, both cases (100%) showed acantholysis. Subcorneal bulla was seen in both cases (100%). Dyskeratosis was seen in only1 cases (50%). Inflammatory cells were seen in the bulla cavity in both cases (100%). Spongiosis was seen in 1 case (50%).

Of 18 patients having pemphigus vulgaris we carried out Tzank smear in 12 patients who presented with vesciculobullous lesions. Out of 12 cases, 9 (75%) cases showed characteristic acantholytic cells of pemphigus vulgaris in Tzank smear and 3 (25%) cases failed to show acantholytic cells.

DISCUSSION

Our series is in accordance with other Indian series each of which has maximum cases of pemphigus vulgaris, followed by pemphigus erythematosus and foliaceus [2,6,7,8]. Majority (85.7%) of our patients were between 21-60 years, akin to Indian literature[7, 8]. Pemphigus vulgaris affects males and females equally, although in the present study, there was a slight female predominance in the ratio 1.6.:1.the female to male ratio was 1.04:1 and 1.4:1 in studies by Ameneh Yazdanfar and Deval Vora et al respectively[3, 8].

Nature and distribution of lesions as well as mucosal involvement in different types of pemphigus in our series has followed the pattern seen in earlier studies [3, 8, 9]. Flaccid bullae were seen in all the cases.

The salient histological features of pemphigus vulgaris were (1) intraepidermal suprabasal blisters, (2) presence of acantholytic cells, (3) "Row of tombstone appearance" (4) Acantholysis. Salient features of pemphigus foliaceus were (1) acantholysis (2) subcomeal cleavage (3) an inflammatory infiltrate.

Tzank smear is also useful to diagnose pemphigus vulgaris. Of 12 cases of pemphigus vulgaris 75% cases showed acantholytic cells and provided successfully a quick, easy, convenient tool for aiding the clinical diagnosis & it is accordance to earlier study of Medhi G. et al [10].

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