International Journal of Research in Dermatology
Parmar D et al. Int J Res Dermatol. 2017 Dec;3(4):478-480
http://www.ijord.com

DOI: http://dx.doi.org/10.18203/issn.2455-4529.IntJResDermatol20175368

Original Research Article

Prevalence of pemphigus incidence in the Bhuj, Kutch, Gujarat: a cross-sectional study

Devendra Parmar[1]*, Keyur J. Patel[2]

[1]Department of Dermatology, Gujarat Adani Institute of Medical Science, Bhuj, Gujarat, India
[2]Government Medical College and New Civil Hospital, Surat, Gujarat, India

Received: 28 July 2017
Accepted: 31 August 2017

***Correspondence:**
Dr. Devendra Parmar,
E-mail: researchguide86@gmail.com

ABSTRACT

Background: Pemphigus is a group of rare skin disorders that cause blisters and sores on the skin or mucous membranes, such as in the mouth or on the genitals. In Brazil, the prevalence of the disorder is 3.4% in regions such as the Amerindian reservation of Limão Verde and approximately 15,000 patients are known to have pemphigus foliaceus. This prompted us to conduct a study to estimate the incidence of pemphigus in the central Bhuj district of the state of Gujarat in India.
Methods: The present study was planned and conducted between January 2016 to December 2016. The entire team of dermatologists licensed in the district were included in the study and full efforts were made to reach out to them. All the participating dermatologists were asked to fill up a simple perform prepared by the experts in the field. The Performa included the details like date, investigations required for pemphigus, diagnosis based for pemphigus.
Results: The total population of Bhuj district is estimated to be 5 millions. At the end of the study period that is one year, all the collected performa from the licensed dermatologist were evaluated and results were prepared accordingly. A total of 24 patients who fulfilled all the inclusion criteria were included in the study.
Conclusions: This original research proves that the incidence of pemphigus in Bhuj district was 7.2 per million in the period of one year. This incidence is much higher than the incidence rates reported from the Germany where only 14 cases were reported over a period of 8 years. This study does give us a preliminary idea about the incidence of this rare disease in this geographic area.

Keywords: Pemphigus, Vulgaris, Epidemiology, Bhuj district

INTRODUCTION

Pemphigus is a group of rare skin disorders that cause blisters and sores on the skin or mucous membranes, such as in the mouth or on the genitals. Pemphigus is not to be confused with bullous pemphigoid, another blistering skin condition.[1,2] Pemphigus is a group of organ specific autoimmune diseases characterized by the production of auto antibodies to desmogleins (a). They are mucocutaneous blistering diseases that demonstrate a loss of cohesion of the epidermal cells (acantholysis) which result in the formation of clefts in the epidermis.[3-5] Autoantibodies directed against adhesion molecules cause epidermal keratinocytes to separate, resulting in intraepidermal bullae. Pemphigus is an uncommon disease with an incidence rate ranging from 0.5 to 3.2 per 100,000 per year.[6,7]

Of the four main types of pemphigus, pemphigus vulgaris is the most common; pemphigus foliaceus, pemphigus vegetans, and pemphigus erythematosus are less common. There may indeed be only two main forms of

pemphigus because pemphigus vegetans may be a variant of pemphigus vulgaris and pemphigus erythematosus may be a localized variant of pemphigus foliaceus.[8,9]

There is role of other exogenous factors that can provoke the disease in genetically predisposed individuals. Patients with pemphigus have been found to expose to pesticides three times more often than healthy subjects.[10] A study showed that pemphigus patients had more number of pregnancies than controls. A dose dependant relation has been found in cases using cosmetics. In Brazil, the prevalence of the disorder is 3.4% in regions such as the Amerindian reservation of Limão Verde and approximately 15,000 patients are known to have pemphigus foliaceus.[11] All these studies suggest that there is a probable environmental factor triggering the disease. This prompted us to conduct a study to estimate the incidence of pemphigus in the central Bhuj district of the state of Gujarat in India.

METHODS

The present study was planned and conducted between January 2016 to December 2016. The entire team of dermatologists licensed in the district were included in the study and full efforts were made to reach out to them. All the participating dermatologists were asked to fill up an simple perform prepared by the experts in the field. The Performa included the details like date, investigations required for pemphigus, diagnosis based for pemphigus. Whenever the new patient from the Bhuj district were diagnosed, various details were filled by the licensed dermatologist and were send it to the principal investigator. Patient who were from outside the Bhuj district they were excluded from the study. However the people who were temporary residing into the Bhuj district and who happened to diagnosed with Pemphigus was included in the study.

RESULTS

The total population of Bhuj district is estimated to be 5 millions. At the end of the study period that is one year, all the collected performa from the licensed dermatologist were evaluated and results were prepared accordingly. A total of 24 patients who fulfilled all the inclusion criteria were included in the study. The average age was found to be 52 years and 43 years in females. Six patients were diagnosed with pemphigus foliaceus; out of them 4 were male and 2 were female. Twelve cases were found to be diagnosed with pemphigus vulgaris, out of the 12; there were 8 males and 4 to be females. Rest six cases were diagnosed with pemphigus erythematosus; out of which 5 were males and 1 female. No cases of pemphigus vegetans were not recorded. All the pemphigus vulgaris were diagnosed clinically, histopathological confirmation and direct immune immunoflurescence. For pemphigus foliaceus, tzanck smear test and clinical diagnosis were used. For confirmation direct immunoflurescence method was used. For confirmatory diagnosis of pemphigus erythematosus, direct immunoflurescence method was used. This study shows that the incidence of pemphigus in Bhuj district, Gujarat is 7.2 per million and there is male predominance [65.5%] in evaluating the incidence of pemphigus. The distribution of lesions was on chest, scalp, face, axillae, neck, extremities, and abdomen. There was ill defined and light to dark brown pigmentation in 20 and hypo pigmentation in 4 cases without scarring or atrophy. They were observed in buccal mucosa, palate, gingiva, tongue and genitalia. Palm, soles, nose, eyes and ear canal were not involved.

Table 1: Sex distribution of our original research.

No.	Diagnosis	Males	Females
1.	Pemphigus vulgaris	8	4
2.	Pemphigus erythematosus	5	1
3.	Pemphigus foliaceus	4	2
4.	Pemphigus vegetans	0	0
5.	Total	17	7

Table 2: Method of diagnosis for each type of pemphigus.

No.	Diagnosis	Method for diagnosis	No. of cases
1.	Pemphigus vulgaris	Biopsy, DIF	12
2.	Pemphigus erythematosus	DIF	6
3.	Pemphigus foliaceus	Tzanck Smear, DIF	6
4.	Pemphigus vegetans	None	0
5.	Total		24

Table 3: Site of pemphigus lesion.

No.	Site of lesion	Number of lesions
1.	Chest	3
2.	Face	4
3.	Axillae and neck	2
4.	Scalp	5
5.	Abdomen and extremities	2
6.	Buccal	4
7.	Gingiva and palate	2
8.	Tongue	2

DISCUSSION

Pemphigus is a group of organ specific autoimmune diseases characterized by the production of autoantibodies to desmogleins (a). They are mucocutaneous blistering diseases that demonstrate a loss of cohesion of the epidermal cells (acantholysis) which result in the formation of clefts in the epidermis.[12,13] Autoantibodies directed against adhesion molecules cause epidermal keratinocytes to separate, resulting in intraepidermal bullae. The basic abnormality in all forms of pemphigus is the separation of keratinocytes (acantholysis). This process leads to the formation of a cleft within the epidermis, which then enlarges into a bulla, which then breakdown into coalescing ulcers.[14,15]

All forms of pemphigus are associated with the presence of circulating and fixed autoantibodies—referred to as intercellular antibodies—against keratinocyte cell-surface antigens. It is most common in the fourth and fifth decades and equally affects males and females. PF occurs in middle-aged persons and presents with flaccid bullae over an erythematous base or as scaly crusted erosions without any blister formation. Here, the cleft is in the superficial epidermis, especially in the granular layer.[16]

Pemphigus vulgaris (PV) is the most common form of pemphigus (up to 80% of all pemphigus (c)). Pemphigus foliaceus (PF) is the second most common form of pemphigus (b), (5:1 in the US, but more common in Finland, South Africa, Mali (e)), and is endemic in some areas of Brazil and Columbia, where an insect vector is suspected but none yet identified. The prognosis of untreated pemphigus foliaceus is more favourable than that of pemphigus vulgaris, as the lesions of pemphigus foliaceus are not as deep, and there is less chance for infection, fluid loss, and metabolic disturbance.[17]

This original research proves that the incidence of pemphigus in Bhuj district was 7.2 per million in the period of one year. This incidence is much higher than the incidence rates reported from the Germany where only 14 cases were reported over an period of 8 years. The average age of the patients was found to be 40 years which is much lower than 60 years, found in European study. However contraindicating to the previous study male predominance is found more in our study as compared to females. Most of the incidences were reported more than the age of 50 years.

In France and Tunisia when study was done, it showed the incidence rates were found to be 1.6 and 6.7 million/year, respectively. The majority of the patients were of pemphigus foliaceus, the age group and percentage of PF were contraindicated as compared to our study. There are studies on the impact of environmental factors on the pathogenesis of pemphigus and their relationship to the incidence of pemphigus in populations. However, we do not have any evidence to propose such hypotheses now due to the preliminary nature of this study.

This study does give us a preliminary idea about the incidence of this rare disease in this geographic area. It is possible that a few patients who migrated out after developing the disease have been missed in this study but that is unlikely to change the incidence of this rare disease. To the best of our knowledge, this is the first study to cover the entire population of a Gujarat district in India or anywhere in the world to assess the incidence of pemphigus in a population.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the institutional ethics committee

REFERENCES

1. Scott JE, Ahmed AR. The blistering diseases. Medical Clin North America. 1998;82:1239-83.
2. Scully C, Muzio LL. Oral mucosal diseases: mucous membrane pemphigoid. Br J Oral Maxillofacial Surg. 2008;46:358-66.
3. Sitaru C, Zillikens D. Mechanisms of blister induction by autoantibodies. Experimental Dermatol. 2005;14:861-75.
4. Grando SA. Pemphigus autoimmunity: hypotheses and realities. Autoimmunity. 2012;45:7-35.
5. Kitajima Y, Aoyama Y. A perspective of pemphigus from bedside and laboratory-bench. Clinical Rev Aller Immunol. 2007;33:57-66.
6. Vakirlis E, Theodosiou G, Apalla Z, Arabatzis M, Lazaridou E, Sotiriou E, et al. A retrospective epidemiological study of skin diseases among pediatric population attending a tertiary dermatology referral center in Northern Greece. Clin Cosmet Investig Dermatol. 2017;10:99–104.
7. Tucker DK. Pemphigus Vulgaris: Pathomechanisms of a Desmosomal Disease and Protection by Plakophilin-1. Emory University, 2013.
8. Lever WF. Pemphigus. Medicine. 1953;32:1-123.
9. Korman N. Pemphigus. J Am Acad Dermatol. 1988;18:1219-38.
10. Ruocco V, Ruocco E, Schiavo AL, Brunetti G, Guerrera LP, Wolf R. Pemphigus: etiology, pathogenesis, and inducing or triggering factors: facts and controversies. Clin Dermatol. 2013;31:374-81.
11. Edelson RL. Pemphigus—decoding the cellular language of cutaneous autoimmunity. Mass Medical Soc. 2000.
12. Sagi L, Baum S, Agmon-Levin N, Sherer Y, Katz BSP, Barzilai O, et al. Autoimmune bullous diseases: the spectrum of infectious agent antibodies and review of the literature. Autoimmunity Rev. 2011;10:527-35.
13. Robinson ND, Hashimoto T, Amagai M, Chan LS. The new pemphigus variants. J Am Acad Dermatol. 1999;40:649-71.
14. Bystryn JC, Rudolph JL. Pemphigus. The Lancet 2005;366:61-73.
15. Velez AMA, Calle J, Howard MS. Autoimmune epidermal blistering diseases. Analysis. 2013;3:21.
16. Chowdhury J, Datta PK, Chowdhury SN, Das NK. A clinicopathological study of pemphigus in Eastern India with special reference to direct immunofluorescence. Indian J Dermatol. 2016;61:288.
17. Alcaide-Martin A, Gallardo-Perez M, Castillo-Munoz R, Fernández MM, Herrera-Ceballos E. Epidemiologic study of 20 cases of pemphigus at Hospital Clinico Universitario Virgen de la Victoria de Malaga, Spain. Actas Dermo-Sifiliográficas (English Ed). 2010;101:524-33.

Cite this article as: Parmar D, Patel KJ. Prevalence of pemphigus incidence in the Bhuj, Kutch, Gujarat: a cross-sectional study. Int J Res Dermatol 2017;3:478-80.