

Incidence of pemphigus in Thrissur district, south India

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ABSTRACT

Background: Pemphigus is a common autoimmune blistering disorder in India. However, there are no population based surveys from India available to study the prevalence of this disease. **Aims:** To estimate the incidence of pemphigus in Thrissur district, Kerala, South India. **Methods:** Questionnaire based survey was conducted among the practicing dermatologists of Trichur district between 1st January 2001 and 1st January 2002 to determine the incidence of pemphigus in the district. **Results:** Ten cases of pemphigus vulgaris, one case of pemphigus erythematosus, and two cases of pemphigus foliaceous were diagnosed during the study period in a population of 2.9 million in Thrissur district. The average age of pemphigus vulgaris patients was 58 years in males [42 to 82 years] and 37 in females [25 to 57 years]. The average age of the two male pemphigus foliaceous patients was 46 years [52 and 40 years] and the only pemphigus erythematosus patient was a 45 year-old female. Five cases of pemphigus vulgaris were investigated clinically histopathologically and by using direct immunofluorescence, while two were investigated clinically and histopathologically, two cases clinically, and one case clinically and with Tzanck smear. Two cases of pemphigus foliaceous were diagnosed with the aid of direct immunofluorescence and the only case of pemphigus erythematosus was diagnosed with the aid of direct immunofluorescence. Pemphigus vulgaris was common among females. The incidence of pemphigus in Thrissur district is 4.4 per million. **Conclusions:** This study indicates that the incidence of pemphigus in Thrissur district is high in comparison to the available studies from Germany, France, and the North African country Tunisia.

Key Words: India, Kerala, Pemphigus

INTRODUCTION

The Greek word “pemphigus” means a bubble or a blister. Pemphigus consists of a group of epidermal diseases associated with bullae and acantholysis. This group of diseases carries a relatively bad prognosis. Experimental studies in mice have proven that antibodies are directly involved as causes of this disease.^[1] Different types of this disease are pemphigus vulgaris (PV), pemphigus foliaceous (PF), pemphigus erythematosus (PE), and pemphigus vegetans (P veg). The main difference between PV and PF is the level of epidermal acantholysis. An increased incidence of Fogo selvagem, an endemic form of PF, has been reported in South America and Tunisia.^[2] In areas where pemphigus foliaceous

and pemphigus vulgaris are not endemic, the cumulative incidence of the two diseases is less than two cases per 100,000 population, and pemphigus vulgaris is usually the more common of the two. In areas where the diseases are endemic, the ratio of the number of cases of pemphigus foliaceous to the number of cases of pemphigus vulgaris is nearly 20 to 1.^[3] 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 foliaceous.^[3] All these studies suggest that there is a probable environmental factor triggering the disease. There is a general impression among dermatologists that the incidence of pemphigus is comparatively high in the Indian subcontinent.^[4] But there has been no systematic study so

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far to prove or disprove this. This prompted the author to conduct a study to estimate the incidence of pemphigus in the central Thrissur district of the state of Kerala in India.

METHODS

The Thrissur Dermatology Club, a scientific forum of all 30 dermatologists practicing in the District, sponsored the study. The study was conducted between 1st January 2001 and 1st January 2002. An effort was made to make sure that no dermatologist in the district was left out from participating in the study. All dermatologists practicing in the district were requested to fill up a Simple Performa (which includes the details, date, basis of diagnosis etc) and send it to the principal investigator of the study whenever he/she saw a new patient with pemphigus from Thrissur District. Patients from other districts, who came to Thrissur district for treatment or for other reasons, were excluded from the analysis. However, patients who were temporary residents (staying in the district for at least one year) of this district and who happened to develop the disease here were included in the study. Patients who had had lesions during the past years that were diagnosed to be pemphigus for the first time, were also excluded from the analysis.

The details of all patients included in this study are shown in Table 1.

RESULTS

In a population of 2.9 million in Thrissur district,^[5] ten cases of PV, one case of PE, and two cases of PF were diagnosed during the study period. The average age of PV patients was 58 years in males [42 to 82 years] and

37 years in females [25 to 57 years]. The average age of the two PF patients was 46 years [52 and 40 years]; both the cases were male and the only PE patient was a female aged 45 years. Five cases of PV were diagnosed clinically, histopathologically and by DIF, two cases clinically and histopathologically, two cases of PF were diagnosed by clinical methods and one case with the help of Tzanck smear. Two cases of PF were diagnosed by DIF and biopsy and the only case of PE was diagnosed by DIF. There was only one patient past the age of 80 years, who was diagnosed to have pemphigus vulgaris. PV was more common among females [seven out of ten cases]. This study shows that the incidence of pemphigus in Thrissur district is 4.4 per million and that there is a female predominance [61.5%] in the incidence of pemphigus.

DISCUSSION

This study proves that the incidence of pemphigus in Thrissur district was 4.4 per million in the period of 2001-2002. This incidence is much higher than the incidence rates reported from Germany where 14 cases were reported over an observation period of eight years [1989-1997] among the 1.46 million residents. In Thrissur district, 13 cases were reported over a period of one year [2001-2002] among the 2.9 million residents in the district. The average age of females with PV was 37 years in this study as compared to 50-60 years in the European study. However, female preponderance in the disease was noted in both the studies. The highest incidence of PV among males was in the age group > 55 years, which is almost on par with European studies.^[5,6]

Studies conducted over a period of six years in France and

Table 1: Details of our cases

| Age | Sex | Diagnosis | Date of diagnosis | Duration | Basis of diagnosis |
|-----|-----|-----------|-------------------|----------|--------------------|
| 45 | F | PE | 1-1-2001 | 5 months | DIF |
| 31 | F | PV | 2-1-2001 | 2 months | Biopsy |
| 82 | M | PV | 21-4-2001 | 3 months | Clinical |
| 32 | F | PV | 2-2-2001 | 6 months | DIF |
| 49 | F | PV | 21-7-2001 | 3 months | Clinical |
| 25 | F | PV | 12-7-2001 | 15 days | Tzanck |
| 50 | M | PV | 7-9-2001 | 1 month | Biopsy |
| 32 | F | PV | 10-10-2001 | 3 months | DIF |
| 52 | M | PF | 28-11-2001 | 6 months | DIF |
| 40 | F | PF | 29-9-2001 | 5 months | DIF |
| 57 | F | PV | 11-9-2001 | 9months | DIF |
| 42 | M | PV | 28-8-2001 | 6 months | DIF |
| 32 | F | PV | 28-5-2001 | 4 months | DIF |

DIF:Direct immunofluorescence; PV: Pemphigus vulgaris; PE: Pemphigus erythematosus; PF: Pemphigus foliaceus; Pveg: Pemphigus vegetans

Tunisia showed that the incidence rates in France and Tunisia were 1.6 and 6.7/million/year, respectively, whereas it was 4.4 million/year in our study. The majority of cases reported in Tunisia were of Article for PF and the age group and gender were strongly in contrast to the Indian study reported here. In our study, the reported cases of Article for PF were males of an average age of 46 years and the percentage of Article for PF of the total pemphigus cases was 15% as compared to 61% in Tunisia. The age group of pemphigus patients was between 25 and 32 years in Tunisia.^[7]

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.^[8]

In a rare disease like pemphigus, a prolonged study involving a larger population may give more precise epidemiological details. However, 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.

There are a few clinic-based studies on the prevalence of pemphigus from India.^[9] To the best of our knowledge, this is the first study to cover the entire population of a district in India or anywhere in the world to assess the incidence of pemphigus in a population.

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