Long-term prognosis of pemphigus in central Kerala, India: A retrospective cohort study

Sir,

There are only a few Indian studies documenting long-term outcome of pemphigus vulgaris. Most of them evaluate the outcome of dexamethasone-cyclophosphamide pulse therapy in pemphigus vulgaris.[1,2] These studies show varying estimates of achieving drug free remission. We tried to document the rate of long-term drug free remission and long-term outcome of pemphigus vulgaris patients from Thrissur district who are receiving currently available treatment from Government Medical College, Thrissur during the last 8 years.

We compiled details of such patients during the period 2002-2010. To make sure that only patients who had adequate exposure to the care from this hospital were included, we selected only those patients who completed at least 2 years of treatment. This also ensured that factors such as short term mortality with resulting loss to follow-up and outcome in patients without inadequate treatment did not affect the study. A trained health worker visited patients and handed over a letter detailing the study protocol from AK and requested them to participate in the study. Patients were also encouraged to contact principal investigator directly. Those who were not willing to participate were allowed to opt out. The author repeated interviews in 20% of the respondents. The study was cleared by institutional review board of the Government Medical College, Thrissur.

Data collected included demographic data and details of treatment, adherence and side-effects of treatment, co-infections, other morbidities, mortality, remission and relapse.

Out of 75 patients who were admitted with pemphigus vulgaris, 37 fulfilled inclusion criteria of being from Thrissur district (22 females and 15 males). We could collect details of 26 out of 37 of these patients. The remaining 11 patients were untraceable. At the time of the study, nine patients (five males and four females) were in complete drug free remission, three were still having skin lesions, four were taking medications even though they were symptom free and 10 had expired. All the nine patients in drug free remission had systemic corticosteroid as a main therapeutic agent. Four of them had treatment with dexamethasone-cyclophosphamide pulse therapy regimen and one was treated with steroid and azathioprine. Average duration of treatment in patients in the drug free remission was 3.33 years (1 year 8 months to 7 years).

Pemphigus vulgaris is considered chronic diseases with fatal outcome and rare drug free remission. A study from USA on 18 patients, found that drug free remission was achieved in three patients using steroids and mycophenolate mofetil within a 3-year period.[3] Another study showed remission in 35 patients (94.6%) with oropharyngeal lesions, of whom 13 (35.1%) were off therapy and 21 (56.8%) were on treatment at the last evaluation.[4] Another study showed drug free remission in 25%, 50% and 75% of patients after 2, 5 and 10 years of treatment, respectively.[4] A study from Italy on oropharyngeal pemphigus showed 35% achieving drug free remission.[5] Study from Korea showed complete or partial remission in 77% at 5 years and 94% at 10 years after initial diagnosis. There are no similar studies on pemphigus except that on patients on dexamethasone-cyclophosphamide pulse regimen from India. Our study shows 9 out of 26 (34.6%) patients achieving drug free remission with in a mean duration of 3. 33 years (1 year 8 months to 7 years). This study also showed that common regimen like daily systemic steroid could induce drug free remission in pemphigus vulgaris.

It is possible that patients who died in the initial days of admission and those who reached remission before 2 years would have affected the study results. We acknowledge this as a possible weakness of the study.

The high mortality of pemphigus vulgaris (10 out of 25) in this cohort is possibly because it included data from patients who were lost on follow-up, which probably may not have been possible in other hospital based studies.

In this study, we could document that a significant proportion of patients with pemphigus vulgaris does go into drug free remission following varying periods of immunosuppression. This includes patients on systemic steroid alone, steroid with immune suppression and dexamethasone-cyclophosphamide pulse therapy regimen. Further larger prospective cohort studies are necessary to define the group of patients who attains remission. A preliminary analysis of our data indicates that the numbers of admissions or the exact regimens used are unrelated to drug free remission. The weaknesses of this study are the small sample size; high level of missing data, retrospective design and exclusion of patients in the early years of illness. We suggest larger long-term prospective cohort studies to overcome these limitations. Yet, this is probably the first study from India, which shows that a significant proportion of pemphigus vulgaris patients can go into drug free remission even when treated using different regimens.

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