

PUBLISHED ABSTRACT

Granulomatous Disease and Lymphoma in a Cohort of 1395 Patients with CVID in the USIDNET Registry

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Background

Granulomatous disease (GD) has been described with a variable incidence (8.0–22.0%) in patients with common variable immunodeficiency (CVID). An increase in malignancies has been reported in CVID patient cohorts, particularly for lymphoma, reported in 1.6–8.2% of the CVID patients depending on the cohorts. Prior analysis of a cohort of 436 CVID patients, included 59 patients with GD (GD+). In these, there was a suggestion of more cases of lymphoma (12.5%) when compared to cases without (GD–) (5.0%) although the difference was not statistically significant ($p = .07$). Our objective is to compare the frequency of lymphoma in GD+ and GD– patients in the CVID patient cohort from the USIDNET Registry.

Methods

We submitted a query to the USIDNET registry requesting de-identified data for patients with the diagnosis of CVID, through August 2018. Statistical analysis was performed on SPSS, with comparisons done with Pearson chi-square or Fisher's exact test, depending on the sample sizes, using an alpha level of .05.

Results

A cohort of 1395 CVID patients from the USIDNET registry was analyzed. Ninety-one patients (6.5%) were GD+. Overall, 152 patients (10.9%) had a malignancy diagnosis, 47 of these (3.4%) with lymphoma. Lymphoma was present in 6/91 GD+ patients (6.6%) versus 41/1304 GD– patients (3.1%) ($p = .12$). Overall malignancy was present in 15/91 GD+ (16.5%) versus 137/1304 (10.5%) ($p = .08$).

Conclusions

In the cohort of 1395 CVID patients from the USIDNET registry, we found a frequency of lymphoma of 3.4%, which is in the range of previously described cohorts. The frequency of lymphoma was 6.6% in patients with GD, higher than the 3.1% frequency for GD– patients, but these differences were not statistically significant. Our identified frequency of lymphoma in GD+ patients was lower than the one previously identified in the 436 CVID patient cohort, but with similar proportional differences between GD+ and GD– patients. Despite no statistical significance, the frequency of lymphoma, as shown here and elsewhere, was higher in CVID patients with granulomatous disease in both studies, with no full understanding of this increased risk of lymphoma. Expanding this analysis to larger groups of CVID patients may help to confirm, or deny a more robust association, which may have a meaningful impact in the outcomes of this particular population.

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