HIGH PREVALENCE OF CANCER AMONG PRIMARY SJÖGREN'S SYNDROME: DATA FROM BRAZILIAN SJÖGREN'S SYNDROME REGISTRY (RE-BRASS)

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BACKGROUND

About 5–10% of patients with Sjögren's syndrome (SS) will develop non-Hodgkin's lymphoma, presenting a 16 to 44 increased risk compared to the general population. The relationship between SS and other types of cancers has not been much described in the literature.

METHODS

A total of 1,010 consecutive patients were included in the Brazilian Sjögren's syndrome registry (RE-BRASS), fulfilling the 2002/2016 classification criteria for primary SS. The patients who developed neoplasia diagnosed according to the National Cancer Institute classification were retrospectively analyzed.

RESULTS

A total of 1,010 patients were included, 975 women, average age 57.21 years, 110 out of 909 patients (10%) presented cancer that was more prevalent in 55-year-old patients ($p < 0.001$). The diagnosis of cancer occurred after the diagnosis of SS in 10 out of 15 (66.7%) of the cases of lymphoma, 37 out of 68 (54.4%) other cancers and in 12 out of 16 (75.0%) of skin cancers. Regarding the subtypes of cancer, 21 (2.5%) patients had skin cancer, 15 (1.5%) had lymphoma, and 68 (6.7%) had other neoplasms. Basal cell carcinoma 13 (59%) was the most prevalent skin cancer, followed by melanoma 5 (0.6%). Lymphoma was the most prevalent cancer after skin cancer in our population, and non-Hodgkin MALT lymphoma 11 (73%) was the most prevalent with respect to lymphoma subtypes. Regarding other cancers, thyroid cancer 9 (0.9%) was the most prevalent, followed by ovarian cancer, 4 (0.4%) cases. Regarding the prognosis 18 out of 21 (90%) with skin cancer, 13 out of 15 (86.7%) with lymphoma and 51 out of 68 (81%) with other cancers progressed to cure, 4 (6.3%) patients had metastases, 4 (6.3%) relapsed and 4 (6.3%) died. The presence of cancer was associated with age (OR = 1.03), disease duration (OR = 1.01) and the presence of anti-Ro/SSA antibodies $p < 0.008$.

CONCLUSION

Skin cancer is the most prevalent cancer as it is in Brazilian general population. Lymphoma was the most prevalent cancer after skin cancer in our population. Among other cancers, thyroid and ovarian cancer were the most prevalent, which differs from the Brazilian female population in which breast cancer is the most prevalent and thyroid cancer is the fifth in prevalence. Patients with SS have different kinds of cancer and its prevalence is superior and differs from the general population without SS.

KEYWORDS

Sjögren's syndrome, Cancer, Cohort.