MORTALITY IN BRAZILIAN CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS: PREDICTORS AND CAUSES OF DEATH

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BACKGROUND

With the introduction of new treatment strategies over the years, improvement on childhood-onset systemic lupus erythematosus (cSLE) survival is widely recognized. However, these estimates are based on isolated, short-period small studies, and information on recent trends are lacking. The objectives of the study were to identify risk factors of mortality in cSLE and to evaluate the association of these factors with clinical manifestations, laboratory features, disease activity and damage, current treatment, survival and causes of death.

METHODS

This was a multicenter, nationwide observational study of a cohort of 1,528 cSLE patients followed in 27 Brazilian pediatric rheumatology tertiary centers. Patients' charts were analyzed according to a standardized protocol at cSLE diagnosis, follow-up, last visit, or death.

RESULTS

A total of 63/1,528 (4.1%) of patients died, 53/63 of them were female (84.1%), with a median age of death of 11 years old and a median period of 3.2 years after cSLE diagnosis. Death certificates were available in 52/63 (82.5%) patients and sepsis was the most frequent cause of death in 27/63 (42.8%) patients, followed by opportunistic infections 6/63 (9.5%) and alveolar hemorrhage in 6/63 (9.5%) of patients. Autopsy was performed in 12/63 (19.0%) of the deceased cSLE patients. Regarding outcome to death, we found as determinant factors: constitutional symptoms, skin vasculitis, pericarditis, arterial hypertension, acute renal injury, chronic kidney disease (CKD), neuropsychiatric involvement (NP-SLE), thrombocytopenia, higher score of current SLEDAI-2K and SLICC/ACR-DI and higher use of cyclosporine, plasmapheresis, and intravenous immunoglobulin. In the Cox regression models, NP-SLE (HR = 2.56, 95%CI = 1.48–4.42) and CKD (HR = 4.33, 95%CI = 2.33–4.72) were significant risk factors associated to death. Overall patient survival after diagnosis at 5, 10 and 15 years were 97.0%, 95.4% and 93.8% respectively.

CONCLUSION

Neuropsychiatric involvement with systemic lupus erythematosus and CKD are risk factors associated to mortality. Lupus nephritis and neuropsychiatric involvement remain major concerns in patient management. The overall mortality rate was low, most related to sepsis or infection.

KEYWORDS

Systemic lupus erythematosus, Mortality, Lupus nephritis, Neuropsychiatric, Childhood.