Systemic EBV-Positive Lymphoproliferative Disease of Childhood

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Introduction

Epstein-Barr virus (EBV) is a common herpesvirus usually causing mild or asymptomatic illness. However, in children and patients with immune dysregulation, EBV can drive excessive lymphoproliferation leading to systemic EBV-positive lymphoproliferative disease. Characterized by unrestricted expansion of EBV-infected T or natural killer cells particularly in children, this aggressive neoplasm can manifest as prolonged fever, hepatosplenomegaly, cytopenias, and, infrequently hemophagocytic lymphohistiocytosis.

Objectives

This study analyzes the epidemiological trends and survival outcomes in the past decade using the SEERstat data.

Method

Cases of systemic EBV-positive T-cell lymphoproliferative disease of childhood (SETLC) were extracted using ICD-10 code 9724/3 from the SEER Plus Research Data, November 2023 sub. Data were stratified by age, gender, income, treatment with chemotherapy, and year of diagnosis (YOD).Survival curves were then compared using the Gehan-Breslow-Wilcoxon test (GraphPad Prism).

Results

64 cases of SETLC were identified from 2010 to 2021, of which 64% were males. Median age was 14 years. Racial distribution was observed as 25% White, 40.6% Hispanics, 7.8% Black, 20.3% Asian/Pacific Islanders, 4.7% American Indians/Alaskans and 1.6% of unknown race.

The overall median of survival (MoS) was 21 months, with a 1-year OS of 55% (CI 95%, 41.86-66.4), and 5-year survival of 42.5% (CI 95%, 29.8-54.6). Survival analysis was significant for chemotherapy, with 23 months for those who received it compared to 2 months without chemotherapy (p value 0.0436). The MoS was higher for cases diagnosed from 2010-2015, while the survival was lower for cases identified between 2016-2021 (p value 0.0294). Survival curves were insignificant based on age, gender and household income.

Conclusion

Systemic EBV-positive T-cell lymphoproliferative disease of childhood is an ultra-rare hematologic malignancy, with only 64 cases reported on SEER database between 2010 and 2021. Superior survival was associated with the use of chemotherapy and YOD (2010-2015), while age, gender and income had no influence on survival. Higher survival with earlier years of diagnoses can be attributed to mutations in viral genome with time leading to more aggressive disease and hence lower survival.This study emphasizing the need for further research to better understand the viral genomics and response to various treatment modalities.