Langerhans Cell Histiocytosis: A Retrospective Analysis Using the SEER Database

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INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare neoplastic disease, characterized by clonal expansion of Langerhans cells in the background of cellular inflammatory infiltrate. It is a heterogenous disease that may involve either a single organ or multiple organs.

AIM

To determine the epidemiology and survival trends for LCH in the past two decades based on SEERstat data.

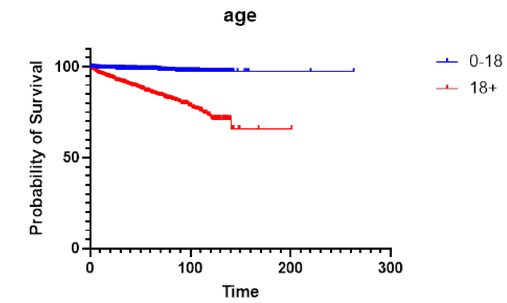
METHOD

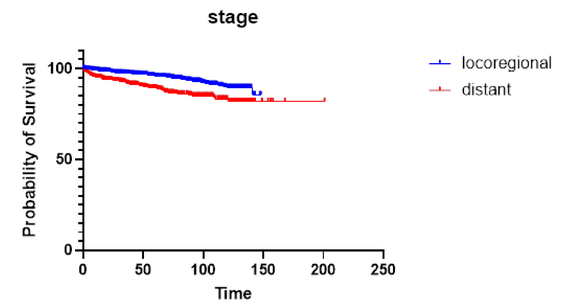
Langerhans cell histiocytosis cases were collected from Surveillance, Epidemiology and End Result database Research Plus Data, 17 Registries, Nov 2023 Sub (2000-2021), using the ICD Code 9751/3. The analysis was stratified based on age, sex, race, year of diagnosis, stage, primary site labelled, laterality, household income and various treatment modalities. Survival curves were compared using the Log-Rank test (GraphPad Prism).

RESULTS

- Total 2022 cases of LCH were extracted, of which 56% were males.
- Racial distribution was noted as: Caucasian (54.5%) followed by Hispanics (27.6%), Asians/Pacific Islanders (8.5%), Blacks (6.7%), Alaskans/American Indians (1.14%) and unknown race (1.63%).
- Median age of diagnosis was 11 years.
- 1-year overall survival was 97.7% (CI 95%, 96.9-97.1), and 5-year OS was 93.4% (CI 95%, 92-94.5).
- The median of survival (MoS) was higher for 0-18 years of age and declined for age 18+ years (p <0.0001).
- Survival based on race revealed higher MoS for Hispanics followed by Asian/PI, Black, Caucasians, and AI/Alaskans (p 0.0004).
- Anatomically, analysis concluded better outcomes for LCH originating from head/face/neck, abdomen and pelvis, and connective tissue and worse survival for thoracic origin (p <0.0001).
- Survival curves based on laterality showed undefined MoS for unilateral (either side) disease compared to 141 months for bilateral disease (0.0001).
- Additionally, locoregional disease had improved survival compared to distant stage disease (p <0.0001).
- Survival analysis was insignificant for gender, income, year of diagnosis, and treatment types.

P.S. Undefined MoS were likely observed due to the lack of significant numbers of death in those categories to calculate 50% survival probability.





CONCLUSION

- Langerhans cell histiocytosis (LCH) is a rare malignancy, with only 2022 cases identified in a period of 22 years, from 2000 to 2021
- Our Study concluded superior survival outcomes associated with younger age, Hispanic origin, unilateral and locoregional disease and neoplasm involving head/face/neck, abdomen and pelvis, and connective tissue.
- Gender, income, year of diagnosis, and treatment types had no influence on survival.

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