**P1139 A POPULATION-BASED STUDY OF ISOLATED ADULT PULMONARY LANGERHANS CELL HISTIOCYTOSIS IN THE UNITED STATES: 2010-2017**

**Topic:** 18. Indolent and mantle-cell non-Hodgkin lymphoma - Clinical

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**Background:** Isolated adult pulmonary Langerhans cell histiocytosis (pLCH) is a rare histiocytic neoplasm that involves the lungs as single system disease. Retrospective studies have suggested that patients with isolated adult pLCH have better overall survival compared to multisystem LCH. While population-based studies have been done for LCH with multisystem disease, there has been no epidemiologic studies done to date on isolated adult pLCH.

**Aims:** To characterize the incidence and outcomes of isolated adult pLCH in the United States.

**Methods:** We queried the Surveillance, Epidemiology, and End Results Program (SEER) 18 Registries (2010-2017) using ICD-0-3 code 9751/3 with the primary site of involvement being the lung. Patients were included if they were ≥ 18 years and had localized pulmonary involvement. Patients with extrapulmonary disease or unknown staging were excluded. The primary outcome was overall survival (OS), which was analyzed using the Kaplan-Meier method. Data on incidence rates (IR) and relative survival (RS) were calculated using the SEER*Stat software. IR (cases/1,000,000) was age-adjusted to the U.S. 2000 standard population. RS was defined as the ratio of the proportion of observed survivors in a cohort of isolated adult pLCH to the proportion of expected survivors in a comparable set of individuals that do not have isolated adult pLCH adjusting for the general survival of the US population for race, sex, age, and time when the diagnosis was established.

**Results:** In SEER, there were a total 147 patients with isolated adult pLCH. Between the years 2010-2017, the median IR of isolated adult pLCH was 0.3 (range 0.1-0.5) and did not change significantly. The median IR from 2010-2013 was 0.2 (range 0.1-0.2) and 2014-2017 was 0.5 (range 0.3-0.5). The median age at diagnosis was 53 years (range 23-79) and 54 (37%) were male. 115 (78%) were white, 21 (14%) were black, and 11 were labeled as other. 116 (79%) were diagnosed on histopathologic evaluation while the remainder were diagnosed without microscopic confirmation. No patients were documented to have secondary malignancies.

With a median follow up of 35 months, the median OS was not reached (95% CI: not reached-not reached) and the 3-year OS was 89%. The 3-year OS between males and females was 93% and 87% (p=0.16) respectively. The 3-year OS between white and non-white was 90% and 87% (p=0.98) respectively. In the general U.S. population, the expected survival for 1, 2, 3 years were 99.2%, 98.5%, 97.8% respectively. In contrast, the RS for patients with isolated adult pLCH at the same time points were 98.3%, 94.8%, 90.9% (Figure). A total of 14 patients died. 3 patients died from isolated adult pLCH while 11 patients died of other causes, including chronic obstructive pulmonary disease (3), cardiac disease (3), chronic liver disease (1), and unknown (4).

**Image:**

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Summary/Conclusion: The incidence of isolated adult pLCH has not changed over the years from 2010-2017. Patients with isolated adult pLCH have lower relative survival compared to the general population. Among patients who died, the majority died from cause other than pLCH.