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Clinical details of early-onset neuroendocrine neoplasms differ by race and ethnicity and overall differ from typical-onset

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BACKGROUND

The epidemiology of neuroendocrine neoplasms (NENs) in younger adults is complex, and it is unclear who presents with which distinct clinical features. Expanding upon our prior work demonstrating a rising incidence of early-onset (EO) NENs overall and differences by race and ethnicity (RE), we characterize detailed clinical features of EONENs by patient demographics in the diverse California population.

METHODS

All patients with malignant NENs diagnosed from 1992-2019 in the population-based California Cancer Registry were identified by histology (ICD-O-3 code 8013, 8041-5, 8150-5, 8240-9). Patients diagnosed by age 49 were designated as EO, and from age 50 as typical-onset (TO). Proportions and age-adjusted incidence rates were calculated for comparisons. Patient demographics, tumor, and other clinical characteristics were compared by onset age. Clinical details of EONENs were compared by patient demographics. Comparisons used Kruskal-Wallis or Pearson chi-squared tests, multivariable logistic regression, and incidence rate ratios.

RESULTS

There were 12,266 EONEN patients identified. Compared to the 107,860 TONEN patients, statistically significantly more of the EONEN patients were women (55% vs 49%), Hispanic (28% vs 12%), or Asian/Pacific Islander (API) (10% vs 7%). More had localized-stage (50% vs 27%), appendiceal primary (12% vs 1%), or rectal primary (15% vs 6%). Fewer of the EONENs were distant-stage (29% vs 52%), or large-cell or small-cell (LSSC) lung NEN (16% vs 49%). Temporal trends in EONENs were similar to TONENs, both showing increasing proportions of patients over time who presented with localized, low-grade, or gastrointestinal primaries, and fewer with metastatic, high-grade, or pulmonary NENs. Furthermore within the EONENs group itself, statistically significant clinical differences were seen across patient demographics. The Hispanic population presented with the most locoregional-stage EONENs (69%; an 18-84% higher odds than other REs), followed by API (67%). The highest incidence rates of small bowel (0.30/100,000 person-years; 177-609% higher than other REs) and extra-gastroenteropancreatic/extra-pulmonary primary (0.39/100,000; 133-155% higher) were among the non-Hispanic (NH) Black population; highest of colon (0.33/100,000; 121-316% higher) and appendix (0.26/100,000; 157-418% higher) among NH White.

CONCLUSIONS

We found differences in the clinicopathologic features of EONENs by demographics and compared with TONENs in California. The increasing proportion of localized disease over time may reflect improvements in diagnostic methods and increased awareness of NENs. Younger patients were much less likely to present with LC/SC lung NENs, which may reflect declining smoking rates. Further research is underway to better understand the mechanisms driving differences in EONENs.

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