Incidence and survival of sebaceous carcinoma in the United States

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Background: Information on risk factors, epidemiology, and clinical characteristics of sebaceous carcinoma (SC) is limited.

Objective: We sought to analyze trends in SC in the United States from 2000 through 2012.

Methods: We used data from the 18 registries of the Surveillance, Epidemiology, and End Results (SEER) Program from 2000 to 2012 to calculate the cause of death, relative frequencies/incidences, 5-/10-year Kaplan-Meier survival, hazard ratios, and incidence rates for SC. Each parameter was analyzed by age, location of occurrence (ocular/extraocular), race, sex, and SEER registry.

Results: Overall incidence was 0.32 (male) and 0.16 (female) per 100,000 person-years. Incidence significantly increased, primarily because of an increase among men. Incidence among whites was almost 3 times the rate among non-whites. Male sex ($P < .0001$), black race ($P = .01$), and extraocular anatomic location ($P < .0001$) were associated with significantly higher all-cause mortality. However, overall case-specific mortality for SC decreased significantly.

Limitations: Underregistration of patients in SEER registries, lack of verification of individual diagnoses, and low levels of staging data because of low stage-classification rate are limitations.

Conclusions: The overall incidence of SC is increasing significantly. Male sex, black race, and extraocular occurrences are associated with significantly greater mortality. (J Am Acad Dermatol 2016;75:1210-5.)

Key words: cutaneous; epidemiology; extraocular; incidence; mortality; ocular; sebaceous carcinoma; skin cancer; survival; Surveillance, Epidemiology, and End Results.

Sebaceous carcinoma (SC) is a rare and very aggressive cutaneous tumor with a diagnosis most often split into ocular and extraocular.[1,2] However, both sites of occurrence have been noted to metastasize to regional lymph nodes.[3] Very few studies have investigated the prognosis, risk factors, and incidence of this disease. In a review of previous case reports of fewer than 200 patients, it was shown

Abbreviations used:

APC:	annual percent change
CI:	confidence interval
DFSP:	dermatofibrosarcoma protuberans
HR:	hazard ratio
SC:	sebaceous carcinoma
SEER:	Surveillance, Epidemiology, and End Results

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that SCs primarily affect the periorbital areas and are the fourth most common neoplasms affecting the eyelid; in addition, more than 70% of reported cases of extraocular SCs in these case reports were located in the head/neck region.[1]

A previous retrospective study based on 1349 cases from the Surveillance, Epidemiology, and End Results (SEER) 1973 through 2004 data reported no observed difference between the prognoses for orbital and periorbital involvement,[4] a predominance of Asians/males in the incidence of SC,[1,4-8] and that the relative 5- and 10-year population-matched survivals were 91.9% ± 1.9% and 79.2% ± 3.7%, respectively.[4] Increasing age, tumor grade (poorly differentiated), and metastasis have been shown to increase risk of death as a result of SC.[9] From 1973 to 2007, 34.5%, 42.8%, and 22.7% of SC cases were located on the eyelid, extraocular head/neck, and other extraocular regions, respectively.[10] In addition, ocular SC has been shown to have greater aggressiveness and rate of metastasis than extraocular head/neck SC.[10] However, relative frequencies/incidence rates, cause-specific survival, and hazard ratios (HR)/cause of death have not been previously stratified by age, race, sex, and site of occurrence.

CAPSULE SUMMARY

- Information on risk factors, epidemiology, and clinical characteristics of sebaceous carcinoma is very limited.
- Overall incidence is increasing significantly; male sex, black race, and extraocular occurrences are associated with greater all-cause mortality.
- Recognition of changing epidemiology of sebaceous carcinoma is crucial in understanding its cause and determining approaches to prevention and treatment.

METHODS

The National Cancer Institute SEER-18 registry was used from 2000 to 2012 for this study. SEER-18 was established in 2000, contains 18 different registries from across the nation, and now accounts for over 28% of the total US population.[11] The SEER registries collect nationwide data on patients with diverse cancers; this information includes demographics, tumor characteristics (eg, primary site, morphology, stage at diagnosis), treatment type, and status.

Patients given a diagnosis of SC as their first cancer were included in this study. SC diagnosis was defined as code 8410/3 in the *International Classification of Diseases for Oncology, Third Edition*.[12] Patients only given a diagnosis by death certificate and autopsied patients were excluded from analysis (2 patients excluded).

We performed descriptive (demographic) analysis of the SEER-18 SC population. The cases were grouped by sex, 5 age-at-diagnosis groups (<19, 20-39, 40-59, 60-79, and >80 years), 4 racial groups (white, black, other, and unknown), and 2 anatomic tumor sites (ocular and extraocular). Cases were further stratified by anatomic site according to age at diagnosis.

SEER*Stat 8.1.5 (National Cancer Institute, Rockville, MD) was used.[13,14] Incidence rates were expressed as cases per 100,000 person-years, and overall incidence/annual incidence rates were calculated for the entire SC population from 2000 to 2012. Overall incidence rates and incidence rate ratios were calculated for each race, sex, and registry. Unknown race was excluded from race analysis because of small sample size (N = 101). In addition, age-adjusted incidence rates (age-adjusted to the 2000 US Standard Population) were calculated at year of diagnosis for males, females, whites, and blacks separately. SEER*Stat 8.1.5 (National Cancer Institute). Joinpoint regression analysis was used to calculate annual percent change (APC) in incidence rates over time for all population subsets (whole population, male/female, white/black). Age-specific annual incidences for the entire SC population and by sex/race were also calculated. Furthermore, peak incidence in each decade was calculated by sex.

Observed and relative 5-/10-year survival were measured with SEER*Stat 8.1.5 using the Kaplan-Meier method. Relative survival refers to the ratio of observed survival in a cohort of patients with SC to the proportion of expected survival in a cohort of cancer-free individuals adjusted for race, sex, and age. Only cases that displayed malignant behavior (*International Classification of Diseases for Oncology, Third Edition* behavioral code 3[12]), were actively followed up, and had a known age were considered for incidence/survival analyses.

The log-logistic accelerated failure time model was used to calculate estimated HR to determine the extent to which age, race, sex, and anatomic site were associated with survival among patients with SC. The reference categories for age, race, sex, and anatomic site used to determine statistical significance were older than 19 years, white, female, and ocular, respectively. In addition, both observed and relative 5- and 10-year survival were calculated for localized and regional staged SC.