**ABSTRACT**

Objective: Malignant melanoma accounts for nearly 75% of all skin cancer deaths, and the incidence is on the rise in the United States. The relative paucity of studies on external ear melanoma (EEM) cases has provided little long-term data regarding the clinical behavior of this melanoma variant. This study analyzes the demographic, clinicopathologic and survival characteristics of EEM.

Methods: The Surveillance, Epidemiology, and End Results (SEER) database (1973-2012) was queried for SNFS cases. Data were analyzed with respect to various demographic and clinicopathologic factors. Survival was analyzed using the Kaplan-Meier model.

Results: External ear melanoma occurred most frequently in the 6th and 7th decades of life. Mean age at diagnosis was 65.5 (±6.8) years. However, the incidence of EEM in adolescents and young adults (ages 15-39 years) has increased by 111.9% from 1973 to 2012. There was a strong male predilection with a male-to-female ratio of 6.4:0.1. The most common histologic subtype was malignant melanoma, NOS (46.8%), followed by superficial spreading melanoma (21.4%), and lentigo melanoma (17.9%). The majority of cases were localized at the time of presentation (88.0%), with rare distant metastasis (9.6%). The most common treatment for modality was surgery alone (97.6%), followed by surgery with RT (2.3%). Ten year disease-specific survival was better among those treated with surgery alone (90.7%), than those treated with surgery plus RT (37.3%).

Increasing Breslow’s thickness and presence of an ulcerating lesion were both associated with poorer survival (p < 0.0001).

Conclusion: This study represents the largest cohort of EEM. It has an excellent survival outcome with surgery alone being the treatment of choice.

**RESULTS**

**INTRODUCTION**

Head and neck melanomas constitute 20% of the primary malignancies with the ear representing 4-6% of this percentage. Head and neck melanomas encompass 1% of all primary cutaneous melanomas. Factors for increased risk are: sun exposure, fair skin, genetic factors, and personal history of melanoma. The current literature discussing EEM comprises only of case reports and retrospective case series. Utilizing the Surveillance, Epidemiology, and End Results (SEER) registry, a powerful resource for evaluation of cutaneous melanomas, was utilized to obtain data for physicians caring for the ear compromised by melanomas diagnosis. The SEER registry was used to evaluate the incidence of EEM as well as patient demographics, treatment modalities, and survival outcomes.

**MATERIALS & METHODS**

Frequency and survival data were obtained from the SEER 18-dataset representing 18 cancer registries covering approximately 28% of the US population. The years included in these datasets were diagnosed between 1973 and 2012. As this database contains only deidentified patient data, this analysis was exempt from institutional review board (IRB) review by the IRB of Rutgers New Jersey Medical School, Newark, New Jersey. The SEER database was queried for EEM cases using the International Classification of Diseases for Oncology, 3rd Edition (ICD-O-3) topographical and morphological codes. Melanoma cases were screened using the morphological codes pertaining to the external ear (C306.0-180). Cases based on the topographical codes, melanoma cases were restricted to the skin of the external ear (Topographical code C369). Frequency scores were stratified and analyzed by age, gender, race, location, histologic grade, tumor size, SEER extent of disease, and treatment modality. SEER extent of disease is categorized into localized, regional and distant disease. Localized tumors are defined as tumors limited to the skin of the ear. Regional tumors are defined as expansion of the tumor to adjacent regions. Distant disease is defined as metastasis to the skin of the body as well as distant metastasis.

**REFERENCES**

**DISCUSSION**

External ear melanoma is a rare entity. This study analyzes the largest cohort of EEM cases to date with over 4,000 cases reviewed in our SEER database. Concerning previously reported literature, there was a strong male predominance (65.5% to 97.6%) found in this cohort. A male predominance of 65.5% to 97.6% was reported in the literature, with an additional predominance found among fair-skinned individuals. One may speculate that later birth in those who may provide greater service-shielding effects, preclude the external ear from easily decreasing the incidence of EEM in females. Despite the incidence is in females, there is no gender disparity in survival, with both groups having nearly identical survival rates at 5 and 10 year time points. The exact reasons for these observations are unclear, but may be a target of future research.

External ear melanoma primarily affects the white population (99.2%) with non-white population having a much lower incidence (0.8%). It is well known that melanoma aggressiveness parallels the risk with the reported effects of UV radiation. Our study supports the negative correlation between degree of skin pigmentation and incidence of EEM.

The mean age of diagnosis was 78.2 years, which was higher than the mean age reported in the literature of 68.4 years. One speciality for the older age diagnosis could be that the external ear is not routinely inspected for dermatologic malignancies. The diagnosis delay resulted in the lesions to grow and become more metastatic at an advanced age. Incidence analysis by age showed a marked increase (111.9%) in incidence of EEM in adolescents and young adults (ages 10-19 years) from 1973 to 2012. A recent SEER study by Milias et al. at Roswell Park Cancer Institute revealed a 25% increase in incidence of melanomas in the entire body in young adults and adolescents (ages 15-24 years) from 1973 to 2011. Although the incidence of EEM in this age group is lower than the incidence of melanomas in the entire body, this could be an increasing trend in the incidence of melanomas diagnosed in adolescents and young adults. Melanoma is the third most common cancer in this young age group, and the incidence has slowly been increasing. More awareness and effective prevention strategies should be instituted to address the issue of melanoma in this young age group.

Majority of the EEM are reported in the western United States (64.2%), followed by southeastern (37.1%) and northeastern (6.6%). Side and Northeastern have previously used the SEER database to analyze the incidence of the entire body. They found that higher UV indices were significantly associated with an increased incidence of EEM. Similarly, many other studies from California, Arizona, New Mexico, etc have had the highest UV indices reported among all states, which may explain the high incidence of EEM in these regions. Furthermore, Ringel et al. at the University of California reported a higher incidence of melanoma in coastal versus inland counties in California. Despite these findings, there is a need to examine the incidence of EEM in other races.

The histographic types of melanomas observed in our patient population were similar to those previously reported (40.3% superficial spreading (40.3% to 68.9%), 33.7% lentigo maligna (15.6% to 40.3%), and nodular (26.6% to 43.7%)). Survival analysis comparing EEM at different time periods revealed that lentigo maligna melanoma had the worst survival rate and nodular melanomas had the worst survival rate (5-year OS 92.9% versus 75.9% to 64.3%). Lentigo maligna melanoma arises in a lentigo maligna, and manifests as a persistent pigmented macule or plaque prior to invasion. However, it could be more easily picked up on physical exam, and potentially treatable at an early, curable stage leading to better survival outcomes with this subtype of melanoma, in contrast, nodular melanomas manifest as a vertical growth phase dorsum, and is known to contain a close of cellular nevus cells capable of metastatic progression leading to poorer survival outcomes. Nodular melanoma is well known to metastasize to regional lymph nodes. Common regional sites of metastasis include both parotid and submandibular lymph nodes, anterior and posterior cervical and paratracheal nodes, and the parotid gland. Nodular melanomas that are associated with lymph node involvement were the most common location of metastasis in our study (10.1% regional lymph node involvement).

Five and 10 year OS rates for EEM were 89.9% and 84.2% respectively. Similar survival rates have been reported in previous literature. Lentigo maligna melanoma had the best survival, and was the most commonly diagnosed melanoma in this study (40.3%). Nodular melanoma had the worst survival rate (5-year OS 92.9% versus 75.9% to 64.3%). Lentigo maligna melanoma arises in a lentigo maligna, and manifests as a persistent pigmented macule or plaque prior to invasion. Therefore, it could be more easily picked up on physical exam, and potentially treatable at an early stage (curable stage). Survival outcomes with this subtype of melanoma can thus be an important factor in the clinical decision-making process of patients with EEM. The SEER database included sites of EEM in this dataset. Hence, the incidence and prevalence of EEM, incidence and prevalence of cases in this dataset are highly dependent on the database.

The SEER program was not able to acquire detailed tumor characteristics and survival information due to the small number of cases. The SEER program was not able to acquire detailed tumor characteristics and survival information due to the small number of cases.

The primary treatment modality for EEM is surgical excision with or without lymph node dissection. Radiation therapy has been used as an adjuvant treatment for patients undergoing surgery with significant cutaneous disease to reduce the risk of metastatic disease. Post-surgical adjuvant radiation therapy may have a more advanced disease requiring the use of adjuvant radiation treatments for better tumor control.