ABSTRACT

Outcome objectives: summarize the epidemiology of sarcomas occurring in the head and neck; identify prognostic factors for patient survival

Setting and methods: cross-sectional analysis of the National Cancer Institute’s Surveillance, Epidemiology and End Results (SEER) Program

Results: A total of 10,578 cases of sarcoma arising in the head and neck were identified. Of head and neck sites, the skin and soft tissue were most commonly affected (n=6432, 61%), followed by the bones of the skull and face (n=1331, 13%) and the oral cavity (n=1078, 10%). Disease-specific 2-, 5-, and 10-year survival rates were 69.5%, 55.9%, and 46.9%, respectively. On multivariate analysis, male gender, the administration of radiation, and advanced stage conferred statistically significant increases in hazard ratios (p<0.001). A propensity-matched model controlling for variables associated with radiation use yielded consistently worse outcomes in patients receiving radiation (p<0.001).

Conclusion: Sarcomas, a heterogeneous group of malignant mesenchymal tumors, are uncommonly found in the head and neck. This study represents the largest analysis of patients with head and neck sarcoma in the literature and demonstrates the impact of age, and gender, histology, tumor location, and radiation status on overall prognosis.

INTRODUCTION

Sarcomas are a heterogeneous group of mesenchymal malignant neoplasms. They account for 2% of all head and neck malignancies, while between 4 and 10% of all adult sarcomas occur in the head and neck. Metastases are relatively uncommon, and are only seen in approximately 10% of patients at presentation. The 5-year survival of head and neck sarcomas has been reported at 60%. While sarcomas not arising in the head and neck largely conform to the disease progression of metastatic disease, patients with sarcoma in the head and neck primarily succumb to local recurrence. This has generally been attributed to the proximity of vital structures in the head and neck, and may be related to the inherent difficulty in obtaining wide margins during surgical resection while limiting concomitant morbidity. Given the relative infrequency of head and neck sarcomas, previous series have been limited by modest sample size. In addition, previous studies have demonstrated a bias toward pathologies treated exclusively by head and neck surgeons, resulting in the exclusion of a large subset of sarcomas in the head and neck-those involving the skin and soft tissues.

METHODS

The SEER 18 registries, comprising data from 1973 to 2010, were used for analysis. Inclusion criteria consisted of the following: records with a diagnosis of a specific subtype of sarcoma or sarcoma NOS; records reporting a primary site in the head and neck, excluding the central nervous system and orbit; records corresponding to one primary tumor or the first primary tumor in patients with two or more tumors. Statistical analyses included extraction of descriptive data and Kaplan-Meier plots. Predictors of overall and cause-specific survival were evaluated using univariate and multivariate Cox proportional hazard models. A propensity score matched model was used to investigate the effects of radiation on survival.

RESULTS

A total of 10,578 cases of head and neck sarcoma were identified in the SEER 18 registries. There was a male preponderance (n=7477, 71%, versus female, n=3101, 29%). The median age group affected was 40-44 years. A majority of cases were diagnosed in the 2000s (n=5553, 53%) over any other decade (1970s, 1980s, and 1990s). The most common primary site was the skin and soft tissues (n=6432, 61%), followed by the bones of the skull and face (n=1331, 13%) and the oral cavity (n=1078, 10%). The most common histologic subtypes were malignant fibrous histiocytoma (MFH, n=2629, 24.9%), followed by Kaposi sarcoma (KS, n=2210, 20.9%), rhabdomyosarcoma (n=862, 8.1%), sarcoma not otherwise specified (n=749, 7.1%), and hemangiosarcoma (n=713, 6.7%) (Figure 1). TNM stage at diagnosis was recorded in only a subset of patients (n=856); of these patients, stage I accounted for 286 patients (33.4%), stage II accounted for 272 patients (31.8%), stage III accounted for 82 patients (9.6%), and stage IV accounted for 216 patients (25.2%).

Radiation status was reported as given (n=7053, 66.7%), not given (n=3254, 30.8%), or unknown/refused (n=271, 2.6%). Surgical or multimodality therapy was inconsistently reported, and was therefore intentionally omitted from analysis. Roughly equal numbers were reported as alive (n=5258, 49.7%) or dead (n=5320, 50.3%) at the time of data query, while cause-specific mortality was expectedly lower (death attributable to sarcoma diagnosis, n=3310, 31.3%; alive or dead of other cause, n=7268, 68.7%). Overall 2-, 5-, and 10-year survival rates were 70%, 57%, and 47%, respectively (Table 1). Kaplan-Meier plots were generated for all-cause mortality as well as cause-specific mortality based on several parameters, including gender, age at diagnosis, decade of diagnosis, histology, site, TNM stage group, and radiation status (log-rank test p<0.001; Figure 2). Both univariate models and multivariate models were used with Cox regression to generate hazard ratios. On multivariate analysis, female gender, decade of diagnosis from 2000-2010, no radiation, and early stage (T1 or T2) conferred lower hazard ratios that reached statistical significance (p<0.001). Histologies notably associated with excellent survival included MFH and liposarcoma, while histologies associated with poorer survival included hemangiosarcoma and Kaposi sarcoma. Sites associated with excellent survival included the skin and soft tissues, while the oral cavity was associated with poorer survival.

The association of radiation with poorer prognosis was investigated further using a propensity score matched model. A propensity score representing the probability of receiving radiation treatment was calculated for each patient using a logistic regression model which identified the variables associated with radiation use. Variables included age, decade of diagnosis, site, histology, stage, presence of metastases, and gender. After 1:1 matching, two groups of n=3254 each (radiation group and matched controls) were obtained. On regression analysis of cause-specific death, radiation given was still associated with a statistically significantly higher hazard ratio compared with no radiation received (HR=1.53, 95% confidence interval, 1.39 to 1.68, p<0.001).

DISCUSSION

Head and neck sarcomas represent a heterogeneous group of malignancies. While the most common entities encountered in our clinical practice as head and neck surgeons include rhabdomyosarcoma, chondrosarcoma, and osteosarcoma, among others, the epidemiologic data presented here demonstrate that MFH and KS represent nearly a majority of head and neck sarcomas. This discrepancy likely reflects the number of sarcomas diagnosed and managed either medically or surgically by providers outside of the specialty of head and neck surgery. The present study reiterates previously reported data-namely, that male gender and advanced TNM stage group (III and IV) are associated with worse prognosis.

This study introduces two novel findings. Decade of diagnosis from 2000 to 2010 was significantly associated with improved survival over other decades studied (1970s, 1980s, and 1990s). Differences may be explained by newer treatment paradigms, including treatment for KS. KS, which dramatically increased in incidence following the advent of the acquired immunodeficiency syndrome (AIDS), is a primarily nonsurgical disease representing a significant portion of our study population.

CONCLUSION

To our knowledge, this study represents the largest series on head and neck sarcomas. Otolaryngologists must remain cognizant of the prevalence of sarcomas routinely diagnosed and treated by providers, including oncologists and dermatologists, outside of our specialty. The epidemiologic and survival data presented here provide insights for patient counseling and clinical decision-making. Further work remains to be performed on determining and refining the optimal treatment of head and neck sarcomas.

DISCLOSURES

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