Tumors of the lacrimal gland represent between 5-25% of orbital tumors and can be subdivided into lymphoproliferative, benign epithelial, malignant epithelial, and malignant non-epithelial lesions. Epithelial lesions represent between 23-70% of biopsied cases, and 50% of these lesions have malignant pathology. While pleomorphic adenoma is the most common benign lesion of the lacrimal apparatus, adenoid cystic carcinoma (ACC) is the most common malignant epithelial tumor. Other malignant epithelial tumors of the lacrimal gland include mucoepidermoid carcinoma, adenocarcinoma, acinic cell carcinoma, and squamous cell carcinoma (SCC). Non-epithelial malignant tumors include lymphomas and rarely sarcomas, malignant melanoma, and other mixed tumor types.

Primary malignant neoplastic neoplasms of the lacrimal gland are rare and are associated with significant morbidity and mortality due to early dissemination via bone. The optimal management of these tumors is controversial with proponents of both orbital sparing conservative surgical approaches followed by post operative radiation to orbital exenteration and craniofacial approaches. Intra-arterial cytotoxic chemotherapy has also been proposed as an adjunct treatment modality. Regardless of treatment modality these tumors continue to portend a grave prognosis, with a reported 5-year overall survival ranging from 40-80% depending on the study.

While there are a myriad of small case series and reports, fewer than 250 cases have been reported in the international literature and population data is scarce. The goal of this study was to evaluate the subtypes of tumors of the lacrimal apparatus, and to evaluate treatment modalities and outcomes as related to patient survival over a 14-year period at a single institution. Given the rarity of these tumors there are few series available for review. We compared our results to the published literature.

Materials and Methods

Retrospective case review of the records and pathology of 42 patients with tumors of the lacrimal gland at UCLA medical center. The study was reviewed from 1998-2012. Of these 42 patients, 21 were found to have cancer of epithelial origin. The medical records of these patients were reviewed to determine patient age, presenting symptoms, risk factors, tumor stage, treatment modality. Follow-up of 10 months to 130 months was available in all 21 cases. Clinical staging of disease was performed according to the American Joint Committee on Cancer (AJCC) staging system. Statistical analyses were performed in SPSS 21 (IBM Corp., Armonk, NY).

Introduction

Results

The histopathology of 42 patients with tumors of the lacrimal apparatus was analyzed. 21 (50%) of these tumors represented mucosal associated lymphoma tumors or lymphoma. Of the 21 epithelial tumors, 7 (33%) were represented by squamous cell carcinoma, 5 (24%) were represented by adenoid cystic carcinoma, and 2 (10%) each were represented by mucoepidermoid carcinoma and mixed tumors.

Discussion

Tumors of the lacrimal gland are a rare oncological entity with varied histopathology. Optimal treatment of these tumors is controversial and is likely to vary according to the histopathology of the tumor. Currently, our knowledge of the behavior of these tumors is based on case reports and small single-institution studies. Our data reinforces that ACC represents the most common epithelial malignant of the lacrimal apparatus. In this study, ACC represented 33.3% of epithelial malignancies. Our analysis revealed a surprisingly high rate of SCC of the lacrimal gland, which represented 33.3% of the epithelial malignancies. Previously SCC of the lacrimal apparatus has only been noted in case reports. Primary SCC of the lacrimal gland, similar to the salivary glands, is thought to be a rare entity in comparison to metastatic SCC from primary SCC of the skin. According to UCLA Tumor Registry methodology these tumors were coded as primary tumors. Furthermore, in reviewing the records of these patients there was no history of primary SCC of the skin, however, it is possible that the high rate of SCC found in this cohort could be the result of misclassification of metastatic disease.

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Multivariate analysis revealed that ACC diagnosis, age at diagnosis, and treatment with radiation therapy may influence survival, although these results did not reach statistical significance. Whether or not a patient had ACC did not appear to influence overall survival.

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