ABSTRACT

OBJECTIVES: Although the lifetime risk of developing a second primary head and neck cancer is estimated to be as high as 20 percent, the incidence of a synchronous carcinoma and sarcoma within the head and neck is exceedingly small, especially in the absence of previous radiation therapy. We report the unique case of a primary angiosarcoma of the scalp and a synchronous second primary squamous cell carcinoma (SCC) in situ of the right tonsil in a 69-year-old male, and discuss diagnosis and management.

STUDY DESIGN: Retrospective chart review.

METHODS: The medical records of a single patient treated at a tertiary academic medical center for angiosarcoma of the scalp and SCC of the tonsil in situ were reviewed.

RESULTS: A 69-year-old male presented to the Head and Neck Surgery Clinic with a one-month history of bleeding scalp mass. He denied tobacco use or prior exposure to ionizing radiation, but did endorse a history of heavy alcohol use. Physical examination revealed a 6 x 6 cm scalp vertex mass with biopsy confirming epithelioid angiosarcoma as well as a 2.5 cm friable, tender right tonsillar mass. He underwent a wide local excision of the scalp, direct laryngoscopy, and tonsillectomy. Negative margins were achieved, and pathology for the tonsillar mass returned as SCC in situ. He was referred for postoperative radiation therapy for both primary sites.

CONCLUSION: Synchronous malignancies of the head and neck are defined as malignancies in two separate anatomic sites diagnosed within 6 months of each other. Though rare, they pose a unique challenge for the otolaryngologist. Optimal management involves considering each malignancy separately while remaining cognizant of the impact of the treatment of one on the other.

INTRODUCTION

A double primary, as defined by Warren and Gates, must meet three criteria: (1) each tumor must present a definite picture of malignancy; (2) each tumor must be distinct; and (3) the possibility that one was a metastatic lesion from the other must be excluded.1 Double primaries of the head and neck are relatively common, and tend to occur within the oral cavity, up to a rate of 10-20% within the first 10 years from initial diagnosis of one primary.2 This incidence increases to 30% after 10 years from initial diagnosis.3 In general, double primaries of the head and neck tend to be of similar histologic origin, notably epithelial malignancies such as squamous cell carcinoma. The concept of field cancerization as introduced by Slaughter speculates that upper aerodigestive tract mucosa, once exposed to disease-modifying irritants, undergoes molecular changes that predispose the remaining "field" to higher risk of malignancy.4

In the rarest of cases, a double primary consisting of an epithelial and a mesenchymal malignancy may be observed.5 We report the unique case of a 69-year-old male patient who was diagnosed with a primary epithelioid angiosarcoma of the scalp and a synchronous second primary squamous cell carcinoma (SCC) in situ of the right tonsil.

CASE PRESENTATION

The patient is a 69-year-old male who presented to the head and neck surgery clinic with a slow-growing, bleeding lesion on his scalp for 1 month. He sought evaluation by his dermatologist, who performed a shave biopsy which returned as epithelioid angiosarcoma. The patient had no other complaints. He has a history of Parkinson disease and prostate cancer (treated with prostatectomy). A complete head and neck exam demonstrated a 4.5 cm friable, bleeding, indurated lesion on the scalp vertex just above nuchal incision. Incidentally, he was also noted to have a 3 cm exophytic, minimally tender, friable mass emanating from the right tonsillar fossa. No cervical lymphadenopathy was appreciated.

Magnetic resonance (MR) imaging of the brain and neck demonstrated that the posterior parietal scalp mass did not appear to erode into the underlying calvarium. The same scan also highlighted a 1.5 x 2.7 x 2.8 cm right tonsillar fossa mass (Figure 1). Further imaging of the chest, abdomen, and pelvis did not reveal evidence of metastatic disease.

The patient subsequently underwent a wide local resection of the posterior scalp down to periosteum, with negative margins on intraoperative frozen section analysis. There was no involvement of bone. Frozen sections were also taken of the right tonsillar mass, which returned as SCC; he thus also underwent tonsillectomy. The scalp defect was reconstructed using an inferior island trapezoid myocutaneous flap. He tolerated the procedure well. Final pathology for the scalp lesion returned as high-grade angiosarcoma, with a tumor size of 4.7 cm (T4bN0M0, stage IA) (Figure 2). SCC in situ was found in the right tonsil (Tis).

He underwent adjuvant radiation therapy at the scalp after surgery, but developed sternal metastases and local recurrence at the scalp as detected on post-treatment positron emission tomography. He is currently undergoing evaluation for systemic chemotherapy.

DISCUSSION

There have been an exceedingly low number of reports of double primary malignancies involving both epithelial and mesenchymal progenitor cells. Komorn and Obermeyer individually reported on a synchronous SCC and adenocarcinoma of the larynx.5 6 Similarly, Medina-Banegas treated a patient with a SCC in situ and chondrosarcoma of the larynx,7 while Tomidokoro reported a synchronous SCC and leiomyosarcoma of the larynx.8 Finally, Dios et al. were involved in the care of a patient with an SCC and leiomyosarcoma of the oral cavity.9

Due to the overall rarity of the disease, angiosarcoma of the head and neck does not appear to be associated with any known risk factors. It is a highly aggressive, often multifocal, soft tissue malignancy that is associated with a dismal prognosis despite multimodality therapy, including surgery, chemotherapy, and radiation.10 11 The most common site is the scalp, occurring in up to 50% cases according to one series.10 Much in accordance with what was observed in this patient, who initially presented with resectable local disease, patients who undergo surgery and adjuvant therapy nevertheless may develop distant metastases.

SCC in situ (Tis) is composed of malignant degeneration within the entire thickness of epithelium without basement membrane invasion. Although commonly classified within the spectrum of premalignant lesions, Tis is a frank malignancy and should be treated more aggressively.12 Although complete excision is likely to prevent further malignant change, the surrounding mucosa appears to also be at risk for development into premalignant lesions (i.e., dysplasia). As such, continued monitoring for global changes in the various mucosal surfaces is required. In the case of this patient, who had Tis of the right tonsil, complete excision of the tonsil, including its mucosal covering, may be sufficient for local control.

CONCLUSION

This is the first report of a case of a synchronous double primary involving an angiosarcoma of the scalp and a SCC in situ of the tonsil. In this case, tonsillectomy was sufficient to eliminate the tonsillar primary given its early stage, precluding the risk of further comorbidity from adjuvant therapy in two separate sites. This case reminds the otolaryngologist to be cognizant of double primaries and highlights the importance of performing a complete head and neck examination in the evaluation of oncologic patients.

REFERENCES