



Epithelioid Sarcoma of the Tongue: An Unusual Tumor Mimicking Squamous Cell Carcinoma

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Objectives:

To present an extremely rare case of primary epithelioid sarcoma of the tongue and review the diagnosis and management of this unusual malignancy.

Materials and Methods:

Chart review and comprehensive literature review using PubMed and Google Scholar.

Results:

A 17-year-old non-smoker was referred for evaluation of an ulcerative lesion of the right anterior-lateral tongue, progressing over several months. Incisional biopsy was concerning for squamous cell carcinoma. He underwent partial glossectomy and bilateral selective neck dissections, with reconstruction using a radial forearm free flap. Final pathology was consistent with epithelioid sarcoma, proximal type, demonstrating perineural invasion and close margins. Post-operative PET scan did not show any persistent disease. He was referred to radiation oncology for post-operative radiation therapy.

Conclusion:

Epithelioid sarcoma is a rare malignancy that usually presents in the extremities of young adults, which uncommonly presents in the head and neck as a primary or metastatic lesion. The infrequency of these lesions has prevented development of evidence-based treatment recommendations. As with most sarcomas, surgery is the mainstay of therapy for epithelioid sarcoma. Radiation has been used in an adjunctive role. Although a rare lesion, epithelioid sarcoma should be considered in the differential diagnosis for atypical epithelioid lesions of the tongue and oral cavity when patient demographics, gross lesion characteristics, or histopathology and immunohistochemical studies are not entirely consistent or convincing for more common lesions, such as squamous cell carcinoma.