Introduction: Angiosarcoma is an exceedingly uncommon tumor comprising about 1% of all soft tissue tumors, with only a handful of cases reported involving the tongue. We present here a rare case of angiosarcoma of the tongue in the eldest patient known thus far in the literature.

Case Description: A 92 year old Caucasian female with atrial fibrillation and CHF presents with a left sided tongue mass that was slowly enlarging over the past 6 months and causing her some discomfort. She confirms denture use for many years but denies any recent trauma, radiation exposure, or smoking history. Oral exam revealed a soft, warty mass, about 2 cm in diameter on the left lateral margin of the tongue. There was no lymphadenopathy. A PET/CT showed a mass in the lateral left mobile tongue with an enlarged enhancing lymph node in the adjacent left submandibular area (SUV max 10.7 and 3.5 respectively). A biopsy of the mass revealed diffuse high grade squamous dysplasia with focal in situ carcinoma along with focal areas of proliferation of malignant appearing spindle cells. Immunostaining performed with S100, Melan A, and SMA was negative. The specimen showed diffuse positivity to Vimentin and focal positivity to CD31 and CD34, thus supporting the diagnosis of high grade angiosarcoma.

Discussion: Angiosarcoma, a tumor originating in the vessels of the blood or lymphatics, typically occurs in the face and scalp region. It tends to occur more frequently in Caucasians, males, and the 6th decade of life. This tumor usually presents as a purplish-blue nodular lesion or ulceration, with about 10% of patients having lymphadenopathy. Since only a few cases of the tongue have been reported, there are no definitive treatment guidelines available. Based on the limited data, the standard of care is complete surgical resection with wide margins along with radiotherapy. The role of neoadjuvant chemotherapy remains unclear at this point but may be considered for younger patients with aggressive features. Due to her advanced age and preference, she received sole radiotherapy to a total radiation dose of 66 Gy spread over 8 weeks with a daily dose of 200 cGy. She responded well to this treatment and the tumor shrunk leaving behind a small remnant. After 1 year follow-up she has the same remnant lesion without any evidence of metastatic disease.

Conclusion: Angiosarcomas account for approximately 15% of head and neck sarcomas, with only a handful of cases involving the tongue having been reported. The 5 year survival rate for angiosarcomas of the head and neck region is less than 40%. Tumors arising from the tongue tend to be more aggressive with extremely high 2 year mortality, depending on the size and disease extent. This case demonstrates the possibility of using sole radiation therapy for patients who may not be good surgical candidates or opt against surgical intervention with this rapidly advancing malignancy. We hereby conclude that radiotherapy alone may be a viable option for treatment of angiosarcoma of the tongue for patients of advanced age with high morbidity and mortality.