**Introduction**

Objectives  
To discuss the clinical presentation, prognosis and treatment of extramedullary plasmacytomas and present a rare case of a uvula plasmacytoma in a patient previously treated for a conjunctival plasmacytoma.

Study Design  
Case report with literature review

Methods  
The PubMed database was searched from 1956 to 2011, limited to the English language, using the keywords: extramedullary, plasmacytoma, head, neck, uvula, conjunctiva

**Case History**

A 41-year-old male presented to the head and neck surgery clinic with a 3 month history of an enlarging mass on his uvula. His only complaint was an intermittent gagging sensation from the mass resting on the back of his tongue. Physical examination was significant for a 1.5 cm pink, mucosally covered mass arising from the posterior surface of his uvula. Nasopharyngoscopy revealed that the mass extended from the tip of the uvula to the posterior soft palate (Figure 1). Otherwise, there were no other palpitap lesions or lymphadenopathy. His past medical history was only significant for an extramedullary plasmacytoma excised from his right conjunctiva 3 years prior to presentation. An incisional biopsy was performed, which revealed a metachronous extramedullary plasmacytoma of the uvula. He was treated with radiation therapy (5040 cGy) with complete clinical resolution of the lesion (Figure 2). Further work-up was negative for any systemic disease, and he is disease free 8 months post-treatment.

**Background**

Extramedullary plasmacytomas are monoclonal proliferations of plasma cells found in soft tissues. They comprise only 1% of head and neck tumors; however, over 80% are located in the upper aerodigestive tract (UAD). Extramedullary plasmacytomas of the UAD are 3 times more common in males than females. The sinonasal tract and nasopharynx are the most common subsites. To our knowledge only 1 uvula plasmacytoma and less than 10 conjunctival plasmacytomas have been reported. Clinical symptoms of head and neck plasmacytomas vary depending on location of the mass.

**Pathology**

Extramedullary plasmacytoma is a slow growing dermo-hypodermic tumor that is composed of plasma cells with large eosinophilic nucleoli and multiple mitoses in a HPF. Immunohistochemistry for kappa light chains is intensely positive (brown cytoplasm). Lambda light chain positivity (red cytoplasm).

**Physical Examination**

- Pre-radiation Nasopharyngolaryngoscopy
- Post-radiation Nasopharyngolaryngoscopy

**Discussion**

Treatment for isolated plasmacytomas is surgery, radiation, or a combination. Wide local excision is often advocated if negative margins can be obtained without compromising function since it spares the morbidity of radiotherapy. Extramedullary plasmacytomas are very radiosensitive and treatment with at least 4500 cGy is recommended for cure.

The five-year local control rate with primary treatment is greater than 80%. In a case series of 68 patients looking at the the long-term outcomes of patients with head and neck EMPs, the 10-year regional recurrence rate was only 8%. Because of this low regional metastatic rate, there is no current consensus on whether to treat clinically negative neck nodes. Including regional irradiation in the primary treatment of UAD plasmacytomas is based on institutional protocols.

There is ongoing debate on whether plasma cell disorders are distinct diseases versus a continuum of the same disease. Regardless, long-term studies of patients with head and neck EMPs reveal that they have an 8-36% risk of developing multiple myeloma. Consequently, a systemic work-up at the time of diagnosis is critical. This often entails serum and urine protein electrophoresis, skeletal survey, and bone marrow biopsy.

**Conclusions**

Extramedullary plasmacytomas are often first diagnosed by otorhinolaryngologists. Although rare, they should be included in the differential diagnosis of mucosal head and neck tumors. A multidisciplinary approach, including hematologists and radiation oncologists, is needed. Our case of two metachronous plasmacytomas in extremely rare soft tissue subsites emphasizes the need for close clinical follow-up, so that second primaries or even multiple myeloma can be detected at an early stage.

**References**