Peripheral T-Cell Lymphoma Involving the Uvula

Chirag R Patel, MD; Senja Tomovic, MD; Evelyne Kalyoussef, MD; Jean Anderson Eloy, MD, FACS

Introduction

Neoplasms of the uvula are exceedingly rare. Squamous cell carcinoma and minor salivary gland tumors are the most common neoplasms involving the uvula. They are usually associated with involvement of the soft palate as well. Lymphomas of the oral cavity and oropharynx have been previously reported, with the majority involving Waldeyer’s lymphatic ring. B-cell lymphomas are the most common however peripheral T-cell lymphomas have been found in the nasal cavity. We present the first reported case of a peripheral T-cell lymphoma presenting as an isolated uvular mass.

Case Report

A 65 year old Filipino male with no significant past medical history presented to the office with complaints of one year history of a gradually enlarging uvula. He also reported intermittent dysphagia and dyspnea. He denied any history of fevers, chills, weight loss, sore throat, upper respiratory tract infection, odynophagia, or pain and was otherwise asymptomatic. On clinical exam he was noted to have a firm 5 x 3 cm mass isolated to the uvula. It was without edema or erythema and was not tender to palpation. It extending inferiorly to touch the base of tongue and epiglottis. There was no palpable cervical adenopathy. Computed tomography (CT) of the neck with contrast was performed which demonstrated an enlarged uvula without abnormal enhancement and with low density characteristics (Figure 1). By CT criteria this was felt to be consistent with edema. The thyroid gland, submandibular glands, and parotid glands were all normal and no cervical adenopathy was noted. He subsequently underwent an excisional biopsy of the uvula for diagnosis. The specimen was sent to pathology fresh to allow for a full work-up. Initial review of the specimen was consistent with a high grade T-Cell lymphoproliferative disorder. Cytospin preparations revealed numerous small and intermediate sized lymphocytes with clefted nuclei admixed with eosinophils. Flow cytometry revealed a homogeneous population of CD2+, cytoplasmic CD3+, surface CD3+, CD4+, CD5+, CD7-T-lymphocytes. There was an increased CD4:CD8 ratio of 17.4:1. In addition, a cluster of Langerhans cells was also present, consistent with a focus of Langerhans cell histiocytosis.

The patient was subsequently referred to the Hematology and Oncology service for further workup and treatment. Positron Emission Tomography (PET) scan demonstrated a 1.5 cm right epitrochlear node with an SUV of 1.8 and bilateral slightly enlarged inguinal lymph nodes with mild uptake. Biopsy of the epitrochlear lymph node confirmed the diagnosis of peripheral T-Cell lymphoma with cytomorphology demonstrating a homogeneous population of CD2+, cytoplasmic CD3+, surface CD3-, CD4+, CD5+, CD7-, CD62L+, HLA-DR+ T-lymphocytes. Peripheral blood smear demonstrated atypical Sezary cells as well. He subsequently went on to develop multiple cutaneous lesions leading to a working diagnosis of Mycosis Fungoides (MF) or Sezary Syndrome. He was treated with weekly Gemcitabine 1000mg/m2 for 6 weeks, two weeks off, followed by Gemcitabine on days 1, 8, and 15 every 28 days. PET scan performed 8 months after initiating therapy revealed improvement in the inguinal adenopathy and no other abnormal findings. 12 months after completing chemotherapy he reported resolution of all skin lesions.

Discussion

Less than 5% of extranodal non-Hodgkin lymphomas are found primarily in the Oropharynx. Waldeyer’s ring is the most common subsite, with involvement of soft palate and/or uvula being quite rare. Non-Hodgkin’s lymphoma of the oropharynx affects all age groups however the incidence increases later in life. The most common initial complaint is of a painless swelling with or without ulceration. Peripheral T-cell lymphoma (PTCL) is a group of rare and sometimes aggressive NHLs that develop from mature T-Cells. They represent approximately 10-15% of all NHLs in the United States. Cutaneous T-lymphoma (CTCL) is subtype of PTCL in which malignant T-cells migrate to the skin and cause various skin lesions to appear. Mycosis Fungoides (MF) is the most common type of CTCL with Sezary syndrome being considered a late stage of MF. These generally affect the skin but visceral involvement can occur over time. To our knowledge, only 35 cases of oral or oropharyngeal involvement with MF have been reported in the English literature and most reported cases were a late phase presentation. Mucosal lesions preceded the development of cutaneous lesions in only 4 of the 35 cases. Two of the 4 patients had a slow course with remission after treatment. Approximately 50% died within 1 year and nearly all remaining 33 patients died within 3 years of diagnosis suggesting an aggressive disease with poor prognosis.

We present the first case of lymphoma identified as a mass isolated entirely to the Previously reported cases of lymphomas involving the oropharynx involved the soft palate. While the development of cutaneous lesions has led to a working diagnosis of CTCL, the slow course of disease and primary mucosal presentation suggests that a variant may be more likely.

In summary, tumors isolated to the uvula are quite rare. The most common tumors of the uvula are minor salivary gland tumors or squamous cell carcinoma. However, lymphoma should be considered in the differential diagnosis. Biopsy specimens should be handled accordingly to ensure an accurate diagnosis.

References


Abstract

Objectives: Malignant neoplasms of the uvula are rare, with squamous cell carcinoma and minor salivary gland tumors being the most common. Peripheral T-cell lymphoma is a type of non-Hodgkin’s lymphoma (NHL) arising from mature T-cells. Patients commonly present with lymph node involvement; however, extranodal sites may be involved. Involvement of the uvula by this entity has not previously been described in the English literature. We present the first case of a peripheral T-cell lymphoma presenting in the uvula.

Study Design: Case Report

Methods: Retrospective chart review of a patient with peripheral T-cell lymphoma involving the uvula and a review of the literature.

Results: We present the case of a 65 year old Filipino male who presented with an enlarging uvular mass over the course of one year. Excision of the mass revealed a peripheral T-cell lymphoma. Excisional biopsy of an epitrochlear lymph node confirmed the diagnosis.

Conclusions: Lymphoma should be considered in the differential diagnosis of a uvular mass. Surgical specimens should be handled accordingly to ensure an accurate diagnosis.

Figure 1:
A: Axial CT image demonstrating narrowing of the oropharyngeal airway by the uvular mass.
B: Sagittal CT image demonstrating the vertical length of the mass.
C: Coronal image demonstrating the uvular mass in relation to the rest of the airway.