Nodal Recurrence of Salivary Polymorphous Low-Grade Adenocarcinoma

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**Outcome Objectives:**
Polymorphous low-grade adenocarcinoma (PLGA) is an uncommon type of low-grade salivary adenocarcinoma. Risk factors for aggressive phenotypes of PLGA are not definitively known. PLGA with a predominantly papillary pattern may have a higher risk of nodal metastasis. The objective of this report is to examine two cases of late nodal metastasis of polymorphous low-grade adenocarcinoma.

**Methods**
A retrospective chart review was performed at a tertiary, academic medical center. Patient demographic data, medical history, presentation, pathology, treatment modalities and follow-up information were collected.

**Results**
Two patients were identified with late recurrence of polymorphous low-grade adenocarcinoma, isolated to the cervical lymph nodes. After wide local excision, the first patient recurred nine years later with a level II neck mass demonstrating PLGA. After resection and post-operative radiation, he has been doing well without evidence of disease recurrence. The second patient presented with a soft palate and tonsillar lesion. He underwent an uncomplicated resection of the primary site with a final pathology consistent with his previous salivary malignancy.

**Conclusion**
Polymorphous low-grade adenocarcinoma is described in the literature as a rare tumor with low-grade metastatic potential. Recent evidence suggests not all PLGA tumors behave the same. In order to counsel and manage patients with PLGA effectively, further study is needed to determine risk factors for the development of recurrence of this low-grade malignancy.

**Introduction**
Polymorphous low-grade adenocarcinoma (PLGA) was first described as a distinct clinicopathologic entity in 1984 by Evans and Batsakis. Typically, these neoplasms present on the palate and demonstrate indolent behavior. The notable histologic traits of PLGA include uniformity on a cellular level lack of nucellar atypia, small-medium cell size, regular cell shape, and rare mitotic figures. Histologically, PLGA is diverse, and a number of growth patterns have been described including solid, tubular, fascicular and papillary subtypes. The vast majority of patients have localized disease upon presentation, and distant metastases are extremely rare. Cervical metastases are unusual and may occur late—nearly a decade after initial presentation. Prognostic factors within PLAC are not well-defined. Perineural invasion does not appear to be a high-risk feature or prognostic of recurrence. Tumors less than or equal to 2 cm at presentation and hard palate primary site may be associated with better outcomes. Positive or unknown margin status has been associated with an increased risk of local recurrence. Anecdotal evidence suggests that patients with papillary subtype are more likely to develop cervical metastases. As with most cancers, recurrent tumors display a more aggressive phenotype with histologic features including more mitoses and, interestingly, increased papillary growth patterns.

Reports describing cervical metastases of PLAC in the existing literature are limited. Furthermore, risk factors for a poorer prognosis are not definitively known. There has been some suggestion in the literature that the papillary subtype may display a more aggressive clinical course. In this report, we present two cases of cervical metastases from PLGA one of which demonstrated papillary growth pattern.

**Patient 1**
A 37-year-old male presented to our institution with dysphagia and throat pain for 2.5 months. A large soft palate mass extending onto the tonsillar was visualized upon physical exam. There were no nodal or distant metastases upon presentation. He underwent a transhyoid neck dissection, mandibulotomy approach to the left soft palate, oropharyngeal reconstruction. Three years later, he recurred with a large level IB neck mass, with pathologic features consistent with his previous salivary malignancy. The recurrent lesion was treated with revision neck dissection and post-operative radiation therapy. Fifteen months post-operatively, he was without evidence of disease and doing well.

**Patient 2**
A 61-year-old male presented to our institution with dysphagia and unknown. A large soft palate mass extending onto the tonsillar was visualized upon physical exam. There were no nodal or distant metastases upon presentation. He underwent a transhyoid neck dissection, mandibulotomy approach to the left soft palate, oropharyngeal wall and lateral tongue base tumor. Reconstruction was performed with a radial forearm free flap. This final pathology of the primary site was reported as polymorphous low-grade adenocarcinoma well-to-moderately differentiated with papillary features and few calcifications (Figure 2). The neck dissection was free of nodal disease. His post-operative course was uncomplicated. He was decannulated and tolerated a regular diet. No adjuvant therapy was administered. Three years later he developed a neck mass (Figure 3). The recurrent lesion was treated with revision neck dissection and post-operative adjuvant radiation therapy. Final pathology demonstrated PLGA with a papillary growth pattern, and there were no other positive nodes. One year post-operatively from his recurrence, he was without evidence of disease.

**Discussion**
Cervical metastases of PLGA are considered to be a rare entity. Few cases exist in the literature, and the reported rates of regional spread vary greatly. The largest cohort of PLGA patients was recently described. Patel et al. utilized the Surveillance, Epidemiology and End Results (SEER) database to report 460 cases of PLGA and their clinical outcomes. Regional spread of disease was defined by the SEER Local/Regional/Distant Staging system as 25.1% in their population, significantly higher than prior reports.\(^2\)\(^3\)\(^4\) Excluding those with direct regional spread of the primary site, 5.0% of patients had N1 or N2 disease at the time of database inclusion.\(^5\) Other authors report approaches to the neck vary from elective neck dissection only in the case of planned major radiation therapy to routine excision of the primary site is usually sufficient for elective neck dissection only in the case of planned major reconstruction.\(^6\) The reported incidence of cervical metastases for all PLGA tumors is low, but the papillary subtype appears to be more aggressive and may merit some additional initial consideration of the neck.

**Conclusion**
Cervical metastases of PLGA are rare, and there is limited literature on the subject. Definitive risk factors for disease progression are unknown. Two cases with cervical metastases are reported here. Accumulating evidence suggests that the papillary subtype may display a distinct, and more aggressive, clinical course.