Polymorphous low-grade adenocarcinoma at the base of the tongue: An unusual location

Alfio J. Tincani, MD, PhD; Albina Altemani, MD, PhD; Antonio S. Martins, MD, PhD; Gilson Barreto, MD; João B. Valério, MD; André Del Negro, MD; Priscila P.C. Araújo, MD

Abstract
Polymorphous low-grade adenocarcinoma (PLGA) is a malignant neoplasm of low aggressiveness that occurs almost exclusively in the minor salivary glands, primarily those in the palate. We report a case of PLGA that arose in the base of the tongue and subsequently metastasized to the neck. The tumor was resected through the oral cavity with wide margins and dissection. The neck metastasis was treated with radical neck dissection and radiotherapy. The patient recovered and remained disease-free at follow-up 30 months later. This case shows that PLGA, which has a variable morphologic appearance, can occur at sites other than the salivary glands.

Introduction
Polymorphous low-grade adenocarcinoma (PLGA) is a malignant neoplasm that occurs almost exclusively in the minor salivary glands, especially those in the palate. This tumor is of particular interest because of its low degree of aggressiveness, its slow growth, and its conspicuous architectural polymorphism—features that for many years have complicated its recognition as a tumor type distinct from other adenocarcinomas.

In this article, we describe a case of PLGA that arose in an unusual location—the base of the tongue—and metastasized to the neck. Our review of the literature revealed that although this tumor is rare, it should nevertheless be distinguished from other neoplasms of the minor salivary glands.

Case report
A 69-year-old man presented with complaints of voice change and dysphagia that had developed within the previous year. He was a social drinker, but he was not a tobacco smoker.

On physical examination, an ulcerated 4 × 3-cm tumor was noticed in the tongue, close to the right side of the lingual V (figure 1). No cervical nodes suggestive of metastasis were found. Findings on radiography of the thorax and routine laboratory tests were normal. Magnetic resonance imaging (MRI) revealed that the tumor had invaded the superficial tongue muscles and had reduced the oropharyngeal lumen (figure 2). A biopsy yielded a diagnosis of epithelial neoplasm of the minor salivary glands. The small size of the biopsy fragment made it impossible to determine whether the tumor was a PLGA or a pleomorphic adenoma. The patient underwent tumor resection through the oral cavity.

Eighteen months postoperatively, a lymph node metastasis 2.5 cm in diameter at level II was diagnosed on the right side of the patient's neck. The patient was treated with radical neck dissection and postoperative radiotherapy. The patient did well during 30 months of follow-up, and no further treatment was necessary.

Discussion
The most common sites of tumors of the minor salivary glands are, in order, the palate, the oral mucosa, and the...
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retromolar fossa. This distribution probably reflects the higher number of glands in this area. Approximately 80% of minor salivary gland tumors are malignant; adenocarcinomas account for 25% of these malignancies.

The term polymorphous low-grade adenocarcinoma was first used in 1984 by Evans and Batsakis to describe a tumor of the salivary glands that had as its primary histologic characteristic a variety of architectural patterns associated with cytologic uniformity. PLGA had previously been referred to as terminal duct carcinoma in view of its probable origin in the ductal system of the salivary glands. Similar to terminal duct carcinoma, PLGA is formed by luminal epithelial, myoepithelial, and basal cells.

The most common clinical aspects of PLGA are a male preponderance (2:1), manifestation during the seventh decade of life, and an almost-exclusive occurrence in the oral cavity; the major salivary glands and the seromucosal glands of the nose and nasopharynx are rarely involved. To the best of our knowledge, only 5 cases of PLGA involving the base of the tongue have been previously reported (table). The most common symptom at this site is a painful mass that may hamper swallowing and may occasionally be associated with bleeding and discomfort. Otalgia and obstruction of the airways may also occur. The tumor has the potential to infiltrate bone. There are no known etiologic factors that predispose to PLGA.

Because of its morphologic pleomorphism, PLGA has often been wrongly diagnosed as pleomorphic adenoma or adenoid cystic carcinoma. However, PLGA differs from pleomorphic adenoma in that PLGA is characterized by infiltrative margins and an absence of myxochondroid stroma. The primary difference between PLGA and adenoid cystic carcinoma is cytologic. In PLGA, the cells are more basoid, with angled and hyperchromatic nuclei, and the cytoplasm is scarcer and paler. It is important to distinguish adenoid cystic carcinoma from PLGA because the former is associated with low long-term survival rates.

However, the differential diagnosis may be difficult to make when a biopsy sample is small, as occurred in our case. PLGA is a low-grade malignancy, and its biologic behavior is apparently not influenced by the different morphologic and cell differentiation patterns that it may exhibit. The only exception to this behavior is seen with tumors that have a predominantly papilliferous arrangement; these tumors are more aggressive and would be better classified as papillary cystadenocarcinomas.

The recommended treatment for PLGA is surgery with ample margins; radiotherapy may be used in cases of local recurrence and/or lymph node metastasis. In our case, we opted for resection through the oral cavity because of the superficial location of the tumor close to the lingual V. After 18 months of postoperative follow-up, the patient exhibited evidence of metastasis in the neck, and he was treated with radical neck dissection and postoperative radiotherapy. At the 30-month follow-up, he exhibited no evidence of recurrence.

We recommend that the possibility of PLGA be consid-

Figure 1. At presentation, the tumor (arrow) is seen at the base of the tongue.

Figure 2. Coronal MRI shows the tumor invasion (arrow) into the superficial tongue muscles.

Figure 3. Histology shows the combination of solid, ductal, and tubular growth (H&E, original magnification ×80).

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