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Case Report

# Polymorphous Low-Grade Adenocarcinoma: A Case Report

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## Abstract

Polymorphous low-grade adenocarcinoma (PLGA) is a malignant neoplasm featuring low aggressiveness and almost exclusively originated from minor salivary glands. A case of PLGA is reported in an 81-year-old female patient who attended the stomatology clinic reporting an increase in volume of palate. Intraoral examination revealed a tumor on the right posterior palate, hard in consistency and painless. Radiographic examination didn't reveal abnormalities. An incisional biopsy was performed with histopathologic diagnosis of salivary gland malignant neoplasm. Immunohistochemically the tumor cells were positive for EMA, CEA, p63, CK14, vimentin, S-100 protein and Ki-67 (< 1%), but negative for smooth muscle actin and GFAP. The patient underwent surgery with tumor-free margins, confirming PLGA. An obturator prosthesis was placed after surgery. PLGA is a slow-growing, distinct, uncommon neoplasm of the minor salivary glands. Because it is characterized by a diverse morphologic pattern that resembles adenoid cystic carcinoma or pleomorphic adenoma, the initial diagnosis is usually inconclusive.

**Keywords:** Malignant Neoplasm; Salivary Gland; Palate

## Introduction

The polymorphous low-grade adenocarcinoma (PLGA) is a kind of uncommon malignant tumor that occurs in the head and neck region, with predilection for smaller salivary glands. The PGLA represents from 10 to 15% of the intraoral salivary gland tumors, being responsible for 20 to 25% of malignant neoplasms, which occur on palate in 60% of the cases [1].

Clinical manifestations include a slow growing nodular lesion with 1 to 4 cm of diameter that has low aggressiveness. It can be asymptomatic, but it's usually accompanied by pain and lining mucosa erosion. Imaging exams reveal a moth-eaten bone aspect. It has a low metastatic potential, however, it can have relapses after long periods of time. It's more prevalent in females and age range between sixty and seventy years old, being more frequent on the palate [2].

The histopathological examination of the PLGA shows morphologic diversity and cytologic uniformity, in which can be found clear cells, mucosa cells and crystalloid bodies like the ones present in

pleomorphic adenomas, characterizing its difficult diagnosis. The main differential diagnosis include pleomorphic adenoma and cystic adenoid carcinoma and the distinction between the PLGA from other neoplasms is necessary once their prognosis and therapeutic conduct are different [3].

The initial treatment for PLGA consists of complete surgical excision, that can be combined with radiotherapy in severe cases to enhance the prognosis [3]. The purpose of this article is to report a case of polymorphous low-grade adenocarcinoma involving the palate, and to describe its clinical, radiography and histopathological features.

## Case Presentation

Female patient, 81 years old, was attended in stomatology clinic reporting a swelling on the palate.

Intraoral examination revealed a tumor on the right posterior palate, hard in consistency and painless (Figure 1). Panoramic and occlusal radiographs didn't reveal abnormalities (Figure 2).

**Figure 1:** Clinical features: Tumor presents as a painless mass of hard consistency and located on the right posterior palate.

**Figure 2:** Radiographic features: Panoramic radiograph showed no abnormalities.

Considering the diagnostic hypothesis of salivary gland neoplasm, an incisional biopsy was performed with histopathologic diagnosis of salivary gland malignant neoplasm (Figure 3A), suggesting PLGA or mucoepidermoid carcinoma.

Immunohistochemically the tumor cells were positive for EMA, CEA, p63, CK14, vimentin, S-100 protein, and Ki-67 (< 1%), but negative for smooth muscle actin and GFAP. Such additional report suggested PLGA or pleomorphic adenoma as the diagnostic hypothesis.

The patient underwent surgery with tumor-free margins, confirming PLGA (Figure 3B-3D). An obturator prosthesis was placed after surgery and the patient was referred to speech therapy.

**Figure 3:** Histopathologic features: A. Cribriform growth pattern; B. Solid growth pattern showing hyalinization of the stroma show; C. Cords and island of cuboidal cells and the stroma showing areas of mucinosis; D. Medium size and uniform cells and oval nuclei and nucleoli were observed.

## Discussion

The PLGA is described in the literature as a neoplasm of smaller salivary glands, affecting the soft and hard palates in 65% of the cases. Its incidence is more significant in elderly, predominating in female. The PLGA has an indolent, low growing behavior and low recurrence probability. It is a relatively recent neoplasm once it was described for the first time in 1983, establishing relevance to its diagnosis, behavior, clinical characteristics, and histopathological aspect [4,5].

It is described in this report a typical presentation of PLGA affecting a female patient, 81 years old, with a painless mass on posterior palate. These features corroborate bibliographical references. When investigating the case, an incisional biopsy was performed to clarify the differential diagnosis between PLGA, adenoid cystic carcinoma and pleomorphic adenoma, that share similar aspects [3].

Immunohistochemically, the tumor expressed a low rate of Ki-67 (< 1%), corroborating the studies that quoted an indolent growing for this kind of tumor. In the same way, in Regezi., *et al*. study, smooth muscle actin was negative, while the vimentin was positive in PLGA cases. With these results and with positivity for EMA, CEA, p63, CK14 and S-100 protein markers, the incisional biopsy suggested, as possible hypothesis, pleomorphic adenoma, and polymorphous low-grade adenocarcinoma [6,7].

Comparing PLGA with pleomorphic adenoma, the absence of tubules with two-layers of cells or squamous differentiation, cartilage lobules and negativity for GFAP on immunohistochemistry are parameters that support the diagnosis of PLGA. Besides, the pleomorphic adenoma does not present perineural nor stromal invasion, while both can be present in PLGA [8].

Usually, distinguishing PLGA from cystic adenoid carcinoma (CAC) histologically can be challenging, once both can present heterogeneous architecture with cribriform and tubular growth varying quantitatively, directing the diagnosis towards CAC when cribriform pattern is seen in the whole tumor. Furthermore, both present perineural invasion, which is more prominent in CAC and presenting itself as a targetoid appearance in PLGA's case. Cystic formations and calcifications also favor PLGA diagnosis [8].

Thereby, considering the exposed parameters, after the tumor removal, the diagnosis of polymorphous low-grade adenocarcinoma was established.

## Conclusion

The polymorphous low-grade adenocarcinoma is an uncommon salivary gland tumor that has a low growth. Once it presents a histological pattern that resembles the cystic adenoid carcinoma and the pleomorphic adenoma, the initial diagnosis is usually inconclusive. It is possible to elucidate the diagnosis by performing histopathological exam and immunohistochemical analysis, which, in this case, was a polymorphous low-grade adenocarcinoma.

## Bibliography

1. Fife TA., *et al*. "Polymorphous low-grade adenocarcinoma: a 17 patient case series". *American Journal of Otolaryngology* 34.5 (2013): 445-448.

2. Da Silva L., *et al*. "Adenocarcinoma Polimorfo de Baixo Grau". *Revista de Odontologia Contemporânea* 3.11 (2019): 16-21.

3. De Araujo VC., *et al*. "Polymorphous low-grade adenocarcinoma: an analysis of epidemiological studies and hints for pathologists". *Diagnostic Pathology* 8.6 (2013): 1-8.

4. Regezi JA., *et al*. "Oral Pathology: Clinical Pathologic Correlations". 6th edition. Missouri: Elsevier (2012).

5. Neville BW., *et al*. "Oral and Maxillofacial Pathology". 4th edition. Missouri: Elsevier (2016).

6. Beltran D., *et al*. "Selective immunohistochemical comparison of polymorphous low-grade adenocarcinoma and adenoid cystic carcinoma". *Journal of Oral and Maxillofacial Surgery* 64.3 (2006): 415-423.

7. Regezi JA and Batsakis JG. "Histogenesis of salivary gland neoplasms". *Otolaryngologic Clinics of North America* 10.2 (1977): 297-307.

8. Chatura KR. "Polymorphous low-grade adenocarcinoma". *Journal of Oral and Maxillofacial Pathology* 19.1 (2015): 77-82.