



POLYMORPHOUS LOW-GRADE ADENOCARCINOMA – A CASE REPORT

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ABSTRACT

Polymorphous low-grade adenocarcinoma (PLGA) is a slow growing malignant tumor of minor salivary glands and is generally of indolent nature. However, according to the most recent WHO Classification of Salivary Gland Tumors (2017), the cancer is classified as Polymorphous Adenocarcinoma (PAC). PAC presents as a less aggressive tumor, though it could on rare occasions demonstrate distant metastasis. PLGAs account for 10% of all tumors and 25% of all malignancies of the minor salivary glands. It has been frequently described as occurring in hard or soft palate minor salivary glands; some cases being described in the tongue and in major salivary glands. Here, we present a case of PLGA on palate in the left side in a 61-year-old male since past 20years.

KEYWORDS : Rare, Adenocarcinoma, Malignancy.

INTRODUCTION:

Salivary gland tumors are uncommon and constitute 2 to 6.5% of all head and neck neoplasms affecting predominantly major salivary glands, especially the parotids. (1) PLGA was firstly described as a distinct entity in 1983 and in 1984, Evans and Batsakis proposed the term 'polymorphous low-grade adenocarcinoma' (PLGA) to refer this tumor in the World Health Organization (WHO) classification of salivary neoplasms in 1990.

Due to its architecturally diverse structure, clinically indolent behaviour, and cytological uniformity, PAC is thought to be a unique entity.

Being asymptomatic with a low metastatic potential, it is commonly seen in elder women, with about 24% of primary tumors associated with major salivary glands (i.e., parotid, submandibular, or sublingual gland) and few in locations other than the head and neck, such as the breast and vagina. (2)

It is the second most common primary minor salivary gland malignancy after mucoepidermoid carcinoma, comprising 9 to 26.4% of all salivary malignancies.

We report a case of PLGA of the palate diagnosed based on histopathology and was treated by subtotal maxillectomy followed by reconstruction of the defect with an obturator.

Case Report:

A 61-year-old male patient presented with a chief complain of a growth present on palate in the left side since past 20 years. He complained of growth which was initially smaller in size and has attained its present-day size gradually.

There is a history of extraction of 15 about a year back.

On extra oral examination, no facial asymmetry or abnormality noted.



Figure 1: Patient Profile Picture

On intra oral examination, a well-defined swelling present in palatal region in the left side measuring 3cm*3.5cm extending antero-posteriorly from 0.5cm behind the incisive papilla up to 1 cm posterior to the line joining from maxillary tuberosity of both sides. It also extends mediolaterally crossing the mid-line from interdental region of 25 and 26 up to 1cm lateral to the midline. Superficial mucosa have normal appearance as that of the surrounding mucosa. On palpation, the growth is sessile, non-tender and firm in consistency attached to the underlying structure, doesn't bleed upon palpation.



Figure 2: Intraoral Findings



Figure 3: Radiographic Findings (cbct- Axial View)

The CBCT of maxilla revealed there is a soft tissue growth present on the palatal region without any invasion into the maxillary sinus or nasal cavity of the left side.

An intra oral incisional biopsy was performed. Microscopic examination of sections revealed myoepithelial component which appear spindled, clear to plasmacytoid cells seen scattered within the myxoid stroma.



Figure 4: Surgical Pictures

He underwent surgery to remove the tumor and excisional biopsy was done which upon microscopic examination revealed a partially circumscribed tumor in which cells display variable architectural patterns of different proportion including solid, tubular, reticular and single filling arrangements.

The defect was later reconstructed with an obturator. Patient is under periodic follow-up.

DISCUSSION:

Clinically, PAC presents as a firm, well-circumscribed, painless, and slow growing mass mostly covered by nonulcerated mucosa resembling a benign neoplasm and could be fixed to underlying structures, eroding and infiltrating the underlying bone, and is even present with perivascular and perineural invasion. (3) The myriad of histologic types of minor salivary gland tumors makes this the most heterogeneous group of neoplasms. PLGA occurs more frequently in females than in males with a male-to-female ratio ranging from 1:1.02 to 1:2.0.13 It affects mainly adult patients from the 3rd through the 7th decades, with a peak prevalence from the 5th to 6th decades of life. (4) It commonly arises in the palate (49-77.8%), followed by either the buccal mucosa or upper lip (7.4-13.4%) and can also involve the floor of the mouth, lower lip, alveolar ridge and tongue. Additionally, PLGA can arise in the lung, parotid gland, submandibular gland and maxilla, and two case reports describe it transforming into higher grade neoplasms. The presenting symptom in PLGA is a lump, may or may not associated with discomfort and/or pain. In our case, PLGA presented as an asymptomatic swelling on the left side of hard palate and

crossing midline without extending into the left maxillary sinus and nasal cavity. Histopathology of malignant minor salivary gland tumors is well-established important diagnostic aid. On gross examination, PLGA appears as a well-circumscribed, nonencapsulated, firm, homogeneous, tan mass. PLGA unified several histologic patterns: Solid, trabecular, glandular, cystic, ductal, spindled, fascicular, cribriform and papillary. Transoral open biopsy is not recommended prior to definitive treatment because of the risk of seeding the oral mucosa.

Because PLGA share many of the histologic characteristics of pleomorphic adenomas, monomorphic adenomas, adenoid cystic carcinomas (ACC), and low-grade papillary adenocarcinomas (LGPA), these tumors should be considered in the differential diagnosis of PLGA. Pleomorphic adenoma and monomorphic adenoma can be easily distinguished from PLGA by its lack of infiltration and by the presence of PLGA characteristics, such as poor circumscription, peripheral infiltration, and the tendency for perineural invasion. The need to distinguish PLGA from ACC arises more often as there are overlapping histologic features (i.e. cribriform, tubular and solid patterns), local infiltration and perineural invasion, but they can be distinguished from one another by cytology; ACC exhibit basaloid cells with scant cytoplasm. In addition, PLGA show spindling, and ACC do not. Finally, while PLGA stain widely with S-100 protein and EMA, ACC stain much less diffusely. (5)

Immunohistochemical staining can assist in making a diagnosis of PLGA, as 90% of cells stain positively with S-100 protein and epithelial membrane antigen (EMA). Muscle-specific actin and carcinoembryonic antigen are poor markers of PLGA because of their high variability in staining. PLGA may stain with glial fibrillary acidic protein (GFAP) but this is usually less consistent than GFAP staining of pleomorphic adenomas. Most PLGAs stain poorly with proliferation markers p53 and Ki-67, which supports the clinical observation that PLGA are slowly growing tumors. (6)

The treatment of choice for PLGA is wide local excision with clear margins. Postoperative radiation therapy is reserved for cases of positive or close surgical margins, but it has not been shown to alter outcome in patients without lymph node metastasis.

CONCLUSIONS:

Minor salivary gland tumors are relatively uncommon lesions encountered in the daily practice. PLGA is the second most common minor salivary gland malignancy, characterized by architectural diversity, cytological uniformity, and indolent clinical behavior. The understanding of the site, gender, age and prevalence, and histological typing aids in proper diagnosis and appropriate management of PLGA.

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