Polymorphous Low Grade Adenocarcinoma of the Hard Palate: A Case Report in a Male

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Abstract: Polymorphous low grade adenocarcinomas are indolent tumours that are localized preferentially to the palate and affect minor salivary exclusively. It was described in 1984 by Evans and Batsakis, when they reported a subset of heterogeneous groups of malignant adenocarcinomas arising from the palate and minor salivary glands and named them “polymorphous low grade adenocarcinomas”. An increasing number of reports in the pathology literature since then have identified this as the second or third most common minor salivary gland malignancy. The natural history of polymorphous low grade adenocarcinoma is distinct from other malignant salivary tumours. The most common presentation is that of a lump in the oral cavity, usually on the palate. Diagnosis is established after biopsy as cytological studies are not helpful. The morphological features can resemble pleomorphic adenoma and adenoid cystic carcinoma. We report a case of a middle-aged man that presented with a fungating mass in the hard palate.

Key words: Palate, minor salivary gland, polymorphous low grade adenocarcinoma.

1. Introduction

In 1984, Evans and Batsakis [1] reported a subset of malignant tumours arising from the hard palate, it was then heterogeneously composed group of adenocarcinomas arising in the minor salivary gland and they named them “polymorphous low grade adenocarcinomas” (PLGAs).

Almost at the same, Freedman and Lumerman [2] reported 12 cases of what they referred to as “lobular carcinomas” of the minor salivary glands. Although the latter term did not survive, both series describe PLGAs as an indolent tumour with local behaviour and having little tendency to metastasize.

Polymorphous low grade adenocarcinoma is a malignant neoplasm of salivary glands, more frequently detected in minor salivary glands.

It usually affects people in the age range between 30-70 years with predilection for female in the ratio 2:1 [3]. It is found almost exclusively in the minor salivary

gland and is rare in extra-oral location including major salivary glands. 60% of cases occur on the hard palate, 13% in the buccal mucosa, 10% in the upper lip, 6% in the retromolar and 9% in the rest of the oral cavity [3].

The lesion is described as a painless, slow growing mass, covered by non-ulcerated mucosa. In most cases, it may be adhered to deep planes and it can reach sizes between 1 and 4 cm [3]. Histology shows a non-encapsulated lesion with infiltrative margins. It is named polymorphous due to its different growth patterns; tubular, solid, papillary, microcytic, cribriform, fascicular and cords. It can infiltrate bone tissue and even perivascular and perineural invasion [4].

2. Case Report

A 55-year old male Fulani farmer presented with a painless ulcerated mass located in the hard palate and part of the left alveolar ridge, which caused an evident disfiguring of his face. The patient said he noticed the mass a year prior to presentation, and that, during this period it had gradually increased in size.

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He had history of foul smelling discharge from the mouth, dysphagia, odynophagia, bleeding, weight loss but no peripheral lymphadenopathy.

On examination, the lesion was firm, circumscribed measuring 3.4×2.8 cm and located in the hard palate. It was painless, not adhered to the deeper structures.

X-ray studies (A-P view of the skull x-ray) showed an irregular radiopaque lesion in the hard palate, but it did not indicate involving alveolar bone. There was no compromise of the ipsilateral maxillary sinus.

The lesion was excised and was sent for histopathological examination. On gross examination, it was an irregular firm to hard mass measuring $3.4 \times 2.8 \times 1.2$ cm and weighing 10 gram. Cut sections show a grey tumour measuring 2.2×1.4 cm in diameter. The margins were free of tumour.

Histological examination showed a tumour growing in solid nests, cords, tubular and cribriform patterns. It is composed of cells having moderately pleomorphic hyperchromatic nuclei and amphophilic cytoplasm (Fig. 1). The intervening stroma is desmoplastic containing tumour cells.

3. Discussion

The term polymorphous low grade adenocarcinoma was first used in 1984 by Evans and Batsakis [1] to describe a tumour of the salivary glands that had a variety of architectural patterns associated with cytologic

Fig. 1 High power view of tumor showing a solid and cribriform pattern with cell having a uniform round to ovoid nuclei with inconspicuous nucleoli and moderate amount of eosinophilic cytoplasm. (H & E stain $\times 100$).

uniformity as its primary histology characteristics.

This lesion is almost exclusively in minor salivary glands within the oral cavity, mostly hard palate, while its extra-oral presentation is extremely rare. Nasal fossa and nasopharynx locations have been described in less than 0.5%-1% of the cases [5].

This characteristic is also found in other salivary gland neoplasms whether it is benign or malignant. There is a female preponderance, particularly affecting women in their fifth and sixth decades. There have been several reports of this tumour in women by Abdul et al. [6] and Ramesh et al. [7], but our index patient in this report is a male Fulani farmer; other researchers like Gourin [8] and Tincani [9] have also reported this tumour in men.

PLGA described in this report corresponds to the experience described in other countries with high incidence in the palate and slow growth [5].

The histological findings of an invasive tumour growing in solid nests, lobular and cribriform patterns; with cells having moderately pleomorphic hyperchromatic nuclei and amphophilic cytoplasm, are similar to those described in the literature.

There is neither perineural, vascular nor lymph node metastasis in our patient as reported by Tanaka et al. [10] and Kurmar et al. [11]. 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 absence of myxochondroid stroma [11]. The primary difference between and adenoid cystic carcinoma is cytologic. In PLGA, the cells are more basaloid, with pleomorphic hyperchromatic nuclei, and pale eosinophilic cytoplasm [11].

It is important to distinguish adenoid cystic carcinoma from PLGA because the former is associated with low long-term survival rates [11]. However, the differential diagnosis may be difficult to make when a biopsy sample is small, as it is in our patient, except there is immunohistochemistry facility

which is lacking in our Centre as at the time of this report.

PLGA is a low grade malignancy, and its biologic behaviour is apparently not influenced by the different morphologic and cell differentiation patterns that it may exhibit [5].

The only exception to this behaviour is seen with tumours that have a predominantly papilliferous arrangement; these tumours 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 [12]. In our case, we opted for surgical resection through the oral cavity because of the superficial location of the tumour. However, since we lack radiotherapy facility, we planned sending the patient to another centre for additional radiotherapy treatment.

Polymorphous low grade adenocarcinoma is a rare malignant neoplasm with predilection for women. But this report has shown that it can be found in black man as it is reported among the Caucasians. Therefore, a high index of suspicion is required on the part of both clinicians and pathologists for this lesion of hard palate in men so as not to miss the diagnosis.

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