 DOI: 10.18231/2395-6194.2017.0026

Polymorphous low grade adenocarcinoma–A diagnostic challenge

N. Balan[1], Edwina J[2,*], Sunita Amruthesh[3], Sudhaamani[4], Yasmeen Ahamed[5]

[1]HOD, [2]PG Student, [3]Professor, [4,5]Reader, Dept. of Oral Medicine & Radiology, Vivekanandha Dental College for Women, Tamil Nadu

***Corresponding Author:**
Email: edwinajoshua@yahoo.com

Abstract

PLGA is considered as a tumour with low neoplastic behavior and varied cytological architecture. PLGA amounts to 12.5% of all salivary gland tumours and 22% of those that are malignant. The accurate diagnosis is challenging due to its diverse clinical and histopathological presentation, but it's necessary for accurate management. In this case report, polymorphous low grade adenocarcinoma of the upper labial mucosa has been presented.

Keywords: PLGA, Minor salivary gland tumour, Malignant tumour.

Introduction

The oral cavity consists of major and minor salivary glands which secrete saliva into the oral cavity. These salivary glands are tubuloacinar in structure.[1,2] Pathological processes like tumours may originate from the major or minor glands. The incidence of these tumours ranges from about 1.0 to 6.5 cases per 100,000 people, 54% to 79% of which are benign and the rest may be malignant. The palate is the most common site for malignant salivary gland tumours.[3] Polymorphous low-grade adenocarcinoma (PLGA) is considered a malignant salivary gland tumor of low neoplastic activity, commonly involving the minor salivary glands.

Case Report

A male patient, who was 32 years old presented to the department of oral medicine and radiology with a painful swelling on the inner side of the right upper lip region since the past 8 months. On eliciting the history, the swelling was initially of smaller size and gradually increased in size to the current state and was almost constant in size for past 4 months. It was associated with chronic pain which presented during manipulation of the swelling for the past 4 months. The patient did not have a history of previous trauma or surgery to the area. The patient had previously undergone consultation from a dentist who prescribed an anaesthetic gel which did not subside the symptoms. His medical history was not significant. Patient previously had the habit of smoking for 10 years. He had quit the habit since the past 2 years.

Physical examination revealed a solitary swelling which was present on the maxillary labial mucosa on the right side. It was pink in colour, irregular in shape, of size 1cm x 1.5cm, the surface was lobulated seen extending, supero-inferiorly, 1cm from the labial vestibule and 1cm from the vermillion border of lips; medio-laterally, 0.5cm from the labial frenum to 2cm from the labial commissure. The borders were ill defined, the base appeared sessile and no secondary changes could be seen. On palpation, the swelling was firm in consistency, tender with indurated base and was non reducible, non-fluctuant and non-compressible. The submandibular, cervical or preauricular lymph nodes were non-palpable.

Provisionally, the diagnosis of benign tumour of labial minor salivary gland was given. The differential diagnosis that were considered were firstly, fibroma of the upper lip. This was due to the fact that the lesion was firm in consistency. Inflammatory hyperplasia of the fibroblasts can commonly occur in the oral cavity. Secondly, fibrosed lipoma was given as adipocytes are present in the submucosa and long standing lipoma, thirdly, neurofibroma was considered, the last differential diagnosis was fibrosed mucocele.

Routine blood investigations were carried out and excisional biopsy with 2 mm margin was done under local anaesthesia.

On histopathology the gross specimen measured 2.5 cm x 2 cm in size, roughly ellipsoidal in shape and brownish white in colour. H&E stained section shows monomorphous epithelial cells arranged in tubular and cribriform pattern. The tubular pattern which show tumor epithelial cells which are composed of cuboidal cells with round to ovoid vesicular nuclei arranged around the duct like structure. Few epithelial islands consists of cystic spaces with basophilic, eosinophilic and mucoid material forming a cribriform pattern. These tumor epithelial nests in few areas encircle nerve bundles, suggestive of neurotropism. The surrounding stroma is myxoid and fibrous with few inflammatory cells. Residual salivary gland is also evident along the periphery. The histopathology diagnosis was confirmed as **"Polymorphous low grade adenocarcinoma".**

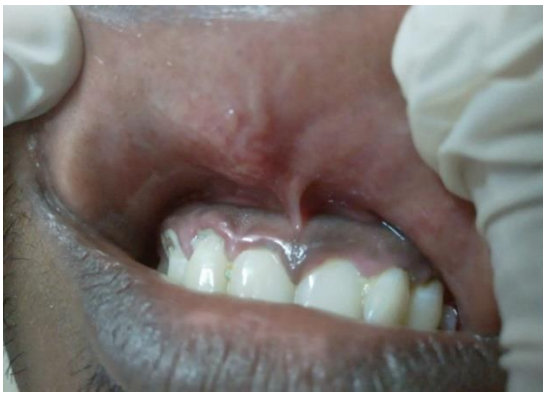

Fig. 1

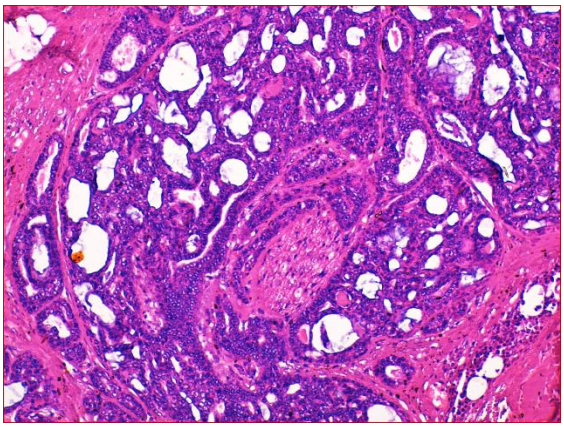

Fig. 2

Discussion

In 1983 Batsakis et al described an intraoral minor salivary gland neoplasm and called it as terminal duct carcinoma as he thought it arised from the intercalated or terminal ducts present in the salivary glands. [4] PLGA was later described using the many terminologies like the lobular carcinoma due to its similarity to lobular carcinoma of breasts.[5] The term polymorphous low grade adenocarcinoma was later proposed due to its diverse morphological and varied microscopical architectures.[5]

In a study of 26,960 cases of salivary gland tumours by De Araujo et al., it was reported that only 431 (1.6%) were PLGAs.[6] Now PLGA accounts for only 12.5% of salivary gland and about 22% of all malignant salivary gland tumours.[7] In a study of 164 cases of PLGA, the female: male ratio was 2:1. Their average age of occurrence was 57.6. The tumour occurs in the palate, the lip, the buccal mucosa, the alveolar ridge followed by other mucosal sites in decreasing order of frequency.[8]

This tumour commonly presents as an asymptomatic mass that slowly increases in size and is painless but may be occasionally associated with pain and secondary changes. The symptoms may present from few days to few years. Clinically and pathologically PLGA closely mimics adenoid cystic carcinoma, which also has slow rate of growth and variable growth patterns with infiltrative borders.[10] Pathologically, PLGA has wide diversity in its architectural patterns but uniformity in cytological patterns can also be seen. A classical tumor can be seen as a well circumscribed mass macroscopically, but microscopically, it presents as a non-encapsulated tumour with infiltrative borders.[10] Several architectural patterns which include solid, trabecular, fascicular, cribriform, tubular, and papillary types can be seen. One may find multiple patterns which appear in a single tumor.[9] The presence of mitotic figures is rare.[10] Occasionally, necrosis may also be present. The cells may be have open ovoid nuclei and small nucleoli. Individual cells may be centered on a nidus, resulting in a target-like appearance. The stroma may frequently shows mucoid changes and has a blue-gray hue.[10]

PLGA in the palate mimics adenoid cystic carcinoma(ACC) and pleomorphic adenoma. Differentiation between PLGA and ACC is based on cytologic and architectural features. The cells of ACC are small and have small hyperchromatic nuclei. The cribriform pattern of ACC is more rigid than that of PLGA. In addition, PLGA tubules are typically lined by monolayered tumor cells, but adenoid cystic carcinoma tubules are lined by bilayered tumor cells. These subtle differences may help in the accurate diagnosis of PLGA.

PLGA and ACC may show recurrence locally, however, ACC is considered to be more aggressive. It also has a greater potential for metastasis. The surgical approach in the treatment of ACC is more radical and often combined with adjuvant radiotherapy.[10] Other tumours which may be confused with PLGA include pleomorphic adenoma, salivary duct adenocarcinoma and epithelial-myoepithelial carcinoma.[11]

PLGA which primarily consists of luminal and non-luminal cells may differentiate into various morphological patterns. The tubules and duct like structures arise from luminal cells. The non-luminal cells transform into solid nests and cribriform patterns with pseudoluminal spaces. Myoepithelial differentiation can also be seen in certain cases.[10] PLGA cases are generally treated with wide local excision. The prognosis of PLGA is good, with a recurrence rate of approximately 25%, and cervical lymph node metastases in 10%.[11]

Collagen IV and laminin, and their integrin ligands, are very useful in demonstrating that neoplastic ductal units of PLGA which are composed of a single cell layer, being distinct from ACC which contains structures composed of two layers of neoplastic cells.[12]

Conclusion

PLGA can show various diagnostic challenges. It is necessary to accurately diagnose PLGA for adequate treatment and management. There is relatively good prognosis. Recurrence rate is about 9% to 17% which can be controlled with re-excision. Death from PLGA is

extremely rare but may occur secondary to direct extension into vital structures.

References

1. Susan Standring. Gray's Anatomy: The Anatomical Basis of Clinical Practice. 39th Edition. Churchill Livingstone. Elsevier. p.603-4.
2. Antonio Nanci. Ten Cate's Oral Histology: Development, Structure and Function. 8th Edition. Mosby. Elsevier.p.253-70.
3. Neville, Damm, Allen, Bouquot. Oral and Maxillofacial Pathology. 3rd Edition. Saunders. Elsevier.p.473-98.
4. Batsakis JG, Pinkston GR, Luna MA, Byers RM, Scuibba JJ, Tillery GW. Adenocarcinoma of the oral cavity: A clinicopathologic study of terminal duct carcinomas. J Laryngol Otol. 1983;97:825–35.
5. Evans H, Batsakis JG. Polymorphous low-grade adenocarcinoma of minor salivary glands. A study of 14 cases of a distinctive neoplasm. Cancer 1984;53:935–942.
6. de Araujo VC, Passador Santos F, Turssi C, Soares AB, de Araujo NS. Polymorphous low-grade adenocarcinoma: An analysis of epidemiological studies and hints for pathologists. Diagn Pathol 2013;8:6.
7. Ellis GL, Auclair PL. Atlas of tumor pathology, third series, fascicle 17. Tumors of the salivary glands. Washington DC: AFIP, 1996;216–228.
8. Castle JT, Thompson LD, Frommelt RA, et al. Polymorphous low grade adenocarcinoma: a clinicopathologic study of 164 cases. Cancer 1999;86:207–219.
9. Pogodzinski MS, Sabri AN, Lewis JE, Olsen KD. Retrospective Study and Review of Polymorphous Low-Grade Adenocarcinoma. Laryngoscope 2006;116:2145-49.
10. Ordonez PB, Linkov I, Huvos AG. Polymorphous low grade adenocarcinoma of minor salivary glands: a study of 17 cases with emphasis on cell differentiation. Histopathology 1998;32:521-29.
11. Simpson RHW, Clarke TJ, Sarsfield PTL, Gluckmant PGC, Baba Jew AV. Polymorphous Low-Grade Adenocarcinoma Of The Salivary Glands: A Clinicopathological Comparison With Adenoid Cystic Carcinoma. Histopathology 1991;19:121-129.
12. Loducca SVL, Raitz R, AraujÂjo NS & AraujÂjo VC. Polymorphous low-grade adenocarcinoma and adenoid cystic carcinoma: distinct architectural composition revealed by collagen IV, laminin and their integrin ligands (a2b1 and a3b1). Histopathology 2000;37:118-123.