

# An Unusual Presentation of Pleomorphic Adenoma- A Case Report

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**Abstract:** Salivary gland swellings can result from tumors, an inflammatory process or cysts. It can sometimes be difficult to establish; whether pathology arises from the salivary gland itself or adjacent structures. Neoplasms of the salivary glands account for less than 1% of all tumors, 3-5% of all head and neck tumors and benign pleomorphic adenoma (PA) of minor salivary glands arising de novo is very rare. PA is the most common tumor of the salivary gland. While the majority arises from the parotid gland, only a small percentage arises from the buccal minor salivary gland. A case of PA of minor salivary glands in the buccal mucosa in a 59-year-old male is discussed. It includes review of literature, clinical features, histopathology, radiological findings and treatment of the tumor; with emphasis on diagnosis.

**Keywords:** Buccal, minor salivary gland, pleomorphic adenoma

## 1. Introduction

Pleomorphic adenoma (PA) is a benign, mixed tumor which most commonly involves the parotid gland. Approximately, 8% of PA involves the minor salivary glands and the palate is the most common site (60-65%) (1). PAs are known to occur in other minor salivary gland sites, including the lip, buccal mucosa and tongue (2). PA of a buccal minor salivary gland, which lies on the external aspect of buccinators, has not been reported previously. We report a case of a PA apparently arising from such a gland and relevant review of literature. An extensive research has revealed only few well-documented cases of PA of a buccal minor salivary gland. This article is presented to share our experience with a case of very rare PA of a buccal minor salivary gland.

## 2. Case Report

A 59-year-old male patient presented with the chief complaint of painless swelling over right side of face since 2 months. The swelling was initially smaller in size; gradually increased to the present size. Patient had no significant medical history. No history of chronic alcoholism or tobacco was given by the patient. There was no previous history of any similar swelling in the same region. On general examination, the patient was apparently healthy. There was no regionally lymphadenopathy. Extraoral examination revealed facial asymmetry due to a swelling on the right side of face [figure 1]. A solitary dome-shaped swelling with diffused margins was present on right midface region, of size approximately 3.5cm × 3 cm, extending antero-posteriorly 2 cm posterior to right ala of nose to 4.5cm anterior to the tragus of right ear. Superior-inferiorly extending from ala-tragus line to the body of mandible [figure 1]. No abnormality was detected on the overlying skin. No local rise in temperature over the swelling noted. On palpation the swelling was firm in consistency, non-tender, non-fluctuant, non-reducible, non-pulsatile, mobile and has well-defined margins. Mouth opening was adequate.

Intra oral examinations the oral hygiene was average. On right buccal mucosa showed a single oval bluish black swelling of size approximately 3 cm x 2.5 cm with ill-defined margins. No visible discharge, sinus opening or pulsations present [figure 2]. On palpation, the swelling was non-tender, firm in consistency, well defined margins and mobile. Additional findings noted were swelling was non-fluctuant, non-reducible, non-compressible and non-pulsatile. There was no evidence of nasal obstruction or ophthalmologic signs of extension of the lesions into these anatomical regions. Magnetic Resonance Imaging (MRI) of face was suggestive of 3.5cm x 3.5cm x 3cm well defined round cystic lesion seen in subcutaneous plane showing T1 hyperintense signals suggestive of protein content or haemorrhage. Polypoid soft tissue component also seen within it [figure 5 & 6]. No extension to oral cavity / paranasal sinus. Ultrasonography (USG) of the swelling was carried out a mixed echoic lesion of size approximately 3.4 cm x 2.4cm with well-defined margins noted in the right midface region. Posterior acoustic enhancement and peripheral vascularity noted [figure 3]. Based on the clinical and imaging findings a differential diagnosis of benign connective tissue neoplasm, benign salivary gland neoplasm and dermoid cyst was made. Fine-needle aspiration cytology (FNAC) from the lesion was carried out which clusters of basaloid cells in a background of blood [figure 4]. Both acute and chronic inflammatory cells can be seen in an eosinophilic background. Surgical excision of tumor was performed. Excisional biopsy specimen revealed a single soft tissue specimen, approximately 2.5 cm × 2 cm in size, greyish pink in color, round in shape and was soft to firm in consistency with rough surface texture. [figure 8]. Scattered areas of hemorrhages and necrosis were also noted [Figure 8]. H & E section showed many duct like structures lined by cuboidal cells, cystic spaces filled with eosinophilic coagulum, myoepithelial cells, solid sheets of round to ovoid cells with basophilic nucleus and scanty cytoplasm. Intervening stroma is hyalinised at areas with many endothelium lined vascular spaces filled with RBC [Figure 7]. Histologically, the features were consistent with

Pleomorphic Adenoma. Although the patient was treated surgically with wide margins of resection and is doing well presently, but knowing the notorious nature of minor salivary gland neoplasms the patient has been kept under a close long-term follow-up.



Figure 1 Figure 2

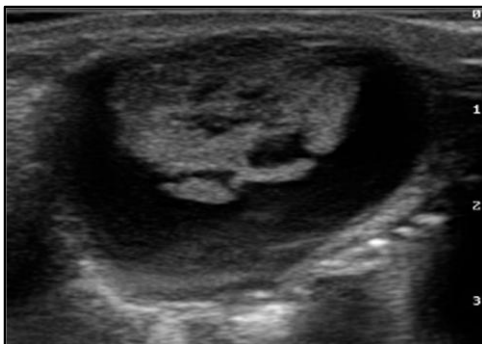


Figure 3



Figure 4

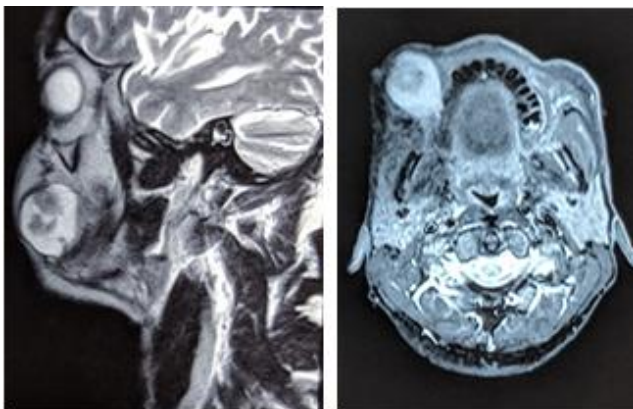


Figure 5 Figure 6

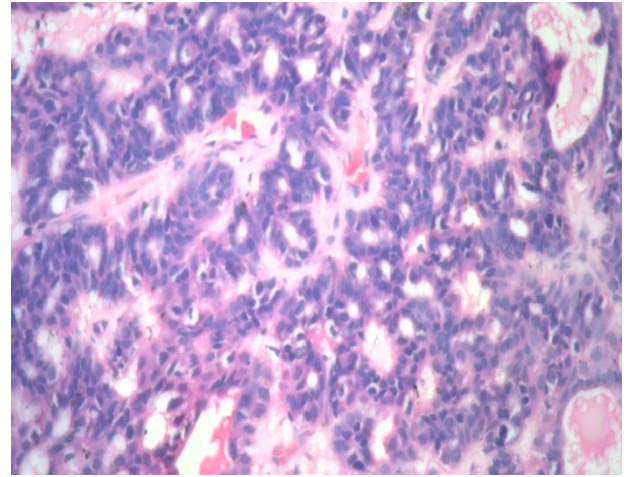


Figure 7

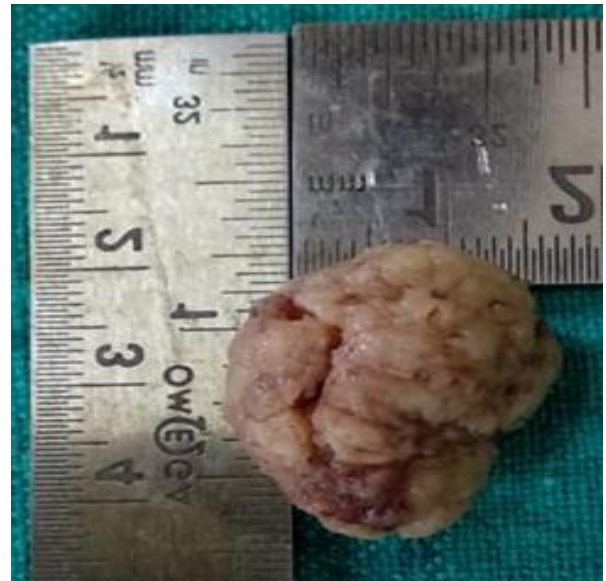


Figure 8

### 3. Discussion

Tumors of the salivary glands represent less than 5% of all head and neck tumors and two-thirds of these tumors are PAs (1). PA is the most common salivary gland tumor that affects both major and minor salivary glands. The parotid gland is the most common site of PA. In the parotid gland, this tumor most often presents in the lower pole of the superficial lobe, about 10% of the tumors arises in the deeper portions of the gland. PA is seen in approximately 8% of the minor salivary glands (2). Most salivary gland tumors spread by local infiltration, perineural or hematogenous spread and less commonly, via lymphatic. Rarely, metastases from other malignancies may involve the parotid glands. The cause of salivary gland tumors remains obscure, but ionizing radiation has been identified as a risk factor. PAs are known to occur in other minor salivary gland sites, including the lip, buccal mucosa and tongue (3). There are 800-1, 000 minor salivary glands located throughout the oral cavity in the tissue of the buccal, labial and lingual mucosa; the soft palate; the lateral parts of the hard palate; and the floor of the mouth. Unlike the major glands, they are not encapsulated by connective tissue, only surrounded by it and usually have a number of acini connected in a tiny lobule (4). The glandular lobules are 1-5 mm in diameter



and are separated by thin connective tissue (5). A minor salivary gland may have a common excretory duct with another gland, or may have its own excretory duct. Their secretion is mainly mucous in nature (except for Von Ebner glands) (4). The patient usually comes with the chief complaint of a small, painless, quiescent nodule which slowly begins to increase in size, sometimes showing intermittent growth. The skin rarely ulcerates even though these tumors may reach a very large size. Pain is not a common symptom, but local discomfort is frequently present. Facial nerve involvement manifested by facial paralysis is rare (2). PAs of the minor salivary glands usually present as painless, submucosal swellings with size ranging from 2 to 6 cm in greatest diameter, but some tumors are massive (6). Grossly, they are usually encapsulated, solitary, well-defined, ovoid or round masses. Larger neoplasms may have a characteristic bosselated surface with necrotic or cystic regions. Their consistency varies from hard to rubbery to soft swelling that may be fluctuant. The cut surface of the tumor is characteristically solid and the color varies from gray blue, pale yellow to tan. There may be gritty areas and gelatinous or glistening foci may be present when there is cartilaginous or myxochondroid differentiation. Willis described PA as the lesion with unusual histologic pattern consisting of cells exhibiting the ability to differentiate to epithelial (ductal and nonductal) cells and mesenchymal (chondroid, myxoid and osseous) cells. It demonstrates combinations of glandular epithelium and mesenchyme-like tissue and the proportion of each component varies widely among individual tumors. Foote and Frazell (1954) categorized the tumor into the following types: Principally myxoid, myxoid and cellular components present in equal proportions, predominantly cellular and extremely cellular. The epithelial components form ducts and small cysts that may contain an eosinophilic coagulum, the epithelium may also occur as small cellular nests, sheets of cells anatomizing cords and foci of keratinizing squamous or spindle cells. Myoepithelial cells have variable morphology, sometimes appearing as angular or spindled, rounded with eccentric nuclei and hyalinized eosinophilic cytoplasm resembling plasma cells. Myoepithelial cells are also responsible for the characteristic mesenchyme-like changes, giving a myxoid appearance. Vacuolar degeneration of the myoepithelial cells result in a cartilaginous appearance. Foci of hyalinization, bone and even fat can be noted in the connective tissue stroma of many tumors (2). FNA biopsy, operated in experienced hands, can determine whether the tumor is malignant in nature with sensitivity of around 90% (4). The differential diagnosis of PA cheek includes buccal abscess, dermoid cyst, sebaceous cyst, neurofibromas, lipoma, mucoepidermoid carcinoma and polymorphous low-grade adenocarcinoma (7). The buccal space abscess shows signs of inflammation, which were absent in present case. The solid nature of PA and lack of tissue showing the three germ layers rule out the possibility of mature dermoid cyst. Sebaceous cyst shows punctum and fixed mass, which differentiate it from PA. As on histological picture, both epithelial and myoepithelial cells were seen; which ruled out mucoepidermoid carcinoma. The negative slip test clinically and absence of lipomatous component histologically rules out lipoma. The absence of perineural invasion and mitotic

figures obscure the chances of polymorphic low-grade adenocarcinoma.

Treatment of pleomorphic adenoma includes simple excision, excision with a margin, and use of adjuvant radiotherapy. Many pleomorphic adenomas of minor salivary glands will have a capsule that is either thin or incomplete. (8, 9) Though these benign tumors are encapsulated, they require resection with an adequate margin of normal surrounding tissue because these tumors are known to have microscopic pseudopod-like extensions into the surrounding tissue due to dehiscence in the false capsule. Hence, incisional biopsy is avoided in these tumors to avoid spillage of the tumor cells. The recurrence of this tumor is a well-known factor. Lou *et al.* reported a recurrence of 7% in 1342 patients with benign parotid neoplasms and 6% in benign minor salivary gland tumors. Recurrences are reported even at 18 years after initial treatment. (9, 10) Inadequate initial surgical procedure was reported to be the main cause of failure. Pseudopodia, capsular penetration, and tumor rupture with spillage of tumor cells are the most frequently encountered surgical issues. (8) If we consider its malignant potential, in a series of 737 minor salivary gland tumors, Wang *et al.* found 53.9% to be malignant. Carcinoma arising from pleomorphic adenoma accounts for about 3% of salivary tumors. (11) In recent years, immunohistochemical markers are playing a major role in distinguishing pleomorphic adenomas from malignant salivary gland tumors. Human epidermal growth factor receptor-2/neu expression, androgen receptor expression, overexpression of P53, and expression of Ki-67 can be used in evaluating malignant salivary gland tumors, but these markers may also be expressed in benign pleomorphic adenomas. Hence, in the absence of compelling histologic features of malignancy, expression of these immunohistochemical markers should not be interpreted as evidence of carcinomatous transformation in a pleomorphic adenoma. (12)

In our case complete removal of the lesion was possible and the salivary gland was spared. This is in agreement with Leverstein *et al.*, 1997 who stated that surgery of PA must be highly customized on the basis of histologic type, extension and patient's age (13).

#### 4. Conclusion

The salivary glands may show a diverse range of lesions presenting a challenge to even the most experienced clinician and pathologist. PA of minor salivary gland is a tumor of rare occurrence and a diagnosis should be made carefully lest a major salivary gland be resected. A point to be noted here is that the ear lobule was not raised, thus clinically indicating that the swelling may not be of parotid gland origin. The swelling was painless, slow growing, mobile with well-defined borders. Histopathologically, plenty of myoepithelial cells and strands of epithelium in myxoid, stroma were evident throughout tumor confirming diagnosis of PA. The complete surgical excision with surrounding dispensable normal tissues is the key to successful treatment of such tumors. Recurrence even after many years of surgical excision as well as malignant

transformation should be a concern, and, therefore, long-term follow-up of these cases is advisable.

## References

- [1] Greenberg M, Glick M. *Burket's Oral Medicine*. Greenberg M, Glick M. Philadelphia: Lippincott; 2008. 172 p.
- [2] Rajendran R, Rajendran R, Sivapathasundaram B. *Shafer's Text Book of Oral Pathology*. 5th ed. New Delhi: Elsevier; 2006. 311-6 p.
- [3] *Textbook and Color Atlas of Salivary Gland Pathology: Diagnosis and Management* | Wiley [Internet]. Wiley.com. [cited 2020 Feb 11]. Available from: <https://www.wiley.com/en-us/Textbook+and+Color+Atlas+of+Salivary+Gland+Pathology%3A+Diagnosis+and+Management+-p-9780813802626>
- [4] *Ten Cate's Oral Histology-8th Edition* [Internet]. [cited 2020 Feb 11]. Available from: <https://www.elsevier.com/books/ten-cates-oral-histology/nanci/978-0-323-07846-7>
- [5] Klijanienko J, Vielh P, Batsakis JG. *Salivary Gland Tumours*. 1st ed. Vol. 15. New Delhi: Karger Publishers; 2000. 10 p.
- [6] Buenting JE, Smith TL, Holmes DK. Giant pleomorphic adenoma of the parotid gland: case report and review of the literature. *Ear Nose Throat J*. 1998 Aug; 77 (8): 634, 637-8, 640.
- [7] Jorge J, Pires FR, Alves FA, Perez DEC, Kowalski LP, Lopes MA, et al. Juvenile intraoral pleomorphic adenoma: report of five cases and review of the literature. *Int J Oral Maxillofac Surg*. 2002 Jun 1; 31 (3): 273-5.
- [8] Mubeen K, Vijayalakshmi KR, Pati AR, Giraddi GB, Singh C. Benign pleomorphic adenoma of minor salivary gland of palate. *J Dent Oral Hyg*. 2011; 3: 82-8.
- [9] Lou SM, Rich AM, De Silva RK, Ferguson MM. Pleomorphic adenoma of minor salivary gland. *Oral Oncol Extra*. 2006; 42: 170-2.
- [10] Varghese BT, Sebastian P, Abraham EK, Mathews A. Pleomorphic adenoma of minor salivary gland in the parapharyngeal space. *World J Surg Oncol*. 2003 Feb 25; 1: 2.
- [11] Wang Y-L, Chang H-H, Chang JY-F, Huang G-F, Guo M-K. Retrospective survey of biopsied oral lesions in pediatric patients. *J Formos Med Assoc Taiwan Yi Zhi*. 2009 Nov; 108 (11): 862-71.
- [12] DeRoche TC, Hoschar AP, Hunt JL. Immunohistochemical Evaluation of Androgen Receptor, HER-2/neu, and p53 in Benign Pleomorphic Adenomas. *Arch Pathol Lab Med*. 2008 Dec 1; 132 (12): 1907-11.
- [13] Leverstein H, Tiwari RM, Snow GB, van der Wal JE, van der Waal I. The surgical management of recurrent or residual pleomorphic adenomas of the parotid gland. Analysis and results in 40 patients. *Eur Arch Otorhinolaryngol*. 1997 Aug 1; 254 (7): 313-7.