

Papillary cystadenocarcinoma – A Very Rare Case Report

Dr.Sherin.A.Khalam¹, Dr.Ninan Thomas², Dr. Anroop A³

ABSTRACT

Papillary cystadenocarcinoma is an extremely rare malignant neoplasm characterized by cysts and papillary endophytic projections¹. It was first defined in 1991 by World Health organization as a separate entity^{1,2}. Major locations of this neoplasm are the parotid gland, the sublingual gland, and minor salivary glands, while occurrence in the submandibular gland is extremely rare³. We present a case of papillary cystadenocarcinoma arising from the submandibular gland in a 67-year-old male patient. Further, we have discussed the cytological and histopathological features of this rare entity and reviewed the current literature.

Key words: Access osteotomy, Excision Biopsy, Fine needle aspiration Cytology, Papillary cystadenocarcinoma, Submandibular Salivary Gland

¹Sr.Lecturer, Oral & Maxillofacial Surgery, PMS College of Dental Science & Research, Vattappara, Trivandrum, Kerala, India.

²Sr.Lecturer, Oral & Maxillofacial Surgery, Mar Baselios Dental College, Thankalam, Kothamangalam, Ernakulam, India.

³Sr.Lecturer, Oral & Maxillofacial Surgery, KMCT Dental College, Mukkam, Kozhikode, Kerala, India.

Corresponding author mail: drsherin666@gmail.com

Conflict of Interest: Nil

INTRODUCTION

Papillary cystadenocarcinoma of the salivary gland is a rare malignant neoplasm. Major locations of this neoplasm are the parotid gland, the sublingual gland, and minor salivary glands, while occurrence in the

submandibular gland is extremely rare. We present a rare case arising in the submandibular salivary gland of a 60-year-old male patient which was histopathologically confirmed where access osteotomy was used to do the excision biopsy.

CASE REPORT

A 60 year old patient reported with pain and swelling in relation to lower right back region of jaw with pus discharge since 15 days (Figure 1).

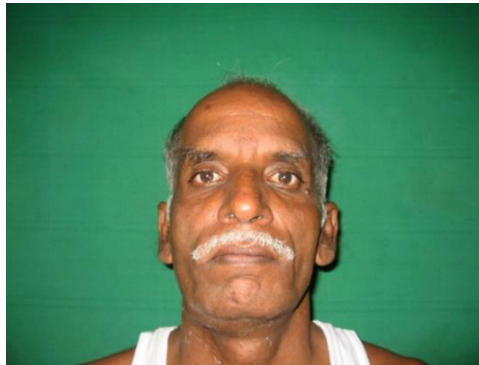


Figure 1: **Preoperative Profile**

Facial asymmetry present, Extra-oral draining sinus present in right side of face in sub-mandibular region with a bulbous tissue overgrowth measuring approximately 1x2 cm ; pus discharge present associated with pain and tenderness that radiates across right pre-auricular region ,right cheek and right sub-mandibular region if pus accumulates(Figure 2).



Figure 2: **Sinus Opening**

Right sub-mandibular region indurated ; no palpable lymph nodes. FNAC was done which reported “chronic sialadenitis”. Orthopantomogram and CT scan was taken which revealed Enlarged and swollen right sub-mandibular gland with calcification/

inspired secretions and thickening of the adjacent buccal mucosa distorting the Oropharyngeal airway and obscuring adjacent Fat planes, no bone involvement is detected. Right sub-mandibular gland inflammation – sialectasis ; normal

mandible with no evidence of infective/
neoplastic process or Lytic/ destructive

Lesion was reported(Figure 3 & 4).

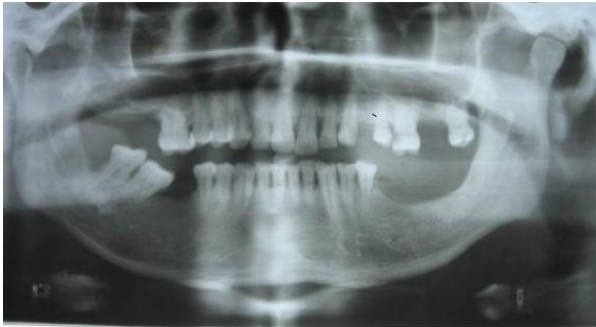


Figure 3: **Pre Operative OPG**

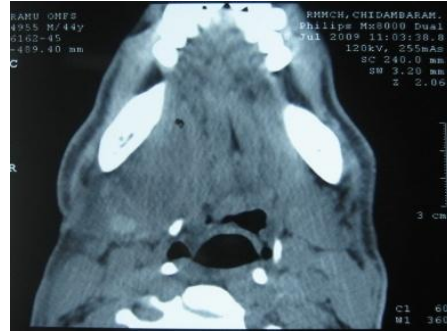


Figure 4: **Preoperative CT**

Under oral intubation, GA was administered Supra hyoid neck dissection – an incision placed, layer by layer dissection done (Figure 5).



Figure 5: **Incision Marking**

Sub-mandibular gland and its content exposed. the facial artery and vein exposed, identified and ligated, The lesion along with sub-mandibular gland ,node removed in Toto (cervically visible part) As the posterior extent of the Lesion was firm and adherent towards the medial aspect of the mandible on right side in relation to medial pterygoid muscle (Figure 6).

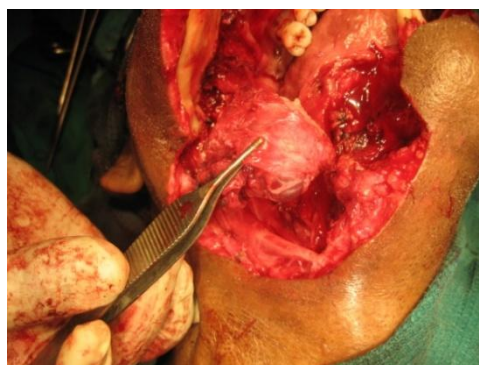


Figure 6: Exposure of Lesion

Modified McGregor lip split incision placed in right side of mandible, Lip was split in the midline Midsymphyseal access Osteotomy was performed in relation to 31 and 41 , the mandible was swung from its position to the right side (Fig 7).



Figure 7: Access Osteotomy

The Lesion attached to the medial pterygoid muscle on right side along with sublingual gland exposed and removed In Toto(Fig 8).



Figure 8: Excised Mass

The wound margins and the site were meticulously Debrided with saline and Metrogl. Replating done using 4-hole with Gap plate (2mm) and 4 (2x10mm) screws(Fig 9).



Figure 9: Replating

Layer by Layer closure done using 2-0 vicryl, 3-0 proline suture material. Tube drain was placed and secured with 3-0 silk suture material(Fig 10).



Figure 10: **Immediate Closure**

Blood transfusion started intra-operatively and patient was extubated uneventfully. Post Operative Follow up was done . The patient was discharged after 10 days, the post operative occlusion was satisfactory(Fig 11 & 12).

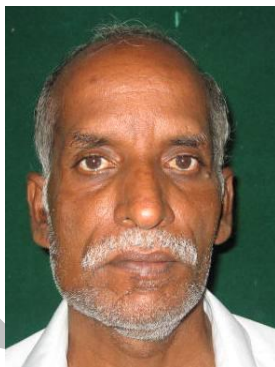


Figure 11: **Post operative Profile**



Figure 12: **Post Operative Occlusion**

Histo pathological Findings

The tumour consists of unilocular or multilocular cyst lined by an epithelial layer and separated from surrounding normal salivary gland tissue by no distinct collagenous capsule. The cyst shows luminous papillary projections(Fig 11).

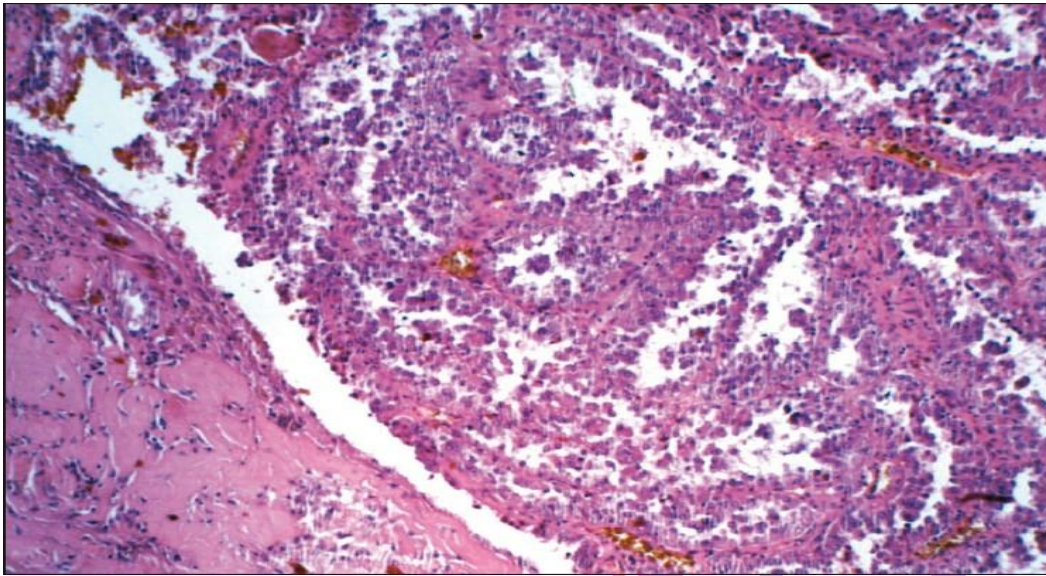


Figure 11: Photomicrograph showing characteristic endophytic papillary projections in papillary cystadenocarcinoma (H and E, 200×)

DISCUSSION

Papillary cystadenoma of salivary glands is an uncommon benign neoplasm. In two large reviews, it constituted 2% and 4.7%, respectively, of all minor salivary gland neoplasms, and 4% and 8.1%, respectively, of all benign epithelial minor salivary gland neoplasms^{4,10,11}. Reviews of clinical, histological, biological features shows that papillary cystadenoma¹ appears to occur more frequently in women, most patients have been older than 50 years of age with several in their seventies. The most common sites are palate and buccal mucosa and least common sites are lips and tongue. The usual presentation is an asymptomatic mass.

In the studies of Armed Forces Institute Of Pathology (AFIP)² the lesion

are widely distributed among major and minor salivary glands. Major salivary glands are more commonly involved than minor salivary glands. On microscopic examination the neoplasm is usually well circumscribed and may be surrounded by a rim of fibrous tissue. There are solid areas and cystic areas into which project papillae lined by cuboidal to columnar cells usually two layers thick. The cells usually have eosinophilic cytoplasm and goblet cells may be present^{3,4}.

Perhaps the most important entity in the differential diagnosis of papillary cystadenoma is cystadenocarcinoma. Sometimes the distinction may be difficult because the neoplasms have same architecture, and also because cystadenoma often shows little

atypia⁴. Both neoplasms usually reveal papillary proliferation of the epithelial lining and are composed of cells that possess bland cytomorphological features.

Differentiation of the tumour types depends largely on the identification of actual infiltration of salivary gland parenchyma or surrounding connective tissue by either cystic or solid epithelium in cystadenocarcinomas. Step sections of a borderline tumour may yield unequivocal evidence of invasion.

The files of Armed Forces Institute of Pathology (AFIP)⁴ have recorded 57 cases of cystadenocarcinoma. The mean patient age in this series was 59 years, and both sexes were affected equally. About 65% of documented tumours occurred in the major salivary glands, and most of these comprised the parotid glands. The preoperative diagnosis of cystadenocarcinoma is complex. When located in the parotid gland, the presentation is usually that of a slowly growing, asymptomatic mass. In a series of 56 cystic lesions of salivary glands including only two cystadenocarcinomas, Layfield and Gopez⁵ reported an overall accuracy of 84%. Other authors described a FNAB diagnostic efficacy of 80% in application to

cystadenocarcinomas. In literature the lesions most often confused with cystadenocarcinomas when performing FNAB are Warthin's tumour or Salivary gland cysts⁹.

The management approach suggested for cystadenocarcinoma to date is similar to that advocated for other low grade salivary gland adenocarcinomas⁶. A number of high and low grade cystadenocarcinomas have been documented in the literature in both animals and humans^{6,7,8} in which complimentary radiotherapy is recommended. High grade tumours frequently show perineural infiltration, vascular or lymphatic channel invasion, infiltration of surrounding connective tissues and regional lymph node metastasis¹¹. In some cases where the mitotic activity is high and there is occasional abnormal mitotic figures and no metastatic lymph node is present, annual revisions were decided for a minimum period of five years.

CONCLUSION

Papillary cystadenocarcinoma of the submandibular salivary gland is an extremely rare entity that can be diagnosed on FNAC. Recognition of cytologic features may be useful in the

differential diagnosis. Computerized tomography is an extremely useful tool for planning the surgery. Access osteotomy is of great significance in inaccessible areas like submandibular salivary gland. Correlations with the clinical and radiological features are also of great importance in arriving at correct diagnosis.

REFERENCES

1. Collins EM. Papillary cystadenoma of accessory salivary gland. *Am J Surg.* 1958 Dec;96(6):749-50.
2. Ellis GL, Auclair PL, Gnepp DR. (eds). *Surgical Pathology of the Salivary Glands.* Philadelphia, PA: Saunders; 1991.
3. Mahler V, Schell H. Papillary cystadenoma: a rare tumor of the minor salivary glands. *Eur J Dermatol.* 1999 Jul-Aug;9(5):387-9.
7. Tsurumi K, Kamiya H, Yokoi M, Kameyama Y. Papillary oncocytic cystadenoma of palatal minor salivary gland: a case report. *J Oral Maxillofac Surg.* 2003 May;61(5): 631-3.
4. Foss RD, Ellis GL, Auclair PL. Salivary gland cystadenocarcinomas. A clinicopathologic study of 57 cases. *Am J Surg Pathol.* 1996 Dec;20(12):1440-7.
5. Layfield LJ, Gopez EV. Cystic lesions of the salivary glands: cytologic features in fine
6. Mills SE, Garland TA, Allen MS Jr. Low-grade papillary adenocarcinoma of palatal salivary gland origin. *Am J Surg Pathol.* 1984 May;8(5):367-74.
7. Pollett A, Perez-Ordonez B, Jordan RC, Davidson MJ. High-grade papillary cystadenocarcinoma of the tongue. *Histopathology.* 1997 Aug;31(2):185-8.
8. Ferrer C, Ramos V, Ferrer E, Sancho R. The role of radiotherapy in the management of salivary gland neoplasms. *Med Oral.* 1998 Aug;3(4):207-221.
9. Klijanienko J, Vielh P. Salivary carcinomas with papillae: cytology and histology analysis of polymorphous low-grade adenocarcinoma and papillary cystadenocarcinoma. *Diagn Cytopathol.* 1998 Oct;19(4):244-9.
10. Alexis JB, Dembrow V. Papillary cystadenoma of a minor salivary gland. *J Oral Maxillofac Surg.* 1995 Jan;53(1):70-2.
11. Waldron CA, El-Mofty SK, Gnepp DR. Tumors of the intraoral minor salivary glands: a demographic and histologic study of 426 cases. *Oral Surg Oral Med Oral Pathol.* 1988 Sep;66(3):323-33.