Papillary cystadenocarcinoma – A Very Rare Case Report

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ABSTRACT

Papillary cystadenocarcinoma is an extremely rare malignant neoplasm characterized by cysts and papillary endophytic projections. It was first defined in 1991 by the World Health Organization as a separate entity. 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 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

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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; inspirited secretions and thickning of the adjacent buccal mucosa distorting the Oro-pharyngeal 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).
Case report
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Dr. Sherin A Khalam et al.

Figure 6: Exposure of Lesion
Modified McGregor lip split incision placed in right side of mandible, Lip was split in the midline Midsymphseal 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 Metrogyl. 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

Figure12: 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).
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. Reviews of clinical, histological, biological features shows that papillary cystadenoma 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) 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 eosinophillic cytoplasm and globlet cells may be present.

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
atypia\(^4\). 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 cystadenocacinoma. 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 Gopez\(^5\) 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 cysts\(^9\).

The management approach suggested for cystadenocarcinoma to date is similar to that advocated for other low grade salivary gland adenocarcinomas\(^6\). 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 metastasis\(^11\). 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
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