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## Papillary cystic acinic cell carcinoma: report of a rare lesion with unusual presentation

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### Abstract

**Introduction:** Acinic cell carcinoma is an uncommon low grade tumour of the salivary glands that constitutes 2.5 to 4% of parotid gland tumours. Acinic cell carcinoma -Papillary cystic variant (ACC-PCV) is histologically composed of tumor with papillary and cystic growth patterns, with varying proportions of one or more cell types. It has been conferred significance because it has a poorer prognosis and is reported to be universally fatal in ten years.

**Case Report:** We present a case of ACC-PCV in a sixteen year old male with unusual unicystic gross appearance, benign cytological picture and characteristic histopathological features. Cystic areas with papillary projection of surrounding cells showing characteristic tombstone or hobnail arrangements were seen.

**Discussion:** The histogenesis and myriad architectural and cellular variations of ACC-PCV have been discussed along with its variegated cytomorphology which may lead to pitfalls in cytodiagnosis. The tumor may pose difficulty in histodiagnosis due to its resemblance to papillary carcinoma of the thyroid.

**Key words:** Salivary gland neoplasm, Papillary cystic acinic cell carcinoma.

## Introduction

Acinic cell carcinomas were considered benign until 1953, when Buxton described their true malignant potential (1). Although it is a low grade malignancy with a 5 year survival of 90%, the papillary cystic variant assumes importance as it has proved to be universally fatal in 10 years (2, 3). This emphasizes the need for prompt and accurate diagnosis.

Acinic cell carcinoma accounts for 2.5 to 4% of salivary gland tumours and a small percentage of these belong to the papillary cystic variant (4). Its relatively slow growing nature and varied histological appearance may lead to misdiagnosis.

## Case Report

A sixteen year old male patient reported with a swelling measuring approximately 5.5x4x3.5 cm in size on the left angle of mandible. The anteroposterior extent of the swelling was from the molar region to the posterior border of mandible and superoinferiorly from middle of tragus to 1.5 cm below the left angle of mandible with elevation of the ear lobule. The swelling was mobile, soft, non tender and well circumscribed (Fig.1). Facial nerve function was normal on examination. A clinical diagnosis of benign salivary gland neoplasm was made.



*Fig 1. Swelling on left angle of mandible elevating lobule of ear*

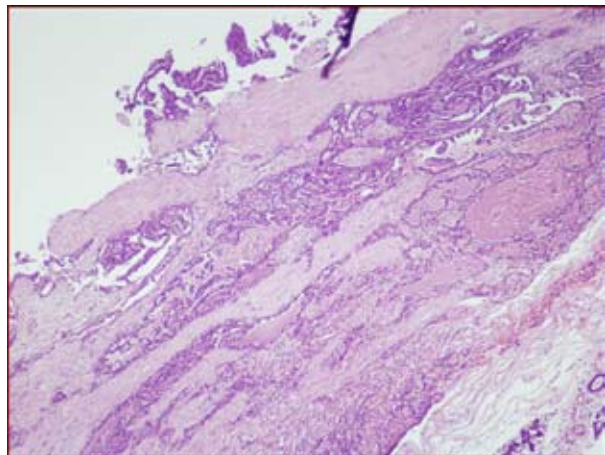
On ultrasonography, isoechoic solid component was seen in the left parotid gland. The thyroid gland appeared normal. Contrast enhanced computed tomography revealed a poorly marginated, lobulated, heterogeneously hypodense mass measuring 5.3x3.5x2.7 cm in

the superficial lobe of left parotid gland with invasion into the left masseter .

Cytological examination showed a few acinar cells with granular cytoplasm and an eccentrically placed nucleus in a proteinaceous background and diagnosis of benign cystic lesion was made. A superficial parotidectomy with preservation of facial nerve was performed. Post operative recovery was uneventful.

The gross specimen obtained was approximately 6x4.5x3 cm in size, bluish grey in colour with a nodular surface. Cut surface showed a large well circumscribed cystic area containing dark chocolate coloured jelly like material that easily separated out from surrounding tissue. An irregular white area of tumour tissue was observed at the superior aspect of this material .

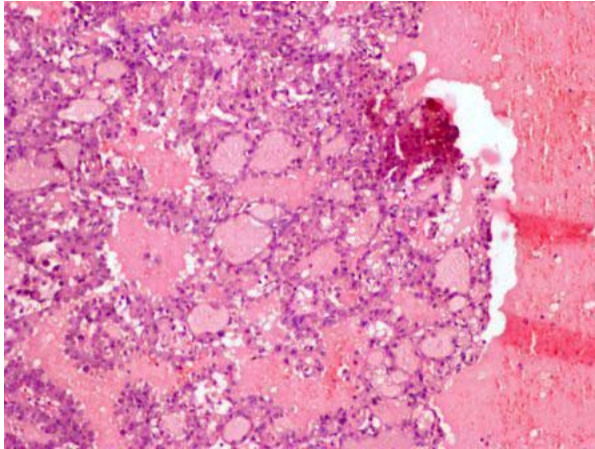
Histopathological examination showed a well circumscribed cystic area surrounded by dense collagenous tissue. Tumour tissue was seen infiltrating the fibrous wall (Fig.2). The lesion predominantly contained homoge-



*Fig 2. Tumour tissue infiltrating fibrous wall*

neous eosinophilic material with a few vacuolated cells at places. The cellular areas showed a follicular arrangement reminiscent of papillary carcinoma of thyroid with cystic spaces containing eosinophilic material lined by cuboidal to flattened cells. Cystic areas with papillary projection of surrounding cells showing characteristic tombstone or hobnail arrangements around the lumen were noted (Fig.3). Parenchymal tissue between the cystic spaces showed cells with basophilic granular cytoplasm and eccentric vesicular nuclei. Few vacuolated cells were also seen.

Periodic Acid Schiff stain with diastase digestion showed positivity for both intracytoplasmic granules and extracellular secretion. Immunohistochemical tests for thyroglobulin and thyroid transcription factor were done to rule out carcinoma of thyroid and found to be negative. Thyroid function tests were performed and found to be normal.



**Fig 3.** Multiple cystic areas containing eosinophilic material with papillary projection of surrounding cells showing characteristic tombstone arrangement

## Discussion

Acinic cell carcinomas of salivary gland origin are destructive neoplasms of individually unpredictable behaviour (5). The tumour may be solid or cystic and distinct morphological growth patterns are seen. These are described as solid, microcystic, follicular and papillary cystic.

The occurrence of these architectural variations indicates that this neoplasm histogenetically differentiates in the direction of terminal ductal-acinar unit of salivary gland; which includes secretory acinar cells, intercalated duct cells, pluripotential reserve cells and myoepithelial cells. The papillary cystic variant is believed to have two forms of genesis: one a retrogressive phenomenon; the other solely neoplastic. The former is a histopathologic reflection of a self destructive quality of some acinic cell carcinomas, the end stage of which is a solitary loculated cyst with attenuated and hydropically altered neoplastic cells in company with neoplastic papillary excrescences in various stages of degeneration (6).

Acinic cell carcinomas occur predominantly in the parotid gland although submandibular and minor salivary glands may also be involved (4, 7). The mean age of occurrence is in the fifth decade, but the papillary cystic variant is reported to occur in younger patients compared to the classic type. Females are involved more commonly. Clinically, Acinic cell carcinoma presents as a slow growing, nontender swelling usually less than 3 cms. in size. Pain may sometimes be present spontaneously or on palpation (4, 7, 8).

The papillary cystic variant is said to have a variegated cytomorphology and is a challenge in salivary gland aspiration. It is composed of large monolayered sheets and numerous small papillary groups; absence of acinar structures, absence of naked nuclei in the background and presence of numerous vacuolated cells in addition to the common cell types (5, 9).

Numerous reasons have been given for difficulty in cytodiagnosis:

- a. Cystic fluid dilutes the overall cellularity leading to a benign diagnosis.
- b. Cytoarchitecture is different from classic type – prominent papillary architecture with more cohesive cells with high nuclear- cytoplasmic ratio.
- c. Change in morphology of tumor cells due to suspension in cystic fluid
- d. Presence of metaplastic oncocyctic or squamoid changes either focally or otherwise (9).

Microscopically, ACC-PCV is usually encapsulated and capsular invasion is a common finding. It displays papillary folds interspersed with cystic spaces. These may be either small cysts with scanty papillary projections or large cystic spaces into which extend delicate papillary growths, stalks, fronds or masses of glandular epithelium supported by thin fibrovascular cords. The most commonly encountered cell types in this variant are the intercalated ductlike and nonspecific glandular cells. However, vacuolated cells are often numerous and acinar cells may be seen. Hobnailing or tombstoning of luminal cells is characteristic of the papillary cystic variant. This is due to bulging of the apical portions of lumen lining cells into the lumen presumably after release of secretions.

The most significant differential diagnosis of ACC-PCV is papillary carcinoma of the thyroid. Thyroid ultrasonography, hormonal assays, and immunohistochemistry for thyroglobulin help in differentiating the two. Other less significant differential diagnosis include cystadenocarcinoma and Polymorphous low grade adenocarcinoma.

Treatment of choice is total parotidectomy. Timely diagnosis and treatment is essential as ACC-PCV has been found to be universally fatal in ten years (3).

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