CASE REPORT

Acinic cell carcinoma, papillary-cystic variant of the parotid gland: A case report with review of literature

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Summary Acinic cell carcinoma (ACC) is a rare, slow growing, low grade neoplasm of salivary glands and some extra salivary sites. ACC-papillary cystic variant is histologically composed of tumor with papillary and cystic growth patterns, with varying proportions of one or more of 5 cell types which include ”hobnail” acinar, intercalated, vacuolated, non-specific glandular and clear cells. The clinical picture is not specific and diagnosis is based on the histopathologic examination. The present case describes ACC—papillary cystic variant and discusses the myriad architectural patterns exhibited by it. © 2005 Elsevier Ltd. All rights reserved.

KEYWORDS
Malignant salivary gland tumor;
Acinic cell carcinoma;
Papillary-cystic variant

Introduction

ACC of the salivary glands are distinctive neoplasms of individually unpredictable behaviour. Salivary gland tumors comprise 0.3–0.9% of all tumors and ACC accounts for 6% of all salivary gland tumors. ACC arising in the parotid was first described by Nasse in 1892. Terms ”acinar” and ”serous” designate the histologic resemblance of tumor cells to the secretory parenchymatous cells of the parotid, grouped in grape like clusters and hence the latin name ”Acinus”. Although previously considered benign and classified as ”Tumors” by the WHO, they are now considered as ”Carcinomas”. The purpose of this presentation is to study and discuss the cytopathology and histopathology of the various architectural patterns of ACC.

Case report

A 26 year old male was referred for a firm, painless, freely movable, atraumatic lump measuring...
Figure 1  Poorly circumscribed tumor infiltrating into the surrounding fibrous capsule (H & E ×100).

Figure 2  Characteristic papillary growth pattern with intervening cystic spaces filled with eosinophilic secretions. Thin papillae are lined by a single layer of "hobnailed" acinar, vacuolated and non-specific ductal cells (H & E ×400).

Figure 3  Tumor cells invading the stromal blood vessel/vascular invasion by the tumor cells (H & E ×400).
2 × 1.5 cm, extending from the lower left ear lobe to the angle of the mandible and slowly enlarging since last 4 years. Provisionally diagnosed as a benign salivary gland tumor, excisional biopsy was performed.

Hematoxylin and Eosin stained sections revealed a serous salivary gland, with an adjacent poorly circumscribed infiltrating tumor (Fig. 1) with papillary and cystic growth patterns. The papillae were thin and lined by a single layer of "hobnail" acinar, vacuolated and non-specific ductal cells. Nuclei were round, regular, hyperchromatic and eccentrically placed with small central nucleoli. Mitotic activity was minimal (Fig. 2). Infiltration into the surrounding stroma, hemorrhage and vascular invasion was noted (Fig. 3).

PAS and D-PAS stain showed positivity for both intracytoplasmic granules and extracellular secretion (Fig. 4). Mucicarmine stain was negative for mucin.

Biopsy for Cytokeratin and S-100 immunocytochemical assay demonstrated broad spectrum keratin (AE1–AE3) positive and S-100 focally positive in tumor cells.

Results lead to the diagnosis of ACC-papillary-cystic variant following which total parotidectomy was performed. The surgical margins were free of tumor tissue. Post-operative recovery and healing were uneventful and there have been no signs of recurrence even after twelve months of follow-up.

Discussion

ACC is a neoplastic growth of epithelial cells showing serous acinar differentiation, presents at a younger age than other salivary gland tumors and shows a female predilection. The most favored site is the Parotid (86%), occasionally the submaxillary gland, minor salivary glands and rarely extra-salivary sites like breast, pancreas and lungs.

Fine needle aspiration biopsy of ACC-papillary cystic variant reveals tightly cohesive fragments of neoplastic epithelium seen as monolayered sheets or with a prominent papillary architecture, neoplastic cells with abundant foamy to coarsely granular cytoplasm, high nuclear: cytoplasmic ratio, ductal type epithelium, cystic material and degenerated cellular debris, cells with squamoid and oncocytic metaplasia, vacuolated cells and lack of a predominant single cell component.

Histopathologically, a distinct fibrous connective tissue usually encapsulates the tumor, but marginal infiltration is a prominent feature. The tumor is composed of diverse cell types and these are acinic cells, vacuolated cells, intercalated cells, non-specific glandular cells and clear cells. Acinic cells resemble the polyhedral cells of normal acini and contain abundant finely granular cytoplasm which may be amphophilic, pale eosinophilic or basophilic. Nuclei are generally small, regular, hyperchromatic with a prominent nucleolus. Characteristic cytoplasmic granularity with strongly PAS and D-PAS positive granules are seen. Vacuolated cells are characterized by 1 or 2 large or multiple vacuoles. Intercalated cells are cuboidal cells with limited amphophilic cytoplasm, distinct borders. Non-specific glandular cells lack specific glandular features, have a faintly eosinophilic or amphophilic cytoplasm, and no granules. Characteristically seen in papillary-cystic variant, they have a tendency to form "hobnail cells", presumably after releasing their secretions into glandular
spaces. Clear cells are characterized by their non-stainable cytoplasm and distinct non-distended cell borders.\textsuperscript{13–15}

The myriad architectural patterns which may be seen in ACC are: solid, solid-lobular, acinar-microcystic, papillary cystic, tubuloductal, follicular/macrocytic and dedifferentiated. Although a single pattern usually dominates, a mixture of histologic patterns may also be observed. Solid is characterized by closely continuous arrangement of acinar type cells surrounded by a scanty, vascular stroma. Solid-lobular additionally shows dense fibrous septae separating tumor cells in sheets. Microcystic pattern exhibits intercellular spaces formed due to coalescence of intracellular vacuoles. Papillary-cystic typically involves transition of usual dense cellularity into papillary folds interspersed with cystic spaces. It may vary from small cysts with scanty papillary projections to large cystic spaces into which extend delicate papillary folds intermixed with cystic spaces. Dedifferentiated exhibits coexistence of both low grade and high grade dedifferentiated component. Few tumors consist of small, round, darkly basophilic calcospherites and lymphoid tissue.\textsuperscript{16–18} It has been concluded that ACC arises from the pleuripotent stem cells found at the acinar-intercalated duct junction and/or in the intercalated duct proper of mature salivary glands and these are responsible for the manifold histologic patterns.\textsuperscript{19}

Various immunohistochemical studies have demonstrated the presence of CK, Carcinoembryonic antigen, $\alpha$-1 antitrypsin, $\alpha$-1 antichymotrypsin, Leu-M, amylase, transferrin, lactoferrin, vasoactive intestinal polypeptide & S-100.

Metastasis is rare and the choice of treatment is total parotidectomy.\textsuperscript{1,4,13,20}

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