Research Paper

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Acinic Cell Carcinoma of Salivary Gland Follicular Variant: A Case Report

Medical Science

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ABSTRACT

Acinic cell carcinoma (ACC) is a rare, slow growing, low grade neoplasm of salivary glands. ACC-follicular variant is histologically composed of closely packed round cystic spaces filled with colloid like material. The follicles are lined by intercalated duct like cells and non-specific glandular cells. The clinical picture is not specific and diagnosis is based on the histopathologic examination. We present a case of acinic cell carcinoma in a 40 year old female who reported with a painful swelling in the parotid region, which was clinically diagnosed as pleomorphic adenoma and histologically diagnosed as acinic cell carcinoma follicular variant and discuss the myriad architectural patterns exhibited by it.

Introduction:

Acinic cell carcinoma is a malignant salivary gland tumor with cells showing serous acinar differentiation. Several variants i.e. solid, microcystic, papillary cystic and follicular are seen.[1] Follicular variant is an infrequent finding. It poses a considerable diagnostic challenge due to its clinical and cytological resemblance to pleomorphic adenoma. We report a case of follicular variant of acinic cell carcinoma of parotid in a 40 year old lady.

Case Report:

A 40 year old female presented with painful enlargement of the right parotid region since 2 years. The swelling was firm to hard and firmly adherent to the overlying skin.

A provisional diagnosis of pleomorphic adenoma was made and superficial parotidectomy was done.

Grossly, the swelling was grey white to grey brown measuring 5x4 cm having a grey white cut surface with few cystic spaces.

Histopathology showed cells with finely granular basophilic cytoplasm arranged in solid, acinar and follicular pattern. The lobular architecture of normal salivary gland was not seen. Small microcystic spaces with few large spaces were seen. Focal collection of round cystic spaces filled with homogeneous eosinophilic material lined by duct like cells were seen. The material was PAS positive and diastase resistant (**Figure 1**)

On immunohistochemistry, there was widespread immunoreactivity for low molecular weight cytokeratins 5/6 alongwith focal immunoreactivity for S 100 (**Figure 2**). Immunonegativity for thyroglobulin ruled out ectopic thyroid neoplasm (**Figure 3**). These results confirmed the diagnosis of Acinic cell carcinoma-follicular variant.

Discussion:

ACC is a primary glandular neoplasm demonstrating differentiation towards the terminal intercalated ductal acinar unit and exhibiting one or more of histologic patterns.[2] This tumor was first recognised by Nasse in 1892.[3] The parotid is the most common primary site involved(90%). ACC accounts for 1-6% of all salivary gland tumors[4] and 7% to 17.5% of malignant salivary gland tumours. The average age of diagnosis is 38 to 46 years, which is a decade younger than with other parotid malignancies and the female to male ratio is 2:1.

ACC arises in different morphologic growth patterns like solid, microcystic, follicular and papillary cystic. A mixture of patterns are common while observing different fields.

Solid or classic pattern comprises of well differentiated acinar cells with prominent basophilic to greyish granularity of cytoplasm. Cells are polygonal with well defined cytoplasmic borders.[5]

Microcystic pattern consist of prominent cellular vacuolization and intercellular cystic change, producing a fenestrated appearance[6].

Papillary cystic variant is a rare variant showing papillary projections. Papillae are covered by hobnailed cells, intercalated duct like cells and cells with eosinophilic cytoplasm. These cells have central nuclei with indistinct cell borders.

Follicular variant is extremely rare comprising of closely packed cystic spaces lined by flattened epithelium and filled with PAS (Periodic Acid Schiff) positive and diastase resistant colloid like material highly reminiscent of thyroid follicles.

The variable histologic appearance coupled with its uncommon occurrence accounts for its diagnostic difficulty.

The tumors exhibit immunoreactivity for cytokeratins, EMA, CEA and amylase. Focal positivity for S100 protein is also seen. Ultrastructural findings confirm the presence of zymogen like granules in tumor cells.

In the case we encountered, the intercalated duct like cells were arranged in solid, cystic and follicular patterns. The differential diagnosis considered were normal salivary gland tissue but ACC lacked the normal lobular architecture, metastatic thyroid carcinoma which was ruled out by immunonegativity for thyroglobulin.

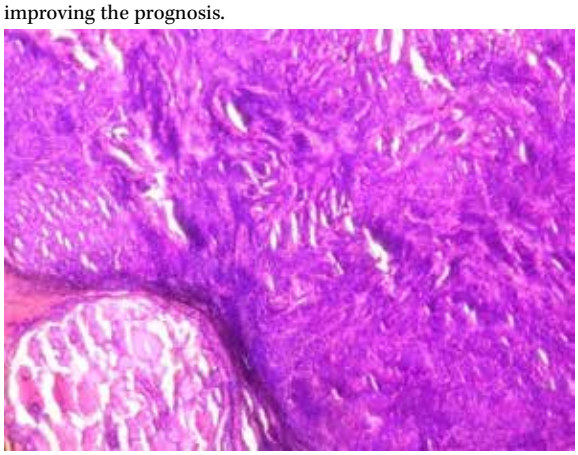
Adenoid cystic carcinoma can be considered in the differential diagnosis but not in our case due to presence of minimal cytological atypia and coarsely granular to vacuolated cytoplasm in ACC. The presence of serous acinar differentiation and absence

of clear cells and oncocytic change rules out mucoepidermoid carcinoma.

The treatment of choice is complete surgical excision, supplemented by post operative radiotherapy if margins are involved.

Conclusion

ACC developes especially in the fifth decade of life, more frequent in women, and with the parotid as the most frequently involved salivary gland. Present case highlights the importance of histopathology and immunohistochemistry in the management of salivary gland tumors as the clinical diagnosis is often misleading. Thus histopathology and immunohistochemistry is useful in the management of the patient at an early stage, thereby improving the prognosis.



Figure–1: ACC - Mixed pattern (Follicular, Acinar and solid pattern) (H & E Stain 4X & 4X Zoom)

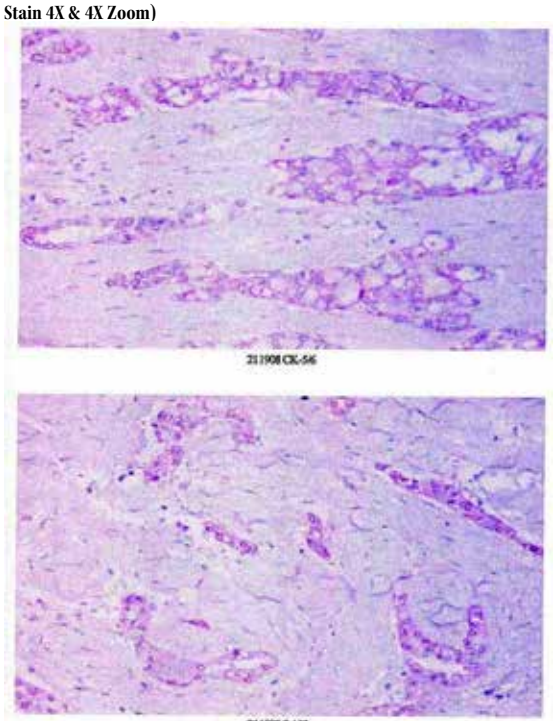


Figure -2 Immunopositivity for cytokeretin 5-6 and S-100 protein (100X).

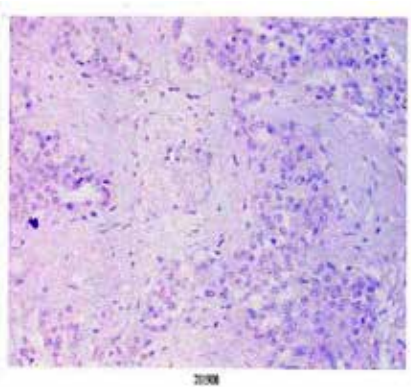


Figure -3 immunonegativity for thyroglobulin (100X)

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