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Case Report

Acinic cell carcinoma, papillary-cystic variant: a rare case diagnosed in fine needle aspiration cytology

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ABSTRACT

Acinic Cell Carcinoma is a rarest malignant epithelial neoplasm arising from pleuripotent stem cells. ACC constitute 6% of all salivary gland tumor. Most common site is parotid. It is slowly growing tumor with female preponderance most commonly occur during fifth and sixth decades of life. Diagnosis by imaging study are complex, but USG is helpful in evaluating tumor. We are presenting a 9year female, complaint of a painless, freely movable, atraumatic swelling on left cheek measuring 2x1cm^2, slowly enlarging since, 8 months. FNAC was performed from the swelling and routine stain (H and E, MGG, Pap) and special stain (PAS) was performed. On cytological smears, overall findings are suggestive of papillary-cystic variant of ACC which is difficult to diagnosed in cytology. PAS stain showed positivity for both intra-cytoplasmic granules and extra-cellular secretions. ACC is a slowly growing multi-potent malignant salivary gland tumor. Management is with surgery.

Keywords: Acinic cell carcinoma, Child, FNAC, Papillary-cystic variant, Parotid gland, Salivary gland tumor

INTRODUCTION

Fine needle aspiration cytology is reliably used to classify most conditions involving the salivary gland. It is useful for establishing, the diagnosis in unusual cases or narrowing the differential diagnosis.[1] Epithelial salivary gland neoplasms are rare in children and adolescents. They represent only the 1-5% of the total number of salivary gland tumors. In the infancy, it has been reported that 35% of salivary gland tumors are malignant and out of these mucoepidermoid carcinoma is the most frequent, while the occurrence of acinic cell carcinoma in children is very rare. The major salivary glands, the parotid and submandibular, are the main site of occurrence. Fine needle aspiration cytology (FNAC) can be considered a safe, rapid and helpful diagnostic test in determining the nature of a parotid mass.[2] Terms "Acinar" and "Serous" designate the histologic resemblance of tumor cells to the secretory parenchymaous cells of the parotid, grouped in grape like clusters and hence the latin name "Acinus". Althought previously considered benign and classified as "Tumors" by the WHO, they are now considered as "Carcinoma". Acinic Cell Carcinoma is a malignant epithelial neoplasm arising from pleuripotent stem cells.[3,4] It is slowly growing tumor with female preponderance most commonly occur during fifth and sixth decades of life.[5,6] Possible causes of acinic cell carcinoma include previous radiation exposure and familial predisposition.[5] Histologically it has got many variants: solid, solid-lobular, acinar-microcystic, papillary cystic, tubuloductal, follicular/macrocystic and dedifferentiated.[4] The cytological picture often is dominated by cells denoted as acinic cells because of their great resemblance to normal acinar cells. We are

presenting a case of an acinic cell carcinoma, papillary-cystic variant in a 9-year-old female.

CASE REPORT

We report an observational study of 9-year-old female, referred to the pathology department at Government Medical College for assessment of firm, painless, freely mobile, atraumatic swelling on left cheek, measuring 2x1cm^2, which was noticed eight months before and it was slowly enlarging in nature. A fine needle aspiration cytology (FNAC) was performed under full aseptic precaution, with consent of her parents. Routine stains (H and E, MGG, Pap) and special stain (PAS) performed. In cytological studies, smears showed epithelial cells arranged in clusters, groups, sheets and scattered singly.

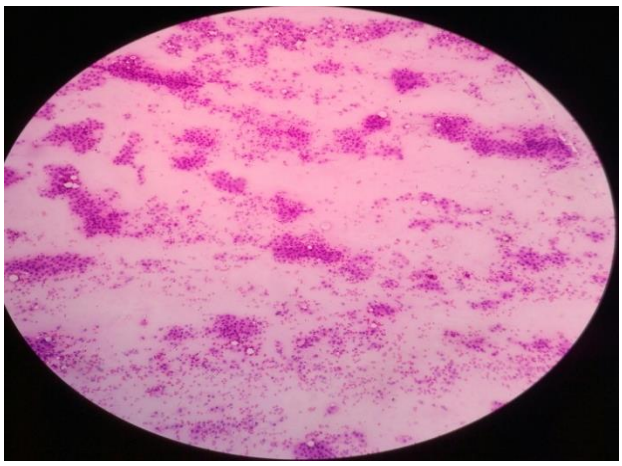

Figure 1: Sheets of tumor cells-acinic cell carcinoma (H and E, stain 10x view).

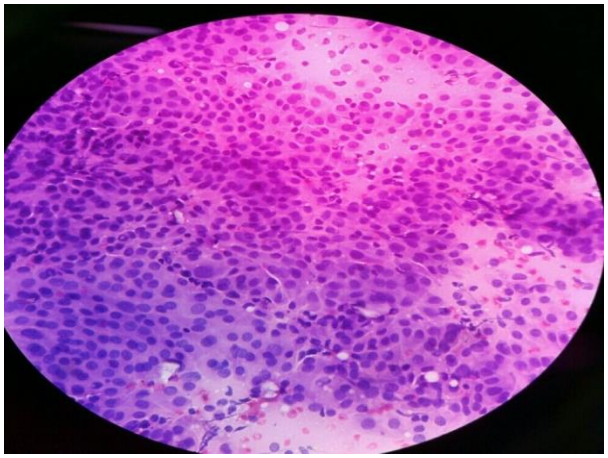

Figure 2: Tumor cells-acinic cell cacinoma (pap stain 40x view).

Cells were round to oval in shape with round to oval nuclei, regular nuclear membrane, finely granular chromatin, inconspicuous to prominent nucleoli with moderate amount of cytoplasm, few cells show finely vacuolated cytoplasm. Occasional mitosis seen. Periodic Acid Schiff stain showed positivity for both intra-cytoplasmic granules and extracellular secretion. Overall cytological features are suggestive of Acinic Cell Carcinoma. Surgery is planned, and mass is excised, and we received the mass in 10% formalin, single lobular structure labelled as "Swelling over left buccal mucosa" measures 2x1.3x1cm^3 in size.

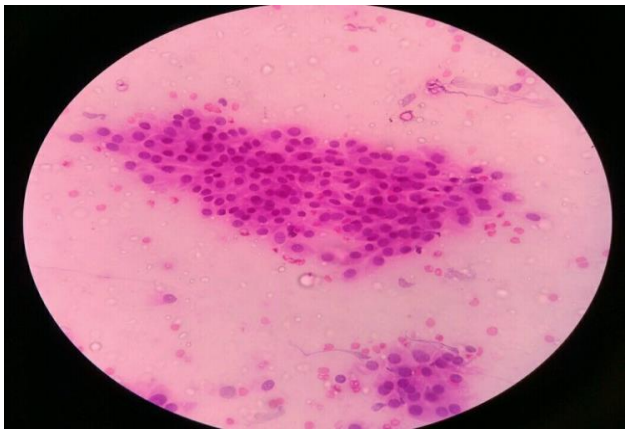

Figure 3: Tumor cells-acinic cell cacinoma (pap stain 40x view).

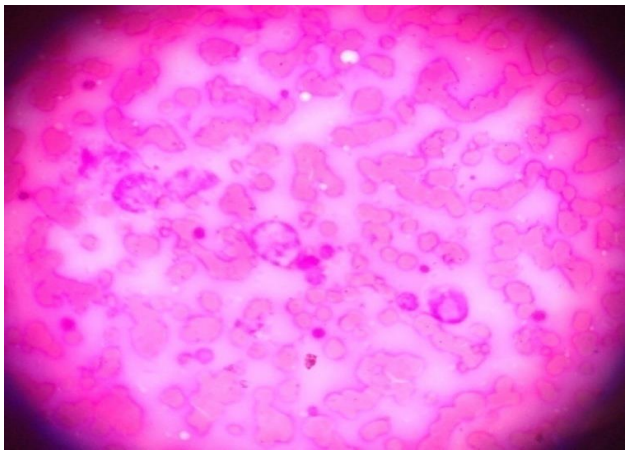

Figure 4: Intra-cytoplasmic positivity seen in tumor cells-acinic cell carcinoma (PAS stain 40X view).

External surface was smooth and nodular, greyish white in color. Cut surface was whitish in color, soft in consistency, dissected and tissue processed, and block prepared. Routine stain (Hematoxylin and Eosin) was performed. Histopathological findings showed tumor cells arranged in papillary cystic, microcystic, solid pattern and sheets. At places cells surrounds the luminal spaces filled with eosinophilic material. Cells were round/oval in shape having central round/oval nuclei, granular/vesicular chromatin, inconspicuous nucleoli with moderate to abundant eosinophilic cytoplasm. Focal areas of haemorrhage and hemosiderin laden macrophages were seen. Marked cytological atypia/mitosis/necrosis/ Perineural invasion was not seen. Surrounding fibrous capsule showed occasional small tumor foci. Histopathological diagnosis was Acinic Cell

Carcinoma-papillary cystic variant. The patient treated successfully with surgery and was subsequently managed with close surveillance. She has had no recurrence of her acinic cell carcinoma. Procedures for staining are as follow

Procedure for hematoxylin and eosin staining

After preparing the smear immediately fix it in methanol for 10 minutes. (Wet Fixation). Dry the smear. First apply Haematoxylin for 2 minutes. Wash the slide 2 times in tap water. Apply Eosin for 5-10 seconds. Wash with tap water for 2 times. Dry the slides and mount with D.P.X. Label it properly.

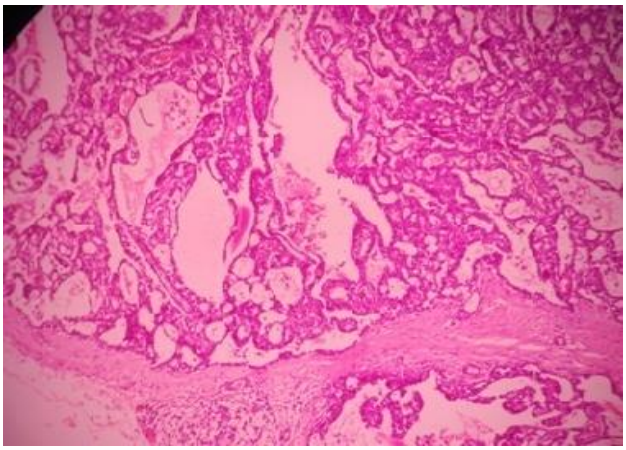

Figure 5: Papillary-cystic architecture-acinic cell cacinoma (H & E stain 10X view).

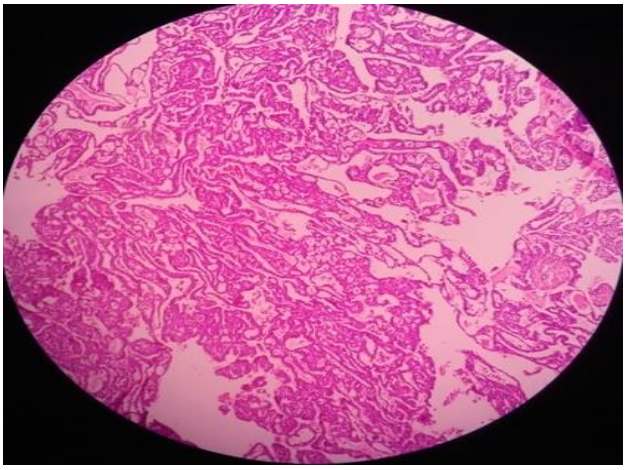

Figure 6: Papillary-cystic architecture-acinic cell cacinoma (H & E stain 4X view).

Procedure for may-grünwald-giemsa staining

After preparing the smears first dry it in air for 5 minutes (Dry Fixation) then fix it in methanol for 10 minutes., Dilute the MGG stain with buffer (1:10), Apply the stain over the smear for 5minute. Remove the stain from smear. Dilute the Giemsa stain with tap water (1:4). Apply the stain over the smears for 15-20 minutes. Wash with tap water. Dry the smears and mount with D.P.X. Label it properly.

Procedure for pap staining

Fix the smears in methanol for 10 minutes after preparing the smears. (Wet Fixation). Wash with tap water for 30 seconds. Apply Hematoxylin for 3 minutes. Wash with tap water for 2 times. Dehydrate the smears with 90% alcohol for 30 seconds. Apply OG6(Orange Gelb) for 5 minutes. Dehydrate with 90% alcohol for 30seconds. Apply EA 36(Eosin Azure) for 6 minutes. Dehydrate with 90% and 95% alcohol for 30 seconds. Dry the smears and mount with D.P.X. Label it properly.

Procedure for PAS stain

Take test and positive control slide SD. Deparaffinize section by keeping in oven for 5 min and clear by 2 changes of xylene for 15 min each.Dehydration with graded alcohol 2 change for 2 min. Bring section to distilled water. Place slide in jar containing 0.5% periodic acid for 5 min. Wash well in several changes of distilled water. Stain with Schiff's reagent for 10-15 min. Tap water washes for 5-10 min. Counter stain with Harrish Hematoxylin for 1 min. Wash in running tap water. Differentiate in 1 % acid alcohol for 1-2 sec. Wash well in running tap water. Dehydrate in alcohol, clear in xylene, mount with D.P.X. Label it properly.

DISCUSSION

Malignant epithelial salivary gland neoplasms are infrequent in adults and rare in children.[1] The major salivary glands, the parotid and submandibular, are the main site of occurrence.[2] Acinic Cell Carcinoma is the third most common malignant tumor of parotid after the muco-epidermoid and adenoid cystic carcinoma.[3,4] Acinic Cell Carcinoma in children is very rare, since it represents about the 3% of all malignant parotid neoplasm.[2] Acinic Cell Carcinoma constitute 17% of primary salivary gland tumor.

Though it is slowly growing, it has got tendency to recur and metastasis. The clinical evalution of a parotid mass can be difficult in children. Clinically, these lesions manifest as a painless, enlarging lesions and grow slowly, without symptoms.[2] Diagnosis of Acinic Cell Carcinoma is very difficult as it produces radiological and cytological similarity.[5,6] Diagnosis of Acinic Cell Carcinoma by imaging study are complex, but ultasonography is helpful in evaluating tumor size, location, nature and invasiveness.[6] The cell of origin is believed to be the reserve cell of the terminal duct, a concept supported by ultrastructural studied by Erlandson and Tandler.[7-9] The probable explanation is the relative rarity of the tumor, its generally benign appearance and its moderate malignancy, if the tumors are considered as a whole.[10] Fine needle aspiration cytology (FNAC) can

be considered a safe , rapid and helpful diagnostic test in determining the nature of a parotid mass.[2]

As it has been reported to have a specificity of 91% and a sensivity of 96% when sufficient cells are present.[2] On fine needle aspiration cytology of acinic cell carcinoma, smears are usually cellular. The smear background lacks cell debris, inflammatory cells or extracellular mucin. Numerous bare nuclei are present, which are differentiated from lymphocytes by lack of a cytoplasmic rim. Fine granularity is seen in the background of tumours with prominent acinar cell differentiation.

This and the bare nuclei are derived from fragile neoplastic cells. The tumour nuclei are 'bland,' round or oval and slightly larger than normal acinar nuclei but marked nuclear pleomorphism is not seen. Neoplastic cells occur in loose sheets of varying sizes that lack the well-defined, rounded outlines of normal acinar cell groups. Capillaries may be seen within cellular fragments. Moderate to abundant, finely granular/vacuolated cytoplasm is present. In tumours with prominent intercalated, ductal or non-specific glandular cells, the amount of cell cytoplasm and granularity vary, but on the whole, there is cellular uniformity. Non-specific glandular cells have scanty, finely granular cytoplasm and are arranged in multilayered cohesive clusters with indistinct cell borders. Infrequent findings are lymphocytic infiltration, cystic change and oncocytic metaplasia.[11]

As it arises from pleuripotent stem cells, there is lack of single cell component. So cytologically it mimics other tumors like retension cyst, warthin tumor, mucoepidermoid carcinoma or cystadeno-carcinoma, but papillary clusters with acinic ells with granular cytoplasm helps in confirmation of Acinic Cell Carcinoma. Cytoplasmic zymogen secretory granules identifiy acinar differentiation. Neopastic acinar cells typically have abundant, pale basophilic cytoplasm with purplish cytoplasmic granules and eccentrically located nuclei. They are usually polygonal with nuclei that vary from darkly basophilic to slightly vesicular. Periodic acid Schiff (PAS) stain highlights the cytoplasmic secretor granules, which are resistant to diastase digestion. The zymogen type granules are unstained by mucicarmine, which helps distinguish them from mucin in mucous cells.[3]

Histopathologically, the tumor is well circumscribed with a distinct capsule, may be solid or cystic and distinct morphological growth patterns are seen. These are described as solid, microcystic, follicular and papillary cystic tumors. They are composed of diverse cell types and include acinic cells, vacuolated cells, intercalated cells, nonspecific glandular cells and clear cells. Acinic cells resemble the polyhedral cell of normal acini and contain abudunt finely granular cytoplasm which may be amphophilic, pale eosinophilic or basophilic. Characteristic cytoplasmic granularity with strongly PAS and D-PAS positive granules are seen. 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 "hobnil cells", presumably after releasing their secretions into glandular spaces. Large lobules or nests of tumor cells with little intervening stroma are characteristics.

Management is easy in low grade tumor as it is slowly growing in which few (20%) patient experience local recurrence and metastasis.[12] Although the immunoprofile is non-specific, acinic cell carcinomas are reactive for cytokeratin, transferrin, lactoferrin, alpha 1-antitrypsin, alpha 1-antichymotrypsin, IgA, carcinoembryonic antigen, Leu M1 antigen, cyclooxygenase-2, vasoactive intestinal polypeptide, and amylase. The zymogen granules in the neoplastic acinar cells are often nonreactive with anti-α-amylase immunostain, an enzyme in zymogen granules of normal serous acinar cells. Reactivity for oestrogen receptor, progesterone receptor, and prostate-specific antigen has been described in some tumours. Approximately 10% of tumours are positive for S-100 protein. In the largest molecular analysis of thesetumours 21 (84%) of the 25 tumours studied showed LOH in at least one of the 20 loci on chromosomes 1, 4, 5, 6 and 17.

The most frequently altered regions were noted at chromosomes 4p,5q, 6p and 17p regions. Chromosomes 4p15-16, 6p25-qter and 17p11 showed the highest incidence of alterations. Another study of multiple spatially obtained samples from one tumour showed evidence for polyclonality suggesting different origins for this tumour.[13] Aggressive metastatic ACC occurs due to deletion of tumor suppressor gene CDKN2A. So, if possible after surgical removal if you do molecular study, then it helps in prevention of metastasis.[14] Size of the original tumor appeared to correlate with overall prognosis.[9] Features of prognostic importance are pain or fixation, gross invasion, desmoplasia, cytological atypia, increased mitotic activity and adequacy of initial excision.[12] Usually, acinic cell carcinomas initially metastasize to cervical lymph nodes and subsequently to more distant sites, most commonly the lung.[13]

CONCLUSION

Papillary cystic variant of Acinic cell carcinoma is rarely seen, especially in young people. Fine needle aspiration cytology is a useful diagnostic procedure that can help diagnose this relatively uncommon type of salivary gland neoplasm and guide its management. Acinic Cell Carcinoma is a slowly growing multi-potent malignant salivary gland tumor and it can be diagnosed early with non-invasive techniques like Fine needle aspiration cytology, and so the surgical excision is possible with good prognosis.

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Ethical approval: Not required

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