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Acinic Cell Carcinoma-Papillary Cystic Variant with numerous psammoma body: A Case Report

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ABSTRACT

Acinic cell carcinoma (ACC) is a rare, low-grade malignant salivary gland neoplasm. Parotid gland is the most common location reaching up to 90% of cases. Papillary cystic variant (PCV) is a much more seldom subtype of ACC. Proper diagnosis of ACC-PCV is only made on histopathology examination. The presence of psammoma body in salivary gland lesions is infrequent finding that may be confused with other benign or malignant lesions. The present case describes ACC-PCV in a 31-year-old Saudi male patient referred to King Abdulaziz University Hospital as post superficial parotidectomy which the patient underwent 3 months ago. This case emphasizes the importance of distinguishing between neoplastic and non-neoplastic lesions with numerous psammoma bodies.

Key words: papillary cystic acinic cell carcinoma–psammoma body–histopathology–parotid gland.

1 INTRODUCTION

Acinic cell carcinoma (ACC) is a rare, low-grade malignant salivary gland tumor. It accounts for 1%- 3% of all salivary gland tumors [1] . The majority of cases occur in parotid gland, approximately 90% of cases. Papillary Cystic Variant (PCV) is even more uncommon subtype of ACC and only histopathological examination can lead to a proper diagnosis. PCV exhibits papillary and cystic growth patterns along with one or more different types of cells. It usually presents in younger patients (16-40 years) when compared to the classic type that characteristically presents in older individuals. Although a low-grade tumor, it is important to recognize this variant-PCV- as it has proved to be universally fatal [2] . Papillary Cystic architecture is associated with an aggressive behavior. Spiro et al documented a 0% survival rate in patients with ACC-PCV [3, 4] . However, no such finding was described in another study conducted by Ellis et al [5] .

Herein, we report a referred case diagnosed as acinic cell carcinoma-papillary cystic variant (ACC-PCV) with numerous Psammoma bodies, which is uncommon finding in salivary gland.

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2 CASE REPORT

A 31-year- old Saudi male patient was referred with left facial swelling post superficial left parotidectomy 3 months ago. During the operation, the cyst was accidentally ruptured and showed clear serous fluid (as per outside report). The patient was diagnosed as having ACC at outside facility. On physical examination, the patient appeared well, conscious, oriented with all vital signs within normal limit. By palpation: Tender wound site was noticed which was interpreted as involvement of left temporal branch of facial nerve. Investigations includes MRI Neck was performed in our hospital and revealed a slight subcutaneous enhanced signal intensity overlying the superficial part of the left parotid gland with a small non-enhancing focus at the superior aspect of the gland. However, this finding could be suggestive of a cyst or an operative change. Both parotid glands were symmetrical in size with no definite evidence of residual or recurrent masses. We received two blocks and two Hematoxylin and eosin stained slides accompanied with histopathology report. As per outside histopathological report, the received piece of parotid gland measuring (2.5x2x2

cm), showed an opened cyst measuring (2x1x1 cm). The cyst wall was about 1mm in thickness. Microscopic examination revealed Sections of papillary cystic lesion lined by epithelial cells with basophilic granular to vacuolated cytoplasm. The cells lining the papillary structures exhibited hobnail pattern. The nuclei were oval with focal clearing but no mitosis noted. Numerous psammoma body calcifications were noted. There was no perineural or lymphvascular invasion identified. Two lymph nodes were seen with no significant pathological impact. A panel of immunehistochemical markers was used, and the tumor cells were positive to DOG-1, CK8-18, CK-PAN and CK7, while negative to S-100, p63, SMA, GCDFP, CD10, TTF-1, CK5/6, and CK20. Ki-67 was approximately 1-2%. Special stains were performed with PAS positivity and resistance to PAS-D in cytoplasmic granules, while mucicarmine was negative. All the above microscopic, IHC, and special stains support the diagnosis of ACC-PCV staging as pTNM, AJCC 8^{th} Edition [6, 7] pT1, N0.

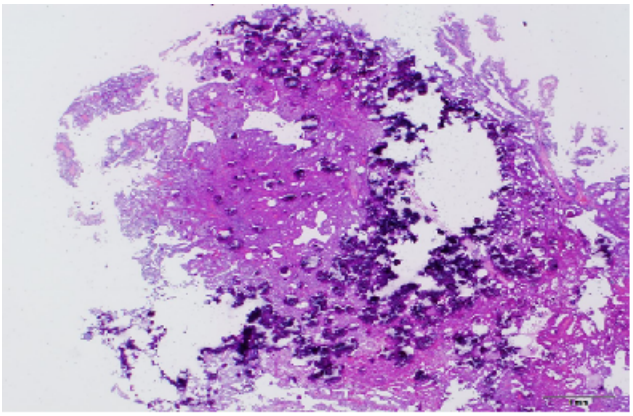

Figure 3. PSAMMOMA BODIES, H&E, 2X

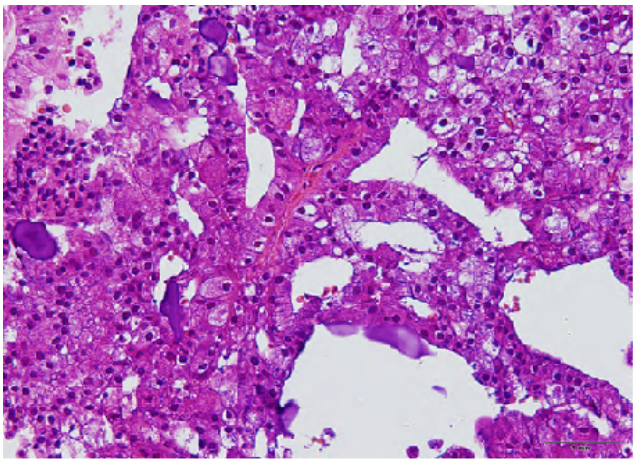

Figure 1. TUMOR, H&E, 20X

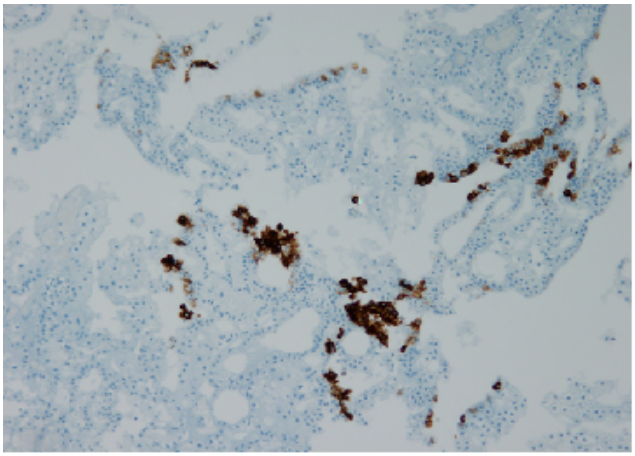

Figure 4. CK7, 10X

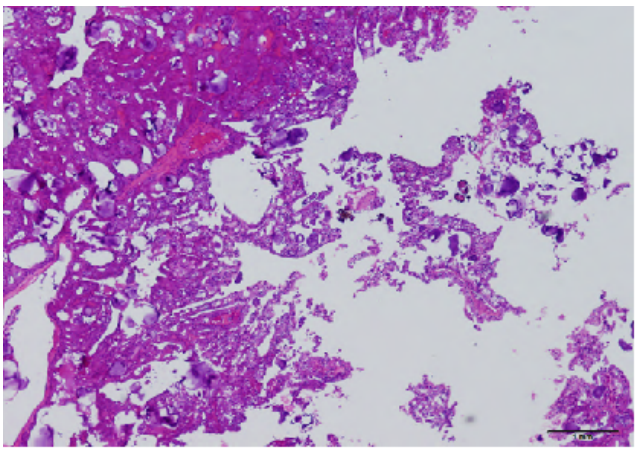

Figure 2. TUMOR, H&E, 4X

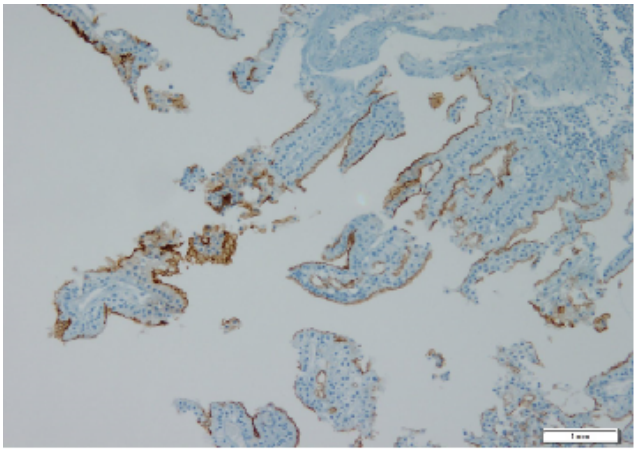

Figure 5. DOG-1, 10X

3 DISCUSSION

Acinic cell carcinoma (ACC) is a malignant salivary gland neoplasm composed of cancer cells with acinar features [8] . It constitutes about 1%-3% of all salivary gland tumors [1] . ACC shows female to male ratio 1.5:1 [8] . The peak incidence is in fifth and sixth decades of life. More than 90%-95% of ACC occur in the parotid gland [8] . Most patients present with slow-growing, solitary, mobile mass; may occasionally be painful or attached to adjacent muscle or skin as 5%-10% develop facial nerve paralysis [8] . Macroscopically, the mass is usually single, well-delineated nodule, solid and yellow, gray or brown in color; it may be multiple or bilateral. Furthermore, cystic lesions may be identified grossly. The classic tumor cells exhibit features of serous cellular components with finely granular, basophilic cytoplasm, and round, eccentric nuclei. In addition, vacuolated, clear, oncocytic, hobnail, and non-specific glandular morphological features can be seen. These types of cells forming a wide variety of growth patterns: Solid, microcystic, papillary cystic, and follicular. Although, various cellular morphologies may exist in the same tumor; there is no significant prognostic impact has been found based on the tumor architecture with different cell types. ACC rarely depicts prominent nuclear pleomorphism, high mitotic figures or necrosis. In which, focal presence of these features makes the tumor dedifferentiated; that is associated with poor prognosis. Papillary cystic variant is a well- known subtype of ACC histologically with a proportion approximately one fourth of ACC [9] . However, histopathological diagnosis has been shown to be more accurate than a diagnosis made through fine needle aspiration biopsy (FNAB). Moreover, cytology finding usually poses a diagnostic problem because of not well recognized descriptions of ACC, papillary cystic variant (ACC-PCV). Some cytology literature reports only a few cases of ACC-PCV; while most case studies were made on histopathology examination. Histologically, vacuolated and intercalated duct cells are frequently seen in papillary cystic variant along with papillary and cystic growth architectures, while acinar cells are more obvious in the solid and microcystic patterns [9] . Psammoma bodies represent a process of dystrophic calcification. Both benign and neoplastic lesions of salivary glands may reveal psammoma bodies, mostly in elderly individuals, but they are usually infrequent. However; one study reports a case with psammoma body-rich papillary cystic acinic cell carcinoma on fine needle aspiration findings [9] . Interestingly, the presence of myriads psammoma bodies in our case of ACC-PCV is a unique finding. The existence of cystic spaces makes the differential diagnosis of low-grade mucoepidermoid carcinoma (MEC) is possible. Also included in the differential diagnosis are other benign cystic salivary gland neoplasms like cystadenoma, intraductal papilloma and Warthin tumour. However, a mucinous background with large, mucin-filled cells and intermediate cells is an important clue to the diagnosis of MEC. On the other hand, the presence of papillary growth nature and neoplastic cells lacking intracellular mucin, absence of intermediate cells, characteristically seen in papillary-cystic variant, they tend to form “hobnail cells”, presumably after releasing their secretions into glandular spaces [10] , are features in favoring a diagnosis of ACC-PCV. In contrast, MEC requires empirical lymph node dissection, and hence this distinction has clinical correlation [9] . The cytoplasmic granules show positive reaction to Periodic Acid-Schiff (PAS) (as in current case); but are not necessary in diagnosis of ACC-PCV. In addition, the negativity of mucicarmine stain in the current case support the diagnosis of ACC. Immuno-histochemistry of non-specific DOG-1 was positive in acinar and intercalated duct cells as shown in the current case. As expected in ACC, very scant mitotic activity is seen as ki-67 (MIB-1) was estimated at 1%-2%. Another lesion that presents with cystic spaces and may contain papillary proliferation and simulate ACC-PCV is benign salivary neoplasm like cystadenoma\ cystadenocarcinoma. The cystadenoma usually lining by a double layer of flattened to columnar epithelium with multifocal oncocytic differentiation and usually there is no atypia, no increased mitoses and no invasion, which helps to exclude a cystadenocarcinoma. The lack a lymphoid stroma making a Warthin tumor much less likely. With presence numerous psammoma bodies and papillary projection the other important differential diagnosis is metastatic papillary carcinoma from thyroid, however the negativity of TTF1 immunostaining make the diagnosis is less likely. Despite low-grade nature of ACC, several reports reveal that papillary cystic variant is associated with aggressive behavior. Many studies emphasize the importance of recognizing this variant-PCV- as it has proved to be universally fatal within 10 years [2] . Local recurrence occurs in about 20% of cases. Otherwise, ACC has excellent prognosis with 90% five-year survival rate. Poor prognostic factors include large tumor size, involvement of the deep lobe of the parotid gland, and incomplete resection. Multiple recurrences, cervical lymph node, and distant metastases predict poor prognosis [8] . Total parotidectomy is considered the best modality of treatment.

4 CONCLUSION

Presence of psammoma body in salivary gland lesions is a very rare finding. It can mimic many benign or neoplastic lesions or even when simultaneously present with papillary pattern; metastatic papillary thyroid carcinoma which should be excluded. High index of suspicion of ACC-PCV should be kept in mind for any cystic lesion in salivary glands as it is very important to identify this subtype; which carries much different prognosis and treatment.

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