ORIGINAL RESEARCH PAPER

Pathology

ACINIC CELL CARCINOMA

KEY WORDS: Carcinoma Acinic cell carcinoma, Parotid gland.

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ABSTRACT

Acinic cell carcinoma (ACC) is a low-grade malignant salivary neoplasm that constitutes approximately 17% of primary salivary gland malignancies. In the head and neck region, the parotid gland is the predominant site of origin and women are usually more frequently diagnosed than men. Previous radiation exposure and familial predisposition are some of the risk factors for ACC. A slowly enlarging mass lesion in the tail of the parotid gland is the most frequent presentation. The diagnosis is usually confirmed with a fine needle aspiration biopsy, and surgical excision is the main treatment of this malignant neoplasm. Other treatment modalities such as radiotherapy may be indicated in some cases. **Conclusion:** ACC has a significant tendency to recur, to produce metastases (cervical lymph nodes and lungs), and may have an aggressive evolution. Therefore, long-term follow-up is mandatory after treatment.

INTRODUCTION

Acinic cell carcinoma (ACC) is a rare, low-grade malignant tumor of the salivary glands, with the parotid gland being the most frequent site of origin.

ACC accounts for 17% of primary salivary gland malignancies and 2–6% of all salivary gland neoplasms.

It typically occurs in the fifth to sixth decades of life, though younger patients may also be affected. Women are more frequently diagnosed than men. Risk factors for ACC include prior radiation exposure and familial predisposition.

Patients commonly present with a painless, slow-growing mass, often localized to the parotid gland.

While ACC generally follows an indolent course, it has potential for recurrence and metastasis.

Diagnosis relies on imaging and histopathological evaluation, and the primary treatment is surgical excision, with radiotherapy reserved for high-risk cases.

This case series describes the clinicopathological features of five patients in time period of 5 years diagnosed with ACC at a tertiary care center focusing on clinical presentation, diagnosis, and management.

CASE SERIES

Case 1

We, in this case report aim to throw light on an interesting case of clinically diagnosed Recurrent parotid tumor of the left parotid gland in a 35-year-old male patient. The patient presented with a slowly progressing painless swelling on the left side of the face for the past 2 years.

The swelling was initially small but had progressively increased in size without any associated symptoms such as discharge, fever, trauma, headache, or difficulty in chewing.

A positive history of tobacco chewing and smoking was noted, but he had ceased these habits 5 years ago.

The patient had no significant medical or surgical history and denied any family history of malignancy or previous surgical interventions.

Clinical Diagnosis: Recurrent parotid tumor.

Ultrasonography Findings: Hypoechoic lesion, superficial lobe

CECT Findings: Heterogeneous lesion ± lymph nodes

- FNAC (Milan system): Recurrent parotid tumor.
- Specimen: Excised Parotid Tumor.

Gross Histopathology: Received a single encapsulated glandular structure attached to skin and fibrofatty tissue. The specimen was grayish-brown, soft to firm, and measured 4.1 × 3.0 × 2.5 cm. On sectioning, white homogenous areas were noted, Multiple lymph node specimens were received, brown in color and soft-to-firm in consistency. The largest measured 6.8 x 3.5 x 2.8 cm, and the smallest was 3.8 x 0.8 x 0.8 cm. Upon sectioning, reddish-gray areas were noted.

Microscopic examination: Sections revealed skin, adnexal structures, and tumor tissue. The tumor comprised neoplastic glands arranged in a papillary configuration within a cystic space. These neoplastic glands were lined by oncocytic cells, and the individual tumor cells had abundant granular cytoplasm. A hobnail appearance was observed, with hypochromatic nuclei and indistinct nucleoli. There were very few mitotic figures, and some cysts contained mucin and debris. No lymphovascular invasion was noted.

A total of eight lymph nodes were isolated, with two showing tumor involvement, and the remaining six were free from tumor deposits.

Histopathology confirmed the diagnosis: Acinic Cell Carcinoma (Papillary Cystic Variant) with Lymph Node Metastasis.

DISCUSSION

Acinic cell carcinoma (ACC) is a rare salivary gland malignancy (1–6%), most commonly arising in the parotid gland.

This patients presented with a slow-growing, painless parotid/preauricular swelling and intact facial nerve function, closely mimicking benign lesions.

Radiological imaging (USG/CECT) suggested benign parotid tumors, highlighting the limited specificity of imaging in ACC.

FNAC findings were benign or indeterminate (Milan IV) in most cases, underscoring the diagnostic limitations of cytology.

Histopathology was diagnostic in this case, demonstrating characteristic solid and microcystic patterns with acinar and clear cell populations; lymph node metastasis was noted.

Surgical excision with facial nerve preservation achieved favorable outcomes in this case, emphasizing histopathology-guided management as the key to optimal care.

This case highlights the diagnostic challenges posed by ACC, particularly its tendency to masquerade as a benign lesion, and reinforces the importance of comprehensive clinicoradiological correlation with histopathological confirmation for accurate diagnosis and appropriate management.

Case 2
A 46-year-old female patient reported to our institution with a swelling on right side of the pre-auricular region. The patient presented with a slowly progressing painless swelling on the right side of the face since 6-8 months.

Clinical Diagnosis: Pleomorphic Adenoma.
Specimen: Right parotid gland.

Gross Histopathology: Received the specimen consisted of a globular soft tissue mass, soft to firm in consistency, reddish-brown in color, measuring 3.0 × 2.5 × 1.5 cm.
On cut section, the mass showed pink, homogeneous areas.

Microscopic examination: Multiple sections examined revealed an outer fibrous capsule enclosing tumor cells arranged in sheets and microcysts.

The tumor showed a biphasic cell population.

The predominant population consisted of acinar cells with granular, basophilic cytoplasm and round centrally placed nuclei.

The second population comprised tumor cells with clear cytoplasm and round central to eccentrically placed nuclei.

Areas of hemorrhage were noted.
Few foci of normal salivary acinar cells were also identified.

Final Diagnosis: The histopathological features were consistent with Acinic Cell Carcinoma of the Parotid Gland.

Case 3
A 28-year-old male reported to our institution with a chief complaint of swelling in the right preauricular region since 9 months. The swelling was first small and then gradually increased in size and was not associated with pain. The swelling was firm, mobile, non-tender, with a smooth surface and regular borders.

Clinical Diagnosis: Benign Parotid tumor.
Specimen: Right parotid gland.

Gross Histopathology: Received a single encapsulated glandular structure attached to fibrofatty tissue. The parotidectomy specimen a multinodular mass measuring 6.2 × 4.3 × 2.0 cm.
On cut section, the tissue was firm in consistency and showed multiple small solid nodules with yellowish to whitish areas.

Microscopic examination: Sections revealed malignant tumor islands composed of an admixture of tumor cell populations, including malignant acinar cells, clear/vacuolated cells, intercalated duct–like cells, and nonspecific glandular cells.

The tumor cells were arranged predominantly in solid and microcystic patterns, infiltrating into a sclerotic stromal background.

Malignant acinar cells were large polygonal cells with abundant basophilic granular cytoplasm and eccentrically placed small hyperchromatic nuclei.

Intercalated duct–like cells appeared cuboidal with amphophilic cytoplasm and centrally placed dark nuclei.
Nonspecific glandular cells showed vesicular nuclei with eosinophilic cytoplasm.

Sheets of tumor cells were traversed by delicate thin-walled blood vessels.

No areas of necrosis were identified.

Special Stains and Immunohistochemistry :
PAS-D staining demonstrated abundant diastase-resistant PAS-positive cytoplasmic granules within malignant acinar cells.

On immunohistochemistry, DOG-1 showed strong cytoplasmic positivity, while mammaglobin was negative in tumor cells.

Case 4
A 52-year-old female patient admitted to ourinstitution with the complaint of cheek mass over left side. The mass was present for 1 years and it was growing slowly, she had no pain complaint. In her medical history, she had no surgery from salivary glands or neck region.

On examination, a firm, mobile, non-tender swelling measuring 3.5 × 3.0 cm was noted in the left preauricular region with smooth surface and well-defined margins. Facial nerve function was intact.

Clinical Diagnosis: Benign salivary gland tumor.
Specimen: Left parotid gland.

Gross Histopathology: Received a single encapsulated glandular structure attached to fibrofatty tissue. The specimen consisted of a globular soft tissue mass measuring 3.8 × 3.2 × 1.6 cm, soft to firm in consistency. The cut surface revealed homogeneous gray-white areas.

Microscopic examination: Sections showed a well-circumscribed tumor composed of cells arranged in solid sheets and microcystic patterns. The tumor exhibited a biphasic population, predominantly acinar cells with granular basophilic cytoplasm and centrally placed nuclei, along with clear cells. Focal hemorrhage was present. No perineural or lymphovascular invasion was identified.

Final Diagnosis: Acinic Cell Carcinoma of the Parotid Gland

Case 5
A 40-year-old male patient reported to our institution with a chief complaint of swelling in his right parotid gland region for 8 month. The swelling was initially small and gradually increased within 8 year. The growth was painless, solitary, well-delineated, intermittent, and slow in nature. There was no significant medical history, radiation exposure, or family

history of malignancy. On local examination, a firm, non-tender, mobile mass measuring 4.2 × 3.5 cm was palpated in the right parotid region with intact overlying skin.

Clinical Diagnosis: Benign parotid tumor.
Specimen: Right parotid gland.

Gross Histopathology: A multinodular mass measuring 4.5 × 3.8 × 2.0 cm was received. On cut section, the tumor appeared solid, gray-white to yellowish, with focal cystic changes.

Microscopic examination: Sections revealed tumor cells arranged in solid, microcystic, and focal papillary patterns. The tumor consisted of malignant acinar cells with abundant basophilic granular cytoplasm, along with clear cells and intercalated duct-like cells. The stroma was mildly fibrotic. No necrosis was identified.

Final Diagnosis: Acinic Cell Carcinoma of the Parotid Gland

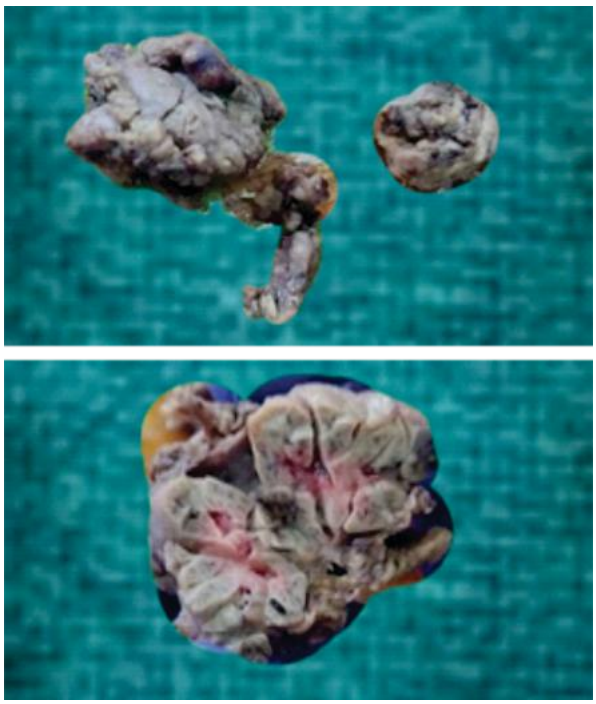
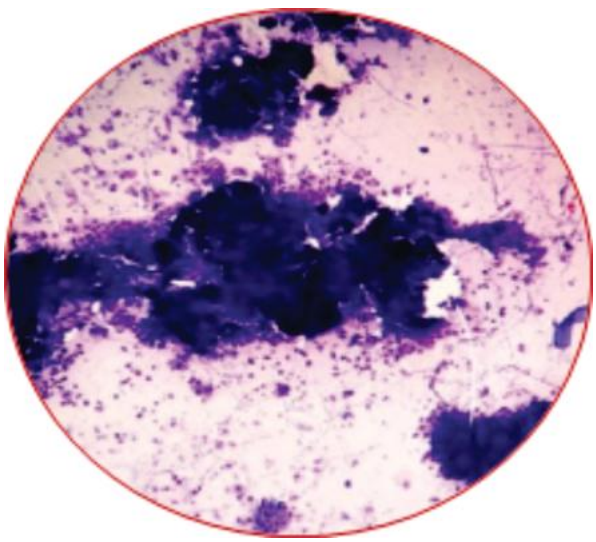
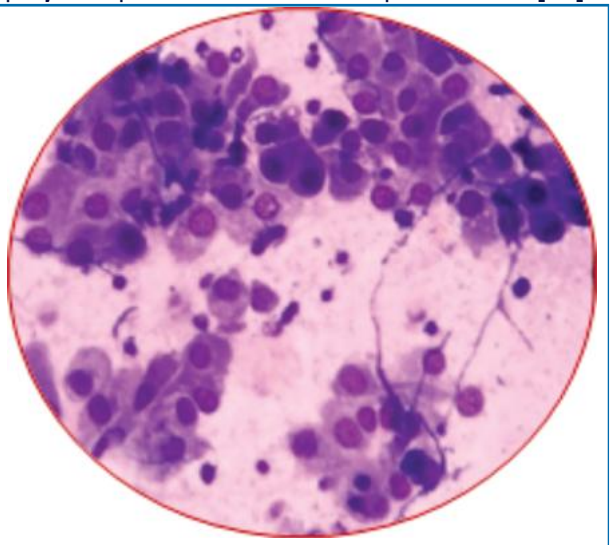


Figure 1. Gross Histopathology Specimens



A)
40x Low Power Magnification



B)
100x high power magnification

Figure 2. Histopathology showing A) salivary acinar cells with inflammatory cells (×40) & B) salivary acinar cells with few lymphocytes (×100) (H&E stain)

CONCLUSION

In conclusion, Acinic Cell Carcinoma (ACC), particularly the papillary-cystic variant (PCV-ACC), remains a challenging diagnosis due to overlapping clinical, cytological, and histological features.

Despite ACC's general association with a relatively favorable prognosis when diagnosed early and treated appropriately, the prognosis of PCV-ACC is poor, with many cases being fatal within 10 years.

Timely and accurate diagnosis, primarily through histopathological examination, is crucial for guiding treatment decisions.

While complete surgical excision remains the standard treatment, the unique clinical course of PCV-ACC necessitates further research to improve understanding, prognostication, and management strategies for this rare malignancy.

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