



## RESEARCH ARTICLE

### ACINIC CELL CARCINOMA OF PAROTID GLAND- A CASE REPORT

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#### ABSTRACT

The Acinic cell carcinoma represents about 1% of all salivary gland tumors. Between 90 and 95% of these tumors are found in the parotid gland. Clinically if the site is parotid gland, it may resemble a pleomorphic adenoma. Some of these tumors metastasize or recur and cause death, it is generally agreed that acinic cell adenocarcinoma should be considered a low-grade malignancy. We report a case of acinic cell carcinoma involving the parotid gland which had a history of more than 5 years. It presented as a huge mass involving the right parotid gland with no history of pain or discomfort. The parotid mass was surgically excised and histopathologically proven to be acinic cell carcinoma.

## INTRODUCTION

Salivary gland (SG) tumors represent 3% of all neoplasms of the head and neck (Acinic Cell Carcinoma of Parotid Gland, 2010). Acinic cell carcinoma is a malignant epithelial neoplasm in which the neoplastic cells express serous acinar differentiation<sup>2,3</sup>. Acinic cell carcinoma represents about 1% of all salivary gland tumors. Between 90 and 95% of these tumors are found in the parotid gland<sup>4</sup>. Clinically if the site is parotid gland, it may resemble a pleomorphic adenoma (Sai Krishna and Sivapathasundharam, 2003). Some of these tumors metastasize or recur and cause death, hence it is generally agreed that acinic cell adenocarcinoma should be considered a low-grade malignancy<sup>3</sup>. This tumor occurs with a higher frequency in women and is usually found in the 5<sup>th</sup> decade of life<sup>4</sup>. This malignant tumor has an aggressive course and carries a very poor prognosis. Local and distant metastases are common<sup>4</sup>.

## Case Report

A 54 year old male patient reported to the department of oral medicine and radiology with the chief complaint of swelling in the right cheek region for the past 5 years. It started initially as a small swelling which gradually increased in size to attain the present size. No pain or discomfort was present. The past medical history revealed that the patient has hemiplegia of right hand and leg. Patient gives a history of head injury before 5 years and was hospitalised for 10 days. The past dental history and family history were not relevant. The patient has mixed diet and had a habit of pan chewing (paanparak) for 5 years but has quit the habit for the past 5 years. Patient is an alcoholic for past 5 years. On general examination, the patient is moderately built and moderately nourished. His vital signs were normal. CNS examinations were revealed slurred speech and loss of memories. Musculoskeletal system examination revealed hemiplegia and limping gait. Extra-oral examination revealed facial Asymmetry due to the swelling present on the right middle and the lower third of the face. On inspection, a single large round well circumscribed swelling was present on the right side of the face measuring approximately 8.5 X 8.0 cm extending superiorly till the tragus of the right ear, inferiorly 1 cm below the lower border of the mandible,

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anteriorly 5cm away from the corner of the mouth and posteriorly extends 1cm behind the ear, lifting the ear lobe. The skin over the swelling appears slightly shiny and stretched. Skin was pinchable over the swelling. The region surrounding the swelling appeared normal and no secondary changes were seen over the swelling. On palpation, all inspeactory findings such as site, size, shape and extent were confirmed. The swelling was soft to firm in consistency, lobulated and non-tender. It was freely mobile on all planes, slightly warm on palpation and no paresthesia was elicited. The swelling lifted the ear lobe on right side and restricted mouth opening was present. Intra oral examination revealed generalized chronic periodontitis and decreased salivary flow rate on the opening of stenson's duct on right side.

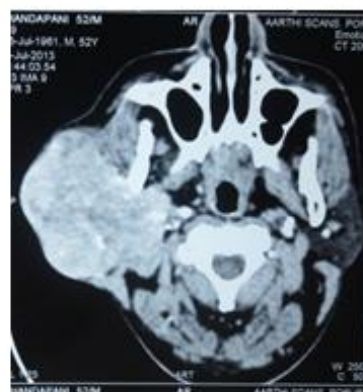
Lymph node examination revealed, 2 oval lymph nodes measuring approximately 2 X 2.5cm palpable on right submandibular region which was non tender and firm in consistency. One node was freely mobile in all planes and another was fixed to the swelling. On the basis of history and clinical examination, a provisional diagnosis of benign tumor of parotid gland was made. Further, radiographic examination was performed. OPG revealed missing 14, 15, 16, 24, 25, 26, 28, 36. Generalised bone loss was present suggestive of generalised chronic periodontitis. Ultrasonography of the swelling was not able to perform as the huge swelling did not allow the penetration of ultrasound waves deep enough into the lesion. Contrast CT of head and neck revealed, 9.2 X 7.5 X 7.3cm size large well-defined soft tissue density mass lesion

### Acinic Cell Adenocarcinoma of Parotid Gland

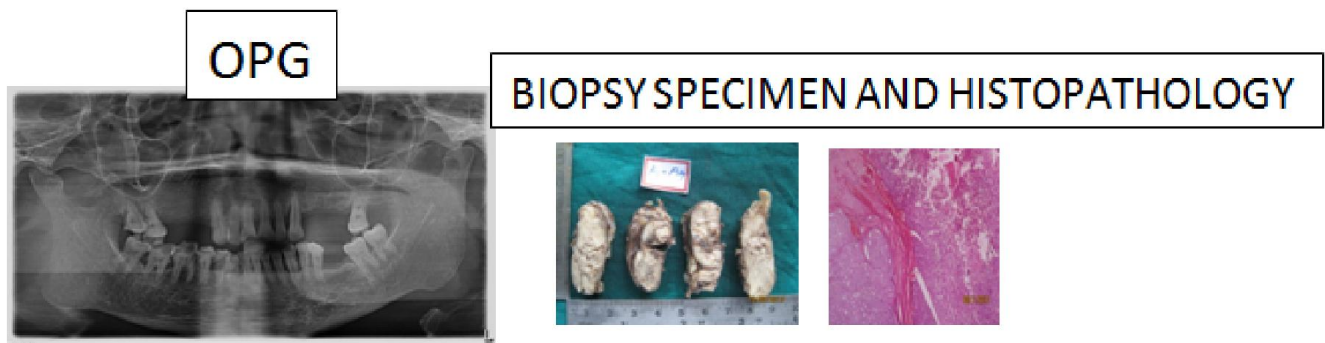
#### PATIENT PROFILE



#### INTRA-ORAL



CT



noted involving the entire superficial and deep lobe of right parotid gland. On iv contrast the lesion shows good enhancement with contrast and some non-enhancing areas within it suggestive of pleomorphic adenoma. Total parotidectomy was performed under general anesthesia on the right side preserving the facial nerve. The specimen was submitted for histopathological analysis and final diagnosis of acinic cell adenocarcinoma was given.

## DISCUSSION

Acinic cell adenocarcinoma (ACC) is a malignant salivary gland tumour with cells showing serous acinar differentiation (Sai Krishna and Sivapathasundharam, 2003)<sup>2</sup>. Interestingly, ACC was originally described as an adenoma, but from the 1950s the term acinic cell adenocarcinoma was coined when its ability to metastasize and recur locally had been recognized. It is hard to diagnose ACC of parotid gland preoperatively (Wonjae Cha et al., 2011). This tumour was first recognized by Nasse in 1982, who described 4 parotid adenomas that closely resembled normal acinar cells<sup>2</sup>. In 1953, Buxton and his groups ascribed the malignant character to many of these tumors<sup>15</sup>. ACC closely resembles pleomorphic adenoma in gross appearance, tending to be encapsulated and lobulated. Around 85% of ACC occurs in parotid gland. It may also occur involving other major glands and the accessory intraoral glands (Sai Krishna and Sivapathasundharam, 2003)<sup>7</sup>. These lesions often presents as slow growing masses<sup>4</sup>. Patients usually visit doctors with the symptom of incidental infra-auricular mass. Facial palsy or pain usually may not exist at the first time (Wonjae et al., 2011). In our case, the tumor appeared as a painless slow growing mass involving both the superficial and deep lobe of parotid in the infra-auricular region. Facial palsy was not present in our case<sup>6</sup>. Bilateral involvement of the parotid gland is reported in 3% of cases (John S. Clarke, 1969; Depowski, 1999).<sup>2,4,9</sup>. The pathogenesis of acinic cell carcinoma is unknown. The neoplasm is postulated to arise from the intercalated duct or the terminal epithelial cluster and tubule. Cyto genetic alterations have been identified in malignant salivary gland tumors, including structural rearrangements of the long arm of chromosome 6, loss of chromosome Y, and gain of chromosome 8. Studies of acinic cell carcinoma have found common deviations, including 6q rearrangements, loss of chromosome Y, and gains of chromosome 7 and 8. However, acinic cell carcinoma may have a normal karyotype as noted by Mark et al. (Depowski et al., 1999). ACC usually occurs in the fifth and sixth decades of life and our patient was in the 5<sup>th</sup> decade.

The tumor occurs with a higher frequency in females with a female to male ratio of 1.5:1<sup>2,3,4,6</sup>. (Nabil N Al-Zaher, 2011). Acinic cell carcinoma constitutes approximately 17% of primary salivary gland malignancies, representing the third most common epithelial malignancy of the salivary glands in adults, and in the pediatrics age group, it is considered to be the second most common epithelial salivary malignancy after mucoepidermoid carcinoma<sup>2</sup>. (Nabil N Al-Zaher, 2011). It behaves most similar to low-grade mucoepidermoid carcinoma<sup>16</sup>. Acinic cell carcinomas can be categorized as conventional and dedifferentiated neoplasms. The conventional type can be graded in 3 tiers. Low-grade tumors are broadly interpreted as those most closely resembling the architecture of a normal salivary acini. They are encapsulated and measure less than 3 cm. The intermediate tumors exhibit intercalated duct differentiation, lack capsule, and measure between 3 and 6 cm. High-grade tumors are poorly differentiated, resemble acini in the early phases of embryonic development, and are highly infiltrative. Dedifferentiated ACCs are composed of conventional lowgrade ACC and areas of differentiated, high-grade adenocarcinoma or undifferentiated carcinoma<sup>12,13</sup>.

Acinic cell carcinomas have variable gross features tending to be gray-tan and firm to soft with solid and/or cystic areas. A capsule may be present. The lesion may have a multi-nodular appearance<sup>4,11</sup>. Over time, various authors have used varied terms to describe the several histomorphologic patterns and cellular features of these tumors. Thus, Batsakis JG *et al.* have described seven different histologic patterns for ACC: Acinar-lobular, microcystic, follicular, papillary cystic, medullary, ducto-glandular and primitive tubular, while Ellis GL and Corio RL recognized only three categories: cystic papillary, follicular, and solid. However, descriptive categories (solid, microcystic, papillary-cystic and follicular) presented by Abrams *et al.* in 1965 have been useful to pathologists over the last 30 years and are still applicable today. We must keep in mind that these designations related to the tumor pattern do not define specific subtypes of ACC for clinical, therapeutic, or prognostic purposes. Most studies indicate solid and microcystic patterns as the major histomorphologic patterns for ACC<sup>2,7</sup>. The most characteristic cell has the features of serous acinar cells, with abundant granular basophilic cytoplasm and a round darkly stained eccentric nucleus. Other cells are the intercalated duct like cells, which are smaller and the vacuolated cells which seem to be unique to acinic cell carcinomas among salivary gland neoplasm. Connective tissue stroma is delicately fibrovascular collagenous tissue. Lymphoid cells are commonly seen in the parotid acinic cell carcinoma

<sup>2,4,5</sup>. Other histologic variants like dedifferentiated, oncocytic, hybrid tumours and well differentiated with lymphoid stroma have also been reported by various authors (Sai Krishna and Sivapathasundharam, 2003). Pre-operative investigations commonly include FNAC, ultrasonography, CT, MRI, sialography and scintigraphy. In many cases it is not possible to determine whether the tumor is benign or malignant using radiological investigations. However, features such as ill-defined margins, invasion, of adjacent soft tissues (such as fat spaces), and destruction of adjacent osseous structure are considered to be typical of malignancy<sup>16,17</sup>. The recommended treatment of acinic cell carcinoma of the parotid is total parotidectomy with preservation of the facial nerve. Infiltration of the nerve or perineural structures may demand its sacrifice and the deficits can be restored with nerve grafting. Neck dissection is best reserved for patients with regional lymphadenopathy although some advocate its routine practice. The results of radiotherapy are inconsistent and the possibility of probable long-term sequelae cannot be overlooked. Irradiation seems therefore most appropriate in locally advanced disease or in high grade tumours. Operative morbidity includes Frey's syndrome and facial numbness. Facial weakness due to nerve injury is seen which was observed in our case due to marginal mandibular nerve sacrifice intraoperatively. Five-year survival rates range from 89% to 96% but fall to 56% at 20 years. Therefore, surveillance must continue for a long time. Poor prognostic feature included pain or fixation; gross invasion; and microscopic features of desmoplasia, atypia, or increased mitotic activity<sup>2,4,12,14</sup>. Local and regional recurrences occur in 34% of cases. Distant metastatic disease is rare and most often arises in the lung and bone. Brain metastasis has also been reported in parotid acinic cell carcinoma<sup>15</sup>.

## Conclusion

In conclusions, parotid ACC is a rare malignancy of head and neck region (Acinic Cell Carcinoma of Parotid Gland, 2010). This malignant tumor has an aggressive course and carries a very poor prognosis. If the condition is appropriately dealt with from the beginning, its morbidity is extremely low and permanent cure is highly probable<sup>14</sup>.

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