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ACINIC CELL CARCINOMA OF PAROTID GLAND- A RARE CASE REPORT.

[1*]Dr. K. A. Kamala*, [2]S. Sankethguddad., [3]S. G. Sujith, [4]Dr. Praveena Tantradi

[1]Associate Professor, Department of Oral Medicine and Radiology. School of Dental Sciences, KIMSDU, Karad, Maharastra, India.
[2]Post Graduate Student, Department of Periodontology, School of Dental Sciences Karad, Maharashtra, India.
[3]General Practitioner.
[4]Associate Professor. Department of OraL Medicine` and Radioloey. Maratha Mandal's Nathajirao G. Halgekar Institute of Dental Sciences and Research Centre, Bauxite Road Belgaum. Karnataka, India.

***Author for Correspondence: Dr. K. A. Kamala**
Associate Professor, Department of Oral Medicine and Radiology. School of Dental Sciences, KIMSDU, Karad, Maharastra, India.

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ABSTRACT
Acinic cell carcinoma is a low-grade malignant salivary neoplasm that constitutes approximately 17% of primary salivary gland malignancies. The literature available in this particular section is very scant in data. It is therefore prime impotent to conduct exact studies in as many groups of acinic cell carcinoma patients as possible to increase knowledge about the different clinical manifestations, optimal diagnosis, and improved therapeutic modalities for this tumor. Here we report a case of acinic cell carcinoma of parotid gland in 48 year old male patient.

KEYWORDS: Acinic cell carcinoma, parotid gland, salivary gland, clinical features, treatment.

INTRODUCTION
Acinic cell carcinoma (ACC) is a rare malignant epithelial neoplasm of salivary gland. The tumor comprises approximately 7% to 15% of all malignant tumors arising in major salivary glands. Majority of ACCs (almost 80%) occur in parotid gland, about 4% develop in the submandibular gland, and less than 1% arise in the sublingual gland. Approximately 13% to 17% involve the intraoral minor salivary glands. In the minor salivary glands ACC develops most frequently in the buccal mucosa, upper lip, and palate and rarely in the tongue, gum, paranasal sinus and larynx.[1]

CASE REPORT
A -48- year old male presented with a gradually increasing left upper third face swelling for 2 year, which had suddenly increased in size in the past 6 month [Figure 1]. He denied any loss of weight or similar swelling elsewhere in the body. Initially, the patient did not seek medical attention, but because of the progressive increase of the size of lesion, he obtained a referral to a head and neck surgeon. Local examination revealed a round, well defined, firm, non-pulsatile swelling fixed to the underlying structures measuring 5 × 5 centimeter (cm) located in pre-auricular region extending posteriorly behind the left ear with elevation of ear lobule. The overlying skin was normal and free from the swelling with no increase in temperature. There were no palpable cervical or supra-clavicular lymph nodes. Intra oral examination [Figure 2] revealed normal mucosa with generalized attrition of teeth. Depending on the history and the clinical examination, we arrived at a provisional diagnosis of pleomorphic adenoma of parotid gland with differential diagnosis of a mucoepidermoid carcinama and acinic cell carcinoma. Radiographic examination (orthopantamograph and lateral oblique) revealed a radiopaque shadow at the angle extending 1cm below the lower border of mandible [Figure 3]. Fine needle aspiration cytology (FNAC) suggested it to be a pleomorphic adenoma. The patient was managed surgically [Figure 4] in the form of a left superficial parotidectomy with preservation of the ipsilateral facial nerve. The post operative period was uneventful. Surgically excised section was sent to histopathological examination [Figure 4] which revealed a highly cellular tumor comprised of small and large cells arranged in prominent organoid pattern, nests, trabeculae and sheets. The individual cell had a centrally placed, round nucleus with coarsely stippled nuclear chromatin. The cells displayed moderate to abundant amount of deeply eosinophilic and finely granular cytoplasm with well defined cytoplasmic borders with both typical and atypical mitoses throughout the tumor. Based on the histomorphological features a diagnosis of Acinic cell carcinoma (ACC) was made. Patient was followed up subsequently after every three months for two years, with no clinical evidence of recurrence of the tumor and a good healing of the surgical wound.

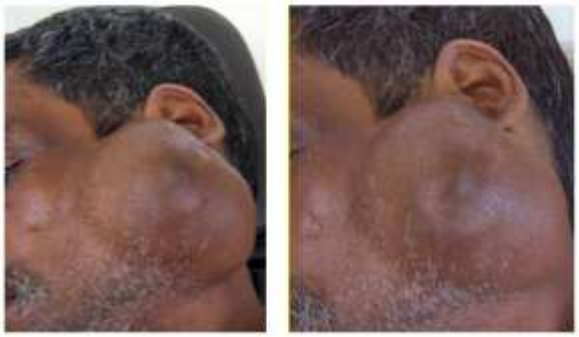

Figure 1. Extra oral view showing large swelling in preauricular area.

Figure 2. Intraoral view showing the severe attision of teeth.

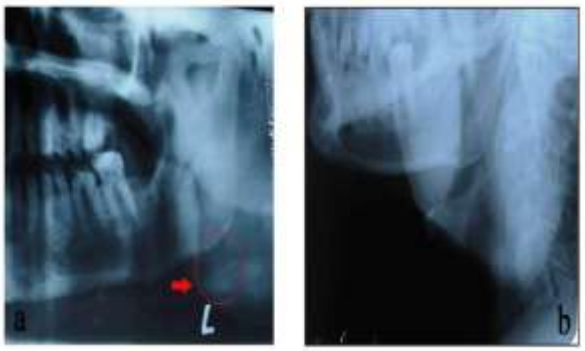

Figure 3. Cropped orthopantamograph (a) and lateral oblique (b) showing radiopaque shadow on left angle extending 1 cm below the lower border of mandible.

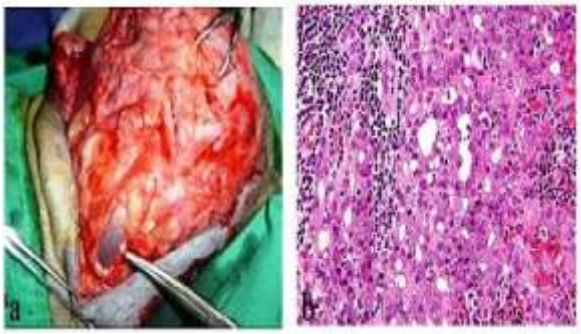

Figure 4: Intraoperative view (a) and photomicrograph (H & E 40X) showing histopathologic feartures of ACC.

DISCUSSION

Nesse first described ACC in 1892, later in 1953 it was recognized as an uncommon salivary gland tumor by Foot and Frazell. The term "acinic cell carcinoma" was established by the World Health Organization in 1992 due to its ability to metastasize. Possible causes of ACC include previous radiation exposure and familial predisposition.[2]

Women are more apt to have this malignant neoplasm, and endogenous hormones have been reported in normal and neoplastic salivary glands, but some of the results have been conflicting. Estrogen and progesterone receptors have been reported in a minority of cases of ACC, mucoepidermoid carcinoma, and salivary duct carcinoma.[3]

Al-Zaher studied ACC in 25 year old pregnant woman and he concluded that salivary gland neoplasm, including ACC might be hormonally dependent, like breast carcinoma.[4] The genetic alterations linked to ACC of the parotid gland include alterations at chromosomes 4p, 5q, 6p, and 17p, suggesting the association of tumor-suppressor genes with the oncogenesis of these tumors.[5] Women are usually more frequently diagnosed (58.8%) than men (41.2%), and according to the National Cancer Data Base Report on cancer of the head and neck, with male-to-female ratio is 1:2 to 1:3.[4] Our patient was a 48 year old male.

ACC typically develops as painless, firm, mobile, solitary, slowly growing mass. Most of the patients report duration of symptoms before diagnosis of less than one year, but cases have been reported in which the age of the tumor was more than ten years. Most of ACC are 1 to 3 cm in dimension, but in some cases can be greater than 10 cm. They are usually circumscribed, but some are ill-defined with irregular peripheries. ACC vary from firm to soft and solid to cystic consistency.[1] Parotid ACC typically presents with a slowly enlarging mass in the parotid region. About 34.33% to 50.75% were palpated in the tail of the parotid gland. Pain and facial nerve palsy were seldom reported.[1,4] Our patient had a gradually increasing painless swelling with rapid increase in size of recent onset without clinical evidence of facial nerve involvement.

The diagnosis of ACCs frequently presents difficulties, owing to its great radiologic and cytologic similarity with benign tumors and with the normal acinar component of the salivary gland, respectively. Fine-needle aspiration cytology (FNAC) has been well established in the diagnosis of salivary gland lesions. This method is highly sensitive in its diagnostic efficacy as it provides essential information on the diagnostic and therapeutic management of these tumors. The cytologic findings in FNACs of ACCs are usually characterized by acinar differentiated tumor cells and by certain cytoarchitectural patterns.[6] In addition to FNAC and other ancillary diagnostic tests, imaging studies are

usually used in the pretreatment assessment and management planning of ACC. These includes ultrasonography (to differentiate solid masses from cystic masses), computed tomography (CT), magnetic resonance imaging (MRI), and nuclear scans.[7] Christe *et al* [8] cunducted MRI study on 84 patients with parotid tumors, analyzed the parameters such as signal intensity, contrast enhancement, lesion margins (well-defined versus ill-defined), lesion location (deep/superficial lobe), growth pattern (focal, multifocal, or diffuse), and extension into neighboring structures, perineural spread, and lymph-adenopathy. He concluded that low signal intensity on T2-weighted images and postcontrast ill-defined margins of a parotid tumor are highly suggestive of malignancy. ACC is hostologically defined as a tumor with predominant differentiation toward serous acinar cells, variably admixed with clear, vacuolated, and intercalated ductal cells. The morphological growth patterns are: solid, microcystic ones are the most common patterns. Acinar cells are large, polygonal cells with slightly basophilic, granular, cytoplasm and round, eccentric nuclei. Immunohistochemically, the tumor cells express Cytokeratin (especially low molecular weight), Carcinoembryonic antigen and amylase. Large tumors and unclear surgical margins indicate poor prognosis.[9]\ In general, management of ACC consists of complete surgical removal of the tumor, by total or subtotal parotidectomy, and postoperative radiotherapy may sometimes be indicated.[3] Other treatment modalities such as radiotherapy may be indicated in some cases. Buiret *et al*[10] treated a case of ACC of parotid gland with invasion of the skull base by external 3D conformational radiation and he concluded that management of the ACC of the parotid gland is surgical with possible secondary radiation therapy. Exclusive external radiation therapy is, however, an option in case of contra-indication for surgery or patient refusal. Postoperative adjuvant treatment in advanced-stage patients or those with positive resection margins usually gives satisfactory control of the disease. The overall five-year disease-specific survival is estimated to be around 91%, and 88% at ten years.[1,4] Lin *et al* [11] conducted retrospective analytic study in 25 patients of ACC and concluded that, surgery and postoperative radiotherapy has ten year disease- free with 84% overall survival rate. 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.[12]

CONCLUSION

ACC of the parotid gland is a rare malignancy that has features of less aggressive behaviour, and good prognosis. Management of acinic cell carcinoma of the parotid gland is surgical with possible secondary radiation therapy. The literature available in this particular section is very scant in data. It is therefore prime impotent to conduct exact studies in as many groups of acinic cell carcinoma patients as possible to increase knowledge about the different clinical manifestations, optimal diagnosis, and improved therapeutic modalities for this tumor.

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