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Epithelial-myoepithelial Carcinoma of the Parotid Gland : Report of a Case and Review of the Literature

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Epithelial-myoepithelial carcinoma(EMC) is a rare salivary gland tumor with relatively low grade malignant potential and rarely metastasizes into the distant organ. Prior to 1991 this neoplasm was not classified as a malignant neoplasm.

There are many confusions in terms of making diagnosis of EMC, probably due to a possibility of bidirectional proliferation and differentiation of intercalated duct cells.

The author reports a case of EMC and reviews literature in order to search optimum treatment for this malignant neoplasm relatively newly classified by the World Health Organization in 1991.

Key Words: Epithelial-myoepithelial carcinoma, Parotid gland

Epithelial myoepithelial carcinoma(EMC) of intercalated duct origin is a relatively rare low grade salivary gland carcinoma with high recurrence rate. It also rarely metastasizes into the brain[1], kidney[2], & lung[3]. Analogous lesions of this tumor can occur in the breast and more rarely arise from adnexae of the skin[4]. In 1972 Donath and coworkers described the distinctive group of salivary gland neoplasm with a clear cell component and designated it as the epithelial-myoepithelial carcinoma of intercalated ducts[5]. This neoplasm was only illustrated as "clear cell monomorphic adenoma" in the 1972 WHO publication on histopathologic typing of salivary gland tumors[6]. It was not until 1991 that this carcinoma was established by the World Health Organization[7] as a distinct diagnostic entity. The WHO defined the EMC as a tumor composed of variable proportions of two cell types, an inner layer of duct-lining cells and an outer layer of clear cells, which typically form double-layered duct-like structure.

The outer clear myoepithelial cells vary in shape from columnar to ovoid and have well-defined cell borders, and eccentrically located vesicular nuclei are located toward the basement membrane. Although mitotic figures are rarely seen, it is not unusual for tumor cells to exhibit vascular invasion and neurotropism[2].

EMC of intercalated duct accounts for less than 1% of parotid tumors and occurs in patient between the ages of 50 and 70 years. According to Lack and Upton, this tumor can occur during childhood; they described two congenital parotid salivary gland tumors that resembled EMC[8].

This author reports a case of locally recurred epithelial-myoepithelial carcinoma of the parotid gland, and discuss the treatment of this rare tumor.

CASE REPORT

A 52 year old female presented with recurrent palpable multiple tumor masses just anterior to the left parotid gland in about 2 years after initial excision of tumor at a local clinic. The patient stated that the surgeon initially regarded it as a epidermal inclusion cyst. Otherwise, the original condition and appearance of the tumor was not clear. The tumor was palpable just anterior to the parotid gland at the anterior border of the masseter muscle. It appeared to contain multiple masses. Patient did not have clinical symptoms of facial nerve involvement.

Computerized tomography (CT) scan of this area showed a cystic tumor mass situated outside of the left masseter

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Fig. 1. Photomicrograph showing typical double layered duct like structure of E.M.C (H & E stain, ×100).

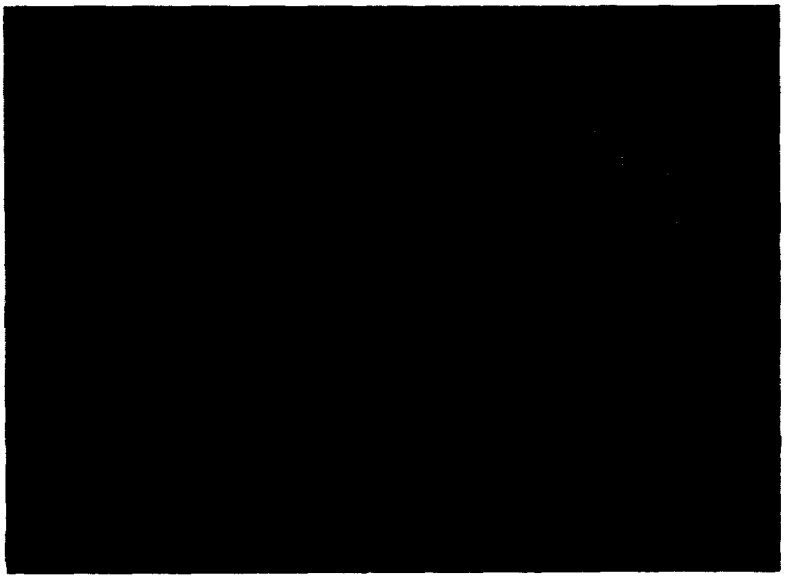

Fig. 2. Higher magnification of E.M.C showing inner layer of duct lining cells and outer layer of clear cell (H & E stain, ×400).

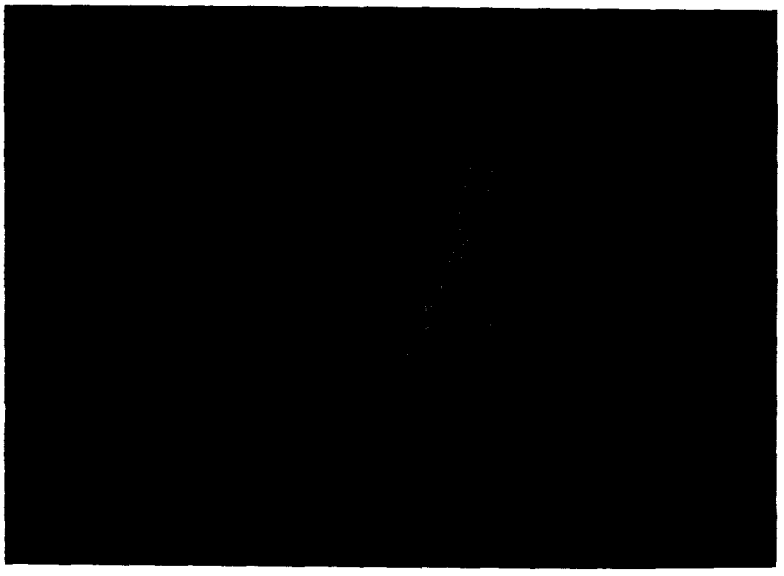

Fig. 3. Photomicrograph showing typical double layered duct like structure next to the nerve (H & E stain, ×40).

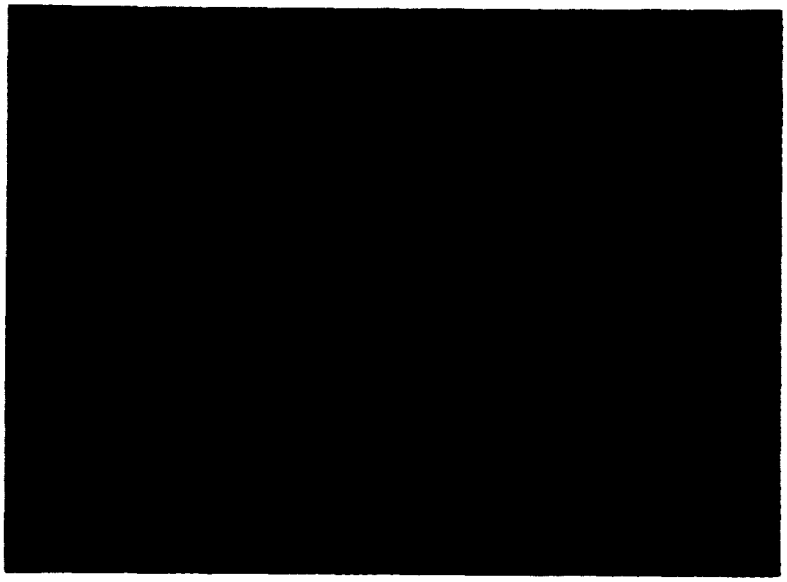

Fig. 4. Photomicrograph showing dense fibrous connective tissue separating tumor (H & E stain, ×40).

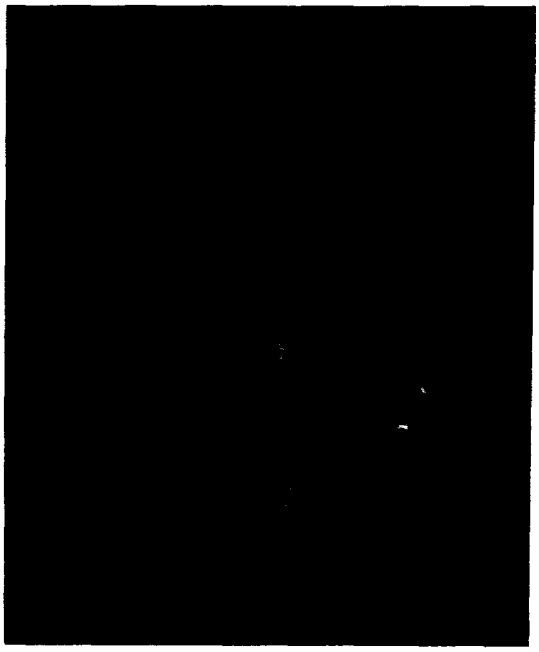

Fig. 5. Clinical photograph showing incisional scar and multiple nodular masses.

muscle. Since this mass appeared to be clinically benign, the tumor was excised through the previous vertical incisional scar. This tumor was multilobulated and adherent to one of the buccal branches of the facial nerve. The entire tumor mass including a branch of the facial nerve was excised. The tumor was very close to the masseter muscle. Histopathology showed the typical findings of EMC intercalated duct origin. It showed a multinodular growth pattern with islands of tumor

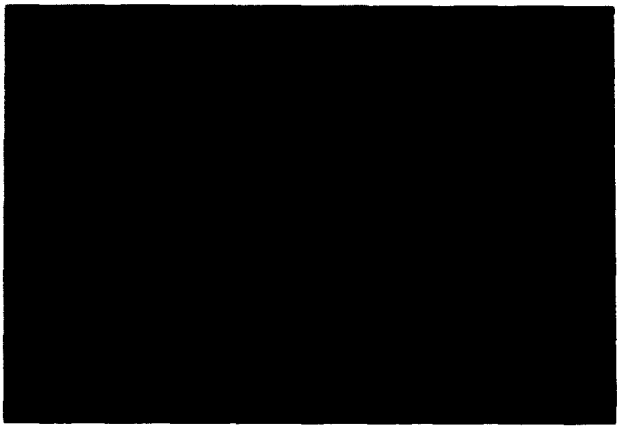

Fig. 6. Multiple nodular recurrent epithelial-myoepithelial carcinoma.

cells separated by dense branches of fibrous connective tissue and double layered tubular cribriform pattern.

The islands of tumor cells were composed of two cell types, an inner layer of duct-lining luminal cuboidal cells with fine granular, dense, eosinophilic cytoplasm and central or basally located nuclei, and an outer layer of clear cells varing in shape from columnar to ovoid, having well-defined cell borders and eccentrically located vesicular nuclei localizing toward the basement membrane.

The PAS stain was positive for the cytoplasm of the clear cells. The central area of necrosis and neural invasion was observed in this tumor(Fig. 3, 4).

Although rare, the EMC of intercalated duct origin has

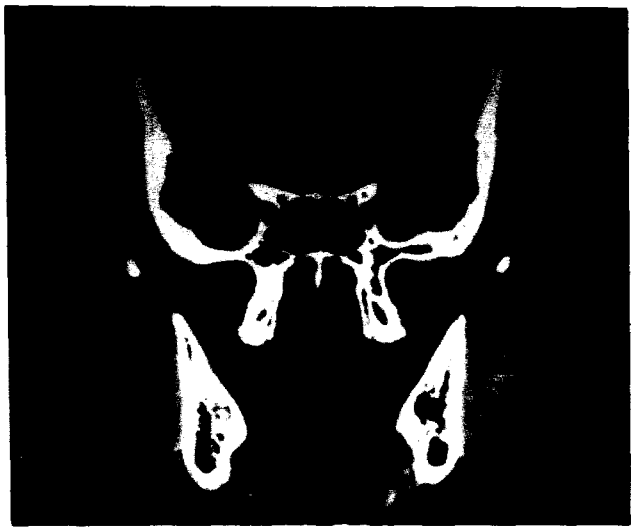

Fig. 7. Computerized tomography showing cystic component of tumor just outside of the masseter muscle, left.

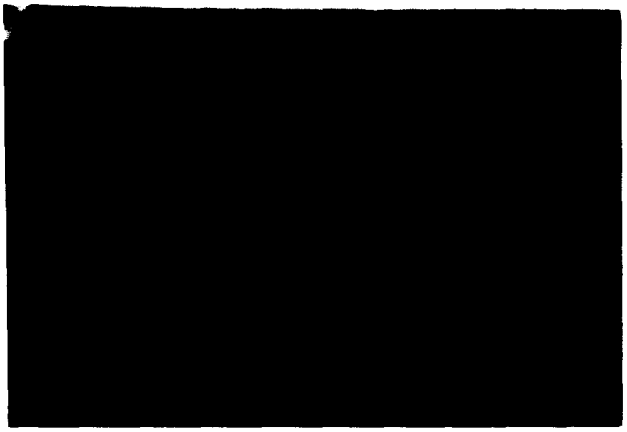

Fig. 8. After total parotidectomy with preservation of facial nerve.

reportedly potentiality of metastasizing into the brain, lung and kidney. The screening test including lung scan, liver function test, kidney scan, and neck CT scan were reported to be negative for possible metastasis.

This patient underwent total parotidectomy with preservation of the facial nerve. The final pathology report failed to show any residual carcinoma in the surgical parotid gland specimen. Six months after the total parotidectomy, the patient does not show any evidence of local recurrence nor distant metastasis.

DISCUSSION

The EMC is a very rare carcinoma and it comprises less than 1% of salivary gland tumor. Inspite of the histologic benign appearance, it is classified as a carcinoma because of the clinical behavior of local recurrence and distant metastasis.

Even though Donath and coworker first used in 1972 the term EMC of intercalated ducts, many other workers used other terminology such as clear cell carcinoma[9], glycogen-rich adenocarcinoma[10], parotid clear cell adenoma[11], monomorphic clear cell tumor[12], which might have affected the treatment of EMC of parotid gland.

Toida et al[13] stated that 97 cases of EMC have been reported in the world literature since the initial report by Donath et al, however, it is not clear how they reached to the number of reported EMC prior to 1991. Some of the reported cases showed a great confusion in the course of making diagnosis of EMC, from adenocystic carcinoma to pleomorphic adenoma finally to EMC of intercalated duct[14]. In one case, the final diagnosis of EMC was reached in 1989 after an episode of lung metastasis from benign mixed tumor in 1975 and recurrent malignant mixed tumor in 1976[15]. It is most likely not that accurate in terms of reported cases of EMC in the literature because of relatively recent establishment of WHO classification as malignant neoplasm.

Previously, this tumor was not classified as a carcinoma and many of these neoplasm were probably treated as benign. It is found in the literature that some cases showed very aggressive nature while others ran relatively benign course. Pathologically typical double layered tubular pattern usually does not give much problem in making diagnosis whereas solid type tumor usually require multiple histochemical and ultastructural investigation for differential diagnosis.

By the use of proliferating cell nuclear antigen(PCNA) immunohistochemistry, Fonesea[16] suggested that it is probably this solid type of EMC which is more aggressive and attributable to distant metastasis.

Very few reports in the literature clearly discussed about the optimum treatment of this carcinoma; many discuss the detail of making diagnosis. Prior to 1991, it was obvious that most of these cases were treated as benign. After its malignant classification, some workers suggested a total parotidectomy with facial nerve preservation with or without radiation

therapy. The role of radiation therapy in this type of neoplasm is still controversial. In the present case, the patient did receive total parotidectomy with facial nerve preservation because of the recurrent nature of tumor and histopathologic evidence of close proximity to the nerve, eventhough pathologic finding clearly showed less aggressive tubular-cribriform type.

This author was not able to find any extensive analytic report on the treatment of EMC. Some authors proposed that proliferation and differentiation of intercalated duct reserve cells which can be bidirectional leading to cells with myoepithelial or epithelial differentiation[17], indicating even with the tumor shows less aggressive tubular-cribriform type can change to more aggressive solid type of neoplasm. Probably, it is dangerous to assume that tubular-cribriform types of EMC requires less aggressive therapy than solid type. In general, wide surgical excision with clear margin is the recommended treatment due to the tumor's propensity to infiltration. Parotidectomy is also recommended, but it is not clear whether superficial parotidectimy is sufficient or total parotidectomy with resection of facial nerve followed by radiation therapy should be done; some cases showed distant metastasis even after parotidectomy. It is, therefore, suggested that one should review treatment of all the reported cases of EMC to obtain any guidance of optimum treatment. We will probably have a difficulty to finding optimum treatment, simply because this particular neoplasm was not classified as a malignant tumor by the WHO prior to 1991.

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