GIANT EPITHELIAL MYOEPITHELIAL CARCINOMA OF PAROTID GLAND- A CASE REPORT.

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Abstract:
Epithelial myoepithelial carcinoma is a rare salivary gland tumor consisting of inner duct lining epithelial cells and outer clear myoepithelial cells, which is characteristic composition of biphasic tubular structure. It was first described by Donath et al in 1972 and classified as a rare low grade biphasic neoplasm of salivary glands. In 1991, WHO recognized this tumor as a distinct entity and subtype of salivary gland adenocarcinoma which became a part of new classification system. Morphological features and immunohistochemistry distinguish the two cell types of this tumor. This is a case report of epithelial myoepithelial carcinoma of parotid gland in a 46 year old male patient.


INTRODUCTION:
Epithelial myoepithelial carcinoma of the salivary glands was first described by Donath et al in 1972. Later in 1991, World Health Organization (ICD-O-code - 8562/3) classify this under low grade malignant epithelial tumor of salivary gland. It is a rare tumor accounting for 0.4 - 1% of all primary salivary gland neoplasms. Most commonly 80% arises in parotid gland. It can also occur in submandibular gland, sinonasal tract and minor salivary glands. There is a female preponderance with female to male ratio of about 2:1, with peak occurrence in the sixth to seventh decade. It usually arises de novo, occasionally develops in a pre-existing pleomorphic adenoma (16%). Distant metastasis is quiet rare (14%). Overall 5-year survival rates is more than 80%.

CASE REPORT:
46 year old male presented with painless swelling on right side of face in the parotid region of 4 years duration. There was sudden increase in size and pain for about one year duration.
There was a no history of facial nerve weakness or palsy.

**RADIOLOGY:**
CT SCAN: Right massive parotid tumor with possibility of right internal jugular vein involvement. MRI: Huge solid mass involving right side of neck and right temperomandibular region. Fat plane between the lesion and deep structure was normal. GROSS: The tumor was well circumscribed, large, globular, grey white soft tissue mass measuring 25 x 18 x 13 cm. Overlying skin showed ulceration (Fig-1). Cut section showed grey white to yellow, solid, firm mass with focal glistening mucoid areas and occasional gritty areas. Deep margin was formed by fascia.

**MICROSCOPY:**
Microscopic examination studied showed tumor lobules separated by fibrous septa. Within the lobules, cells were arranged in solid sheets, compresses of tubules lined by two cell population. Predominant cells were large cell with clear to pale eosinophilic cytoplasm and central to peripherally placed round nucleus - myoepithelial cell. Few cells were small cuboidal cell with eosinophilic cytoplasm and central round to oval nucleus- ductal epithelial cell. There was no nuclear pleomorphism or mitosis. (fig-2) There was no chondromyxoid areas or necrosis. Cut margins all around are free from tumor. Morphologically diagnosed as epithelial myoepithelial carcinoma. However, immunohistochemistry was done to confirm the diagnosis.

**IMMUNOHISTOCHEMISTRY:**
Immunoreactivity with epithelial markers like cytokeratin, epithelial membrane antigen showed focal cytoplasmic positivity in compressed ductular component (fig-3 and 4). The myoepithelial cell showed diffuse positivity with p63 (fig- 5), smooth muscle antigen (fig-6 ) and S100 (Fig 7). Hence, features were consistent with epithelial myoepithelial carcinoma.
**Fig 2** - Ductal epithelial cell showing focal cytoplasmic positivity with cytokeratin (100X and 400X)

**Fig 3** - Ductal epithelial cell showing focal positivity with epithelial membrane antigen (100X and 400X)
DISCUSSION

Epithelial-myoeplithelial carcinoma is quite rare and comprises only 0.4 to 1% of all salivary gland tumors.\(^{(1,2,3,6,7,8)}\) It most often occurs in the major salivary glands, but it can also occur in minor salivary glands and oral cavity.\(^{(9,10,11,12)}\) This tumor behaves as a low-grade malignancy with considerable recurrence rate of about 40%\(^{(3)}\) and capacity for metastasis\(^{(14\%)}\)\(^{(3)}\) to cervical lymphnode, lung, liver and kidney.\(^{(2,3)}\) Usually it presents as asymptomatic, painless slowly enlarging mass. Occasionally facial nerve weakness or pain can occur. Most of the tumors are about 2-8cm in diameter.\(^{(2,8)}\) Mostly it shows good prognosis.

Factors associated with poor prognosis are,\(^{(3,6)}\)
1. Large tumor size
2. Rapid growth
3. Occurrence in the minor salivary gland
4. Solid growth pattern
5. Nuclear atypia more than 20% of tumor areas
6. DNA aneuploidy and
7. High proliferative activity.

Histologically, Epithelial myoeplithelial carcinoma is the prototypical biphasic tumor, tubules, duct like structure- lined by double layered cells. Inner luminal layer with a ductal phenotype and an outer abluminal polygonal myoeplithelial cell layer. Inner luminal cells are single, cuboidal or columnar cells with finely, granular, eosinophilic cytoplasm and central round or oval nucleus. Outer abluminal cells are single or multiple layered, polygonal with abundant clear cytoplasm which are rich in glycogen. They typically have eccentric vesicular nuclei. Classically, the myoeplithelial cells are clear cells or with variant morphologies like oncocytic variant, epitheliod variant.\(^{(2,13)}\) The proportion of each cell type are variable. Cytologic atypia is mild and the mitotic count is usually 2 per 10 high power field.\(^{(4,5)}\)
Cystic and papillary areas are seen in 20% of cases.\(^{(1,4,6)}\) Rarely, solid growth pattern composed predominantly of myoepithelial cells may obscure the epithelial cell layer. In such condition immunohistochemistry helps to confirms the biphasic nature of the neoplasm. Epithelial component show positivity with cytokeratin and epithelial membrane antigen. Myoepithelial component show positivity with smooth muscle antigen, S-100, p63, calponin, Bcl-2 and smooth muscle heavy chain.\(^{(14,15,16)}\)

The main differential diagnosis includes cellular pleomorphic adenoma, myoepithelioma/myoepithelial carcinoma, clear cell carcinoma, basal cell adenoma and adenocarcinoma.\(^{(1,4)}\)

Pleomorphic adenoma may resemble epithelial myoepithelial carcinoma. There is chondromyxoid stroma and outer myoepithelial cells melt into the surrounding chondromyxoid stroma. In our case there was no chondromyxoid area and melting of myoepithelial cells. Hence pleomorphic adenoma was ruled out. Myoepithelioma and myoepithelial carcinoma is a monophasic tumor that consists purely of myoepithelial cells and no ducts. Myoepithelial carcinoma is not well circumscribed. Finding few ductal element rule out myoepithelial carcinoma.\(^{(17)}\)

This present case showed dual population of cells with ductal phenotype and myoepithelial phenotype. Clear cell carcinoma is also poorly circumscribed and the cells are usually more epithelioid, and set in a hyalinized stroma. The cells of clear cells lack S 100 positive myoepithelial differentiation.\(^{(18)}\)

This case showed diffuse positivity with S 100.

Basal cell adenoma and adenocarcinoma, although composed of ducts and myoepithelial cells, are characterized by palisading of outermost layer of tumor nest. In the present case, there was no palisading of outer layer.

This is a rare case of epithelial-myoeoepithelial carcinoma of parotid gland. The tumor size was large- 25cm as compared with a mean diameter of 2 -8cm. Histopathological examination reaveled predominance of myoepithelial cell component and compressed few ductal component. However, immunohistochemistry with cytokeratin, EMA highlighted the compressed ductal component. S-100, p63, and smooth muscle antigen highlighted the myoepithelial layer, thus supports the diagnosis.

**CONCLUSION:**

Epithelial myoepithelial carcinoma is a low-grade epithelial salivary gland tumor, which represents approximately 1% of all salivary gland neoplasms.\(^{(1,2,3,6,7,8)}\) Maximum tumor size recorded in the literature being 12cm.\(^{(2,6)}\) Generally this tumor has excellent prognosis. Factors associated with adverse prognosis include large tumor size, rapid growth, occurrence in minor salivary glands, solid growth pattern, nuclear atypia in >20% of tumor, DNA aneuploidy and high proliferative activity.\(^{(3,6)}\) This present case had the features of large size, rapid growth and solid growth pattern which carries poor prognosis. Hence this case is presented for its rarity and atypical presentation. In this scenario, the chances of local recurrence and metastasis are high which needs adjuvant radiotherapy and regular follow up.

**BIBLIOGRAPHY:**


