Polymorphous Low-Grade Adenocarcinoma of Major Salivary Gland - A Case Report
Maharudrappa Basnaker; GS Kumar

Abstract
Polymorphous low grade adenocarcinoma (PLGA) is a relatively newer entity of salivary gland tumors of low aggressiveness, occurring mainly in the minor salivary glands. The characteristic histopathological feature of polymorphous low grade adenocarcinoma is its cytological uniformity with varied histomorphology. We report a case of PLGA arising from right parotid salivary gland in 35 year old male patient.

Key words: Polymorphous; Low Grade; Parotid; Adenocarcina; Salivary Gland; Tumors

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Introduction
The polymorphous low grade adenocarcinoma (PLGA) is a recently recognized type of salivary gland malignancy, first described by Batsaki et al (1) as terminal duct carcinoma & Freedman et al (2) as lobular carcinoma of intraoral minor salivary gland origin. The population of tumor cells is quite varied, were in many histopathologic growth patterns are observed, hence the term Polymorphous.

WHO classification of salivary gland tumors published in 1991, second edition, classified it under a separate category & recognized it as a specific entity (3). However it is realized that this tumor possess a distinct clinicopathologic features and is one of the more commonly occurring minor salivary gland malignancy. Although PLGA predominantly occurs in oral minor salivary glands, there are few reports of PLGA occurring in major salivary glands. In this article we report a case of PLGA that developed in the parotid gland and was surgically treated with facial nerve preservation and with a brief review of the PLGA’s occurring major salivary glands.

Case Report
A 35 year old male patient presented with 3 year history of swelling in front of his right ear. On examination, the swelling measured about 3.3cms in diameter. The tumor was firm to hard in consistency. It was localized to superficial lobe of parotid gland. A diagnosis of pleomorphic adenoma was done. The lesion was surgically excised through superficial parotidectomy with facial nerve preservation. The patient had regular follow for 2 years. There after the patient is lost for follow up.

Pathology: Macroscopically, the resected tumor mass was well circumscribed, partly encapsulated & measured 2x3x3cms. The cut surface was pale with hemorrhagic foci & several cystic spaces. Histopathology of the lesion revealed round to oval shaped isomorphic cells with bland, scanty hyper chromatic oval nuclei, occasional nucleoli, & mitotic figures were rare. These cells formed diverse patterns, including tubular, trabecular, papillary & follicular & cystic spaces were observed an area of neural invasion was observed.

Discussion
This distinctive neoplasm was described by several investigators and has been described under different terminology as polymorphous low grade adenocarcinoma, terminal duct carcinoma & lobular carcinoma (1, 2). The tumor is so designated because it demonstrates architectural variability in different areas, like glandular, trabecular, tubular, cribriform, Indian file & solid, while exhibiting relative cellular uniformity. However luminal eosinophilic material & myxoid changes were also observed (4). The tumor has infiltrating borders, invading into adjacent soft tissue with low metastatic potential. Perineural invasion & intra neural involvement is characteristic. Mitoses are rarely seen. PLGA is more commonly seen in minor salivary glands with female predilection, with an age range of 23-79 years (2). The recurrence rate of this tumor is approximately 25% which may take up to 10-20 years (1). Approximately 10% metastasizes to local lymph nodes (5).

The polymorphous low grade adenocarcinoma has been described as arising within pleomorphic adenomas of major salivary glands. The reported occurrence of this relatively new entity in major salivary glands are few and
most of the reported cases occurred in parotid salivary glands (4, 6-13) (table-1). Two cases in submandibular glands(14, 15) (Table-2) & only one case reported on sublingual salivary gland till date.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age/Sex</th>
<th>Size</th>
<th>Metastases</th>
<th>Recurrences</th>
<th>Follow up</th>
<th>Papillary Cystic Pattern</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>32</td>
<td>60/M</td>
<td>5cms x5cms.</td>
<td>No</td>
<td>No</td>
<td>36 months</td>
<td>Yes</td>
<td>Ruiz-Godoy, et al(15, 16)</td>
</tr>
<tr>
<td>33</td>
<td>25/M</td>
<td>3cms x3cms.</td>
<td>No</td>
<td>No</td>
<td>-</td>
<td>Yes</td>
<td>Arathi N, et al(4)</td>
</tr>
<tr>
<td>34</td>
<td>35/M</td>
<td>4 cms.x3cms.</td>
<td>No</td>
<td>No</td>
<td>-</td>
<td>Yes</td>
<td>Gelinick I. et al(11)</td>
</tr>
<tr>
<td>35</td>
<td>45/M</td>
<td>20cms.x11cms</td>
<td>No</td>
<td>No</td>
<td>-</td>
<td>Yes</td>
<td>Ashli K et al(12)</td>
</tr>
<tr>
<td>36</td>
<td>35/M</td>
<td>3.3cms x3cms</td>
<td>No</td>
<td>No</td>
<td>2 4 months</td>
<td>Yes</td>
<td>Basnaker MH. et al</td>
</tr>
</tbody>
</table>

Table 2 Sub Mandibular Salivary Gland Tumor Report From The Literature

<table>
<thead>
<tr>
<th>Case</th>
<th>Age/Sex</th>
<th>Size</th>
<th>Metastases</th>
<th>Recurrences</th>
<th>Follow up</th>
<th>Papillary Cystic Pattern</th>
<th>References</th>
</tr>
</thead>
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<tr>
<td>1</td>
<td>72/F</td>
<td>3.5cms x25cms.</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>Yes</td>
<td>Haba R, et al(15)</td>
</tr>
<tr>
<td>2</td>
<td>67/M</td>
<td>2.5cms x2cms.</td>
<td>No</td>
<td>No</td>
<td>20 months</td>
<td>Yes</td>
<td>Yutaka yamazaki et al(14)</td>
</tr>
</tbody>
</table>

Nagao et al(10) were of the opinion that PLGA’s of minor salivary glands & major salivary have similar clinico-pathologic characteristic. On the other hand Mark et al(7) mentioned that some differences do exist in PLGA located in the minor salivary gland with respect to those located in parotid gland. However the reported cases of PLGA’s in submandibular & Sublingual salivary glands were not analyzed. Reviewing these cases we are of the opinion that PLGA’s in submandibular & sublingual salivary glands have clinicopathologic characteristics similar to those seen in minor salivary glands without influencing their anatomic locations. Because of the varied histomorphology exhibited, most of the features of the PLGA’s are similar to those observed in pleomorphic adenoma & Adenoid cystic carcinoma. Distinguishing PLGA from other tumors such as pleomorphic adenoma and adenoid cystic carcinoma is of significance, because these tumors have comparatively worse prognosis. In addition they are most likely to recur, metastasize to regional & distant sites(17).

Myxoid tissue is present both in PLGA & Pleomorphic adenoma. In addition, the myxochondroid & chondroid area present in pleomorphic adenoma are not evident in PLGA. The distinction between PLGA& adenoid cystic carcinoma (ACC) is mainly based on cytological features. Cells in PLGA are cuboidal or columnar, have vesicular nuclei & conspicuous eosinophilic cytoplasm, whereas basaloid cell type is characteristic of ACC. Papillary & fascicular growth patterns are extremely rare in adenoid cystic carcinoma(11). The infiltrative growth pattern, neurotropism are the important histopathologic diagnostic clue which aids in differential diagnosis of PLGA from Pleomorphic adenoma(4).

Immunohistochemical studies on PLGA revealed that glial fibrillar acidic protein (GFAP) is negative, whereas the same is positive in Pleomorphic adenoma, other cytochemical markers reported positive for PLGA are S-100, Vimentin(11, 15, 16).

Analyzing the role of the c-kit IHC in the differential diagnosis of ACC & PLGA Arathi et al(4) are of the opinion that more number of cases needs to be evaluated to draw a firm conclusion. The review of the biologic behavior of PLGA’s arising in minor salivary glands on long term observation was excellent. However the reported cases of PLGA’s arising in major salivary glands are 32 in numbers as reviewed (16). There after 5 case of PLGA’s are reported in parotid salivary including one our case (4, 5, 15) (11)(table-1), two cases of PLGA’s occurring in submandibular salivary glands(14, 15) Table-2. The only case of PLGA was reported in sublingual salivary gland (17).

The age of occurrence of PLGA’s in major salivary glands varied from 25 years(4) to 85 years(18) , with male to female ratio of 1:2. The tumor size varied from 0.8cms(9) to
20cms(12). Although papillary pattern have found in most of the reported cases of PLGA’s in parotid salivary gland(15), & (table-1). Submandibular salivary gland (table-2) & Sublingual salivary gland (table-3) no metastasis was reported except 1 of the case in the series of PLGA’s studied by Kemp et al(9). Recurrences of the PLGA’s in major salivary glands were also reported(4, 9). The follow up of PLGA’s in major salivary glands well not documented in most of the reported case in the literature.

Hence it is difficult to clearly define the biologic behavior of PLGA’s although they are considered to be an indolent tumors occurring in minor salivary glands.

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