CASE REPORT

Epithelio- Myoepithelial Carcinoma of Masseteric Space: a Case Report

Naresh K. Panda¹ · Sourabha K. Patro¹ · Roshan K. Verma¹ · Abdul Wadood¹ · Debajyoti Chatterjee²

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Abstract Epithelio-myoepithelial carcinoma (EMEC) is a very rare malignancy of the salivary gland which comprises less than 1 % of salivary gland tumors. In the parapharyngeal space they can either arise from the deep lobe of parotid gland or de novo from minor salivary glands. We present a case of epithelio-myoepithelial carcinoma of parapharyngeal and masseteric space. The case is important for its rarity of presentation and we discuss the surgical difficulties and morbidities.

Keywords Epithelial myoepithelial carcinoma · Masseteric space and parapharyngeal space · Poor prognosis

Introduction

Epithelio-myoepithelial carcinoma (EMEC) is a rare malignancy of the salivary glands comprising less than 1 % of salivary gland neoplasms. [1] [2] Even though parotid gland is the most common site involved, these can present at other sites involving minor salivary glands like hard palate, soft palate and parapharyngeal space etc. They have a female

predilection and are seen most commonly in the elderly population, usually in the seventh decade [3]. This unusual salivary tumor was first studied and named by Donath et al. in 1972. [4] The tumor is peculiar in its biphasic histopathologic appearance, characterized by tubules lined by an inner layer of cytokeratin-positive bland cuboidal epithelial cells surrounded by an outer layer of S100 positive myoepithelial cells. [4] In 1991, EMEC was recognized as a distinct entity and subtype of salivary gland adenocarcinoma, and it became part of the new classification system. [3] EMEC arises mostly from parotid glands but it can also arise from the submandibular gland and from minor salivary glands. EMEC is a low grade malignant tumor that may recur locally after resection. Distant metastasis rarely occurs [3–5]. Parapharyngeal space is a potential space containing fat, lymphatic tissue, nerves, vessels and minor salivary glands. Hence these can give rise to variety of neoplasms. We present a case of epithelio-myoepithelial carcinoma of the parapharyngeal and masseteric space which is unique for its rarity of presentation and discuss the difficulties treatment and morbidities.

Case Report

A 37 year old female presented to our department with complaints of swelling of the right cheek for 2 years with gradually progressive decrease in mouth opening for 6 months. The swelling was insidious in onset, gradually progressive and did not have any history of pain, bleed from oral or nasal cavity, cranial nerve palsy. It was associated with another swelling in right side of neck for the last 2 months. Clinical examination revealed a 3 × 2 cm ill-defined, non-tender swelling over the right cheek extending superiorly up to a point 1 cm below the zygomatic arch, anteriorly up to 1 cm from the angle of mouth and posteriorly up to 1 cm in front of the

✉ Roshan K. Verma
roshanverma@hotmail.com

¹ Department of Otolaryngology and Head Neck Surgery,
Post Graduate Institute of Medical Education and Research
(PGIMER), Chandigarh 160012, India

² Department of Histopathology, PGIMER, Chandigarh 160012, India

tragus with restricted mobility. Oral cavity examination did not reveal any growth or swelling except for decreased mouth opening of 1 cm between the incisors. Neck examination revealed a 2×1 cm mobile lymph node palpable on right level II. Rest of the general, systemic and cranial nerve examinations were normal. Fine needle aspiration cytology from the cheek swelling was reported as adenocarcinoma, basal cell type with the neck node was reported as metastatic disease. Contrast enhanced computerized tomography of the face and neck showed heterogeneously enhancing soft tissue density with irregular border in right masseteric space extending into the infra temporal fossa with ill-defined plane between the tumor and the muscles of the masseteric and infra-temporal fossa (Fig. 1) whereas similar cuts in the post-operative scan revealed absence of any lesion suggesting a complete radiological excision of the lesion (Fig. 2). There were multiple lymph nodes involving the right levels Ib, II and III. Excision of the tumor along with right selective neck dissection was done for the patient. Intra-operatively primary tumor was found to be 3×3 cm in size involving the masseteric space and infra temporal fossa. There were multiple lymph nodes involving the right levels Ib, II and III in neck dissection with largest being 0.5×0.5 cm at the level II. Histopathologic examination revealed tumor mass invading into the surrounding normal parenchyma in broad fronts, separated by sclerotic stroma (Fig. 3). There were two types of cells comprising of predominantly outer layer cells composed of pleomorphic vesicular nuclei, inconspicuous nucleoli and moderate clear cells (Fig. 3b). Other cells forming the tubule had hyper chromatic nuclei; conspicuous nucleoli and moderate cytoplasm (Fig. 3b and c). SMA immune staining highlighted the myoepithelial cells, while cells in the tubule were negative (Fig. 3d). Lymph nodes showed evidence of metastasis (Fig. 4). Hence, a diagnosis of epithelio-myoeplithelial carcinoma with neck metastasis was made.

Margins were clear on histopathologic study. The patient received post-operative external beam radiotherapy 60Gy in 30# for 6 weeks in view of the aggressive nature of the disease and the histological involvement of lymph nodes for metastasis. Thereafter, the patient is under the follow up of department of otolaryngology since august 2013 for the last 3 years without any evidence of loco regional recurrence.

Discussion

Epithelio-myoeplithelial carcinoma as the name indicates is a histopathologic diagnosis. It was included in the World Health Organization classification of salivary tumors in 1991 and has an ICD-O-3 code of 8562/3. They are known by synonyms like adenomyoepithelioma, clear cell carcinoma or glycogen rich adenocarcinoma. They comprise less than 1 % of salivary gland tumors and more common in females. The age range is between 3 and 89 years with a peak in 6th and 7th decade. It occurs mostly in major salivary glands, mainly in the parotid (60 %) gland [6]. However, the minor glands of oral mucosa and laryngeal region can also be affected. They can also occur in the parapharyngeal space where it can arise either from the deep lobe of parotid or de-novo from the minor salivary glands [7]. They can rarely be seen in the paranasal sinuses [8] and nasal cavity [9] also. Case reports showing origin of these tumors from parotid gland can be found in literature. [6] This patient presented to us with a tumor in the masseteric space. There is only one case report found in the literature showing the possibility of this tumor to arise from the masseteric space [10]. They mostly present as a slow growing painless swelling as it was with this patient, who had a history of 2 years. Once mucosa is affected, they can present with ulceration. Rapid progression, pain and facial palsy are features of high grade malignant transformation. The presence of cheek

Fig. 1 Coronal and axial CT pictures of the lesion involving the masseteric space having infiltrating margins with no definite fat planes with the surrounding muscles

Post operative CT scan

Fig. 2 Post-operative coronal and axial CT pictures

swelling initially and later development of trismus suggests that the tumor arose from the masticator space and later spread to the infra temporal fossa in the course of natural temporal progression of the lesion. Metastasis is known to occur, cervical lymph nodes being the most common local site and lung being the most common distant site. [11] The treatment is wide local excision with adequate margins and neck dissection due to the presence of lymphadenopathy. However, local recurrences are known and the incidence can be as high as 40 % [3].

Macroscopically, they are seen as multi nodular mass, not circumscribed properly and lack a true capsule. There may be

cystic spaces with in the tumor. Microscopic appearance of the tumor is the most convincing feature for diagnosis. The hall-mark of epithelio-myoepithelial tumor is the bi-layered duct like structure. The detection of myoepithelial cells is responsible for the diverse appearance in minor salivary gland tumors. [12] The inner layer comprises of single layered cuboidal cells with dense finely granular cytoplasm and round central or basal nucleus. The outer layer is made of single or multiple layers of polygonal cells with clear cytoplasm and vesicular slightly eccentric nucleus. The duct like structure is surrounded by PAS positive hyaline like eosinophilic material. On immune staining, the outer layer usually stains positive for

Fig. 3 **a** showing the tumor mass invading into the surrounding normal parenchyma in broad fronts, separated by sclerotic stroma (HE 1 00X). **b** Bicellular architecture of neoplastic tubules comprised of a single layer of round to cuboidal luminal(ductal) cells, having bland nuclei and moderate amount of eosinophilic cytoplasm surrounded by one or more

layers(mantle) of large polygonal abluminal (myoepithelial) cells, having abundant water clear cytoplasm (bold arrow) covered by a well-defined basement membrane. (HE400X). **c**- Immuno staining shows positive EMA on the ductal (luminal epithelial) cells and positive SMA on the myoepithelial (clear) cells

Fig. 4 Lymph node structure is effaced with tumour nests in broad fronts along with the clear cells replacing most of the normal pattern. (HE50X)

myoepithelial markers like smooth muscle actin, HHF35, p63 and/or calponin and the luminal cells stain with cytokeratins. [13] On cyto-photometric analysis, these tumors show diploid histograms [14].

Masseteric space and infra-temporal fossa are relatively difficult sites in terms of accessibility. There are various approaches to access tumors at this site. We performed an anterolateral approach by giving a midline lip splitting incision and extending it posteriorly into a transverse cervical incision till the upper border of the hyoid bone and then turning it upward and posteriorly till the mastoid tip. Following which lower cheek flap was raised. This was done as we had to perform neck dissection along with the excision of the primary tumor. The incision was extended intra orally along the gingivo labial and gingivo-buccal sulcus till the posterior border of the tumor could be accessed. At the posterior end of the alveolar process we extended the mucosal incision just lateral to the retro-molar trigone in order to approach the tumor in the masseteric space. The tumor was ill defined and infiltrating into the surrounding muscles of mastication. The stylomandibular ligament had to be cut in order to improve the access. Masseter and lateral pterygoid muscles were involved and had to be cut for providing adequate margin. Even though, there was initial improvement in mouth opening in the post-operative period, patient developed trismus of pre-operative severity following radiotherapy. A trans-parotid approach could have been another option with improved accessibility and exposure. However radiology did not reveal any involvement of both the superficial and deep lobes of parotid, as evident by the presence of intact fat planes between the parotid gland and the tumor. Thus, considering the above mentioned reason and the risks of facial injury involved with a trans-parotid approach in our patient who is a 37 year old young female, a trans-parotid approach was not preferred. Moreover a trans-

parotid approach would have required a much longer incision for when neck dissection is associated with it in order to achieve adequate a level 1 clearance which can be easily done in a lower cheek flap approach. A lateral trans-temporal approach was also not advisable as the tumor was more in the anterior compartment of infra temporal fossa. Owing to the infiltrative nature of disease, high rate of local recurrence and multiple involved lymph nodes, the patient was given post-operative adjuvant radiotherapy. Till date the patient is on follow up and there is no evidence of loco regional recurrence. The patient is free of disease and only complaints of persisting trismus.

Conclusion

Epithelio-myoepithelial carcinoma (EMEC) is a low grade malignant tumor with high rate of recurrence mainly presenting in major salivary gland. Masseteric space and infra temporal fossa are very rare sites of origin for this tumor and is associated with difficult surgical accessibility and post-operative morbidity. Though, epithelial-myoepithelial carcinoma is very rare salivary gland tumor, it should not be missed as under treatment may lead to recurrence and can reduce the survival in such patients. Radical surgery followed by radiotherapy remains the treatment of choice for this cancer.

This material has never been published and is not currently under evaluation in any other peer-reviewed publication. There is no conflict of interest or financial disclosure to be made.

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