Case Report

Epithelial Myoepithelial Carcinoma- A Rare Case Report

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Abstract:

Background: Salivary gland tumors occupy an imperative part while considering the entire spectrum of tumor pathology occurring in the maxillo-facial and cervical areas. They often pose diagnostic uncertainty due to intrinsic architectural and cytologic complexity, overlapping architecture. Epithelial-myoepithelial carcinoma is a rare tumor, constituting 1% of all salivary gland tumors. ***Methods and Results*****:** We present a case of Epithelial-myoepithelial carcinoma in a 55-year-old male who presented with paresthesia of lower lip and swelling of the right side of face . Imaging revealed complete destruction of right ramus with an enhancing soft tissue mass. Patient underwent a surgical excision of the mass, which revealed an Epithelial-myoepithelial carcinoma. ***Conclusion***: Although Epithelial-myoepithelial carcinoma is extremely rare in occurrence, have immense destructive capabilities. Hence a description of clinical presentation, radiographic extension and histopathologic presentation would be of great additive value to the sparse information of this rare tumor

Key words: Epithelial Myoepithelial Carcinoma, Parotid Gland, Salivary Gland, Carcinoma, Neoplasm

Introduction

Salivary gland tumors occupy an imperative part while considering the entire spectrum of tumor pathology occurring in the maxillo-facial and cervical areas. They often pose diagnostic uncertainty due to intrinsic architectural and cytologic complexity, overlapping architecture amongst many benign and malignant tumors.[1] Carcinomas of the salivary glands are uncommon.[2] Epithelial-myoepithelial carcinoma (EMC) is a rare malignant tumor, which constitutes approximately 1% of all salivary gland tumors. It was established as a distinct clinicopathologic entity by the World Health Organization in 1991.[3] The major salivary glands, especially the parotid gland, are the most frequent site involved; however, cases arising in minor salivary glands and extraoral areas have also been reported.[4] We report a case of EMC in a 55 year old male, and discuss the clinical, radiological, histopathological, and immunohistochemical findings.

Case Report

A 55 year old male patient presented to the dental hospital with altered sensation of the right side of lower lip of 3 months duration and swelling of the right side of face since 2 months. Patient was apparently normal about 3 months back when he developed difficulty in mouth opening for about 3 days. Following resolution of difficulty in mouth opening, patient noticed altered sensation of right side of lower lip and chin. Patient had noticed swelling on the right side of face since 2 months which had gradually increased in size. The swelling was asymptomatic. Past medical history was noncontributory.

On extra oral examination diffuse swelling was present in the right side of the face measuring about 5 x 4 cms in size (see Figure I). Swelling was nontender and firm in consistency. Elevation of right ear lobe was noted. Lymph nodes were not palpable. Light touch and pin prick sensation was altered on right side of the lower lip and chin compared to left side. Intra oral examination revealed firm mass along the ramus of mandible. Fine needle aspiration was nonconclusive. Panoramic imaging revealed complete destruction of right ramus with residual condyle (see Figure II). CT scan revealed complete erosion of ramus of right mandible, associated with a soft tissue mass. The soft tissue mass was infiltrating the ipsi-

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lateral masseter muscle but the parotid gland and vascular structure were spared (see Figure III). MRI revealed large expansile lytic lesion with heterogeneously enhancing solid soft tissue involving right ramus, condyle and coronoid process of mandible with large solid soft tissue component appearing hyperintense on T2 (see Figure IV). Bone scan and ultrasound ruled out possibilities of distant metastasis. Excision of mass was carried out with wide margins and right submandibular lymph nodes (see Figure V). Histopathogical section showed partially encapsulated tumor composed of lesional cells arranged predominantly in a lobular pattern. Islands of cells infiltrating adjacent connective tissue were also observed. The cells at the periphery of the ducts, lobules and islands showed clearing of the cytoplasm, some with angular nuclei (myoepithelial cells). Some areas show microcyst formation with papillary projections of lesional cells into the cystic spaces. Nuclear changes such as pleomorphism, hyperchromatism and mitotic figures were seen in the deeper areas of the sections. Areas of necrosis and hemorrhage were also seen. The submandibular lymph nodes however, was free of tumor infiltration (see figure VI A&B). Immunohistochemistry showed positivity for cytokeratin in the epithelial and myoepithelial cells and vimentin in the myoepithelial cells alone. The histopathological features were diagnostic of epithelial – myoepithelial carcinoma. Patient was subsequently followed for a period of one year during which no evidence of recurrence or metastasis. Further follow up could not be done due to logistical reasons.

Discussion

EMC was first described by Donath et al in 1972. It is generally accepted that EMC was previously reported under a variety of names, including adenomyoepithelioma, glycogen-rich adenoma, glycogen-rich carcinoma, clear cell adenoma, and clear cell carcinoma. EMC was listed in the World Health Organization classification as a distinct clinicopathologic entity in 1991.[3] 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 structures. Both electron microscopy and immunohistochemistry disclose that the outer clear cells are of myoepithelial origin.[5]

The most prevalent site of occurrence of EMC is the major salivary glands, especially in the parotid gland. Around 10% of this type of tumour has its origin in the submandibular gland and between the 10-15% in the minor salivary glands.[6] Some studies have reported that the intercalated duct in the minor salivary gland is lacking or shorter than that in the parotid gland. This may explain why most EMCs occur in the major salivary glands. However, it is unknown why EMCs occur more commonly in the parotid gland than the submandibular gland. This difference may be related to variations in the intercalated ducts in the two sites.[3] The etiological factors for salivary tumors are not clearly defined.[2]

As in other salivary gland neoplasms, there is a female predominance, with a female-to-male ratio of approximately 2:1. Most patients affected are adults in the sixth or seventh decade of life, although rare cases have been described in children and adolescents. The most common clinical presentation is a slowly enlarging mass. Occasionally, pain or facial weakness may be present. The mean tumor size is 2-3 cm, with a range from 1 to 12 cm in largest dimension.[1] The lesion may be encapsulated, but the capsule is frequently incomplete and tumor nodules often extend through it.[5] In the present case tumor was partially encapsulated. Gross haemorrhage and necrosis are uncommon.[1]

CT and MR appearances of EMC are nonspecific, and EMC cannot be differentiated from other parotid neoplasms.[4] In this case CT and MRI study indicated that the mass was in the right submasseteric space abutting the parotid gland posteriorly while draped over by the masseter laterally and compressing the medial pterygoid thus suggesting a possible site of origin as extracapsular gland tissue from parotid or accessory parotid. CT and MR imaging of the lesion showed complete destruction of the right ramus indicating a malignant aggressive tumor but this did not correlate with the histological findings of a low grade malignant lesion. Morphologically EMC express both epithelial and myoepithelial differentiation. Tumor presents as bulky, lobulated, slow-growing mass becoming large. Tumor myoepithelial phenotype has been cited as a reason why tumors generally grow in a bulky lobulated fashion, rather than in infiltrating pattern.[4]

In most tumors, cytologic atypia is mild or absent,

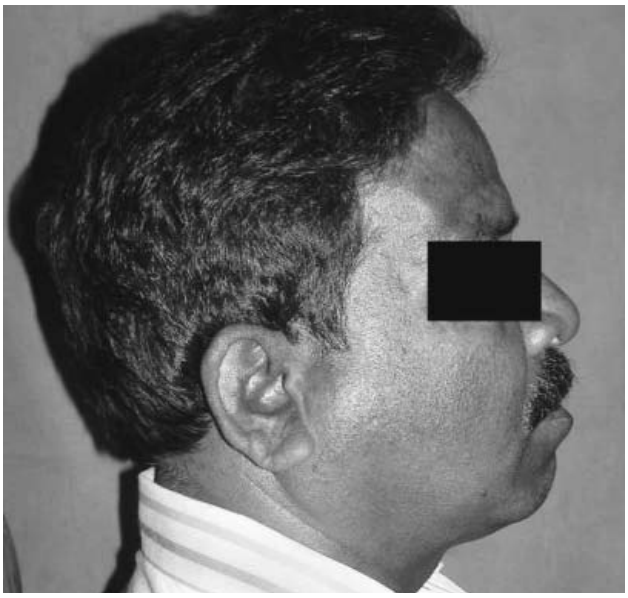

Figure I: Photograph of patient depicting the extent of swelling.

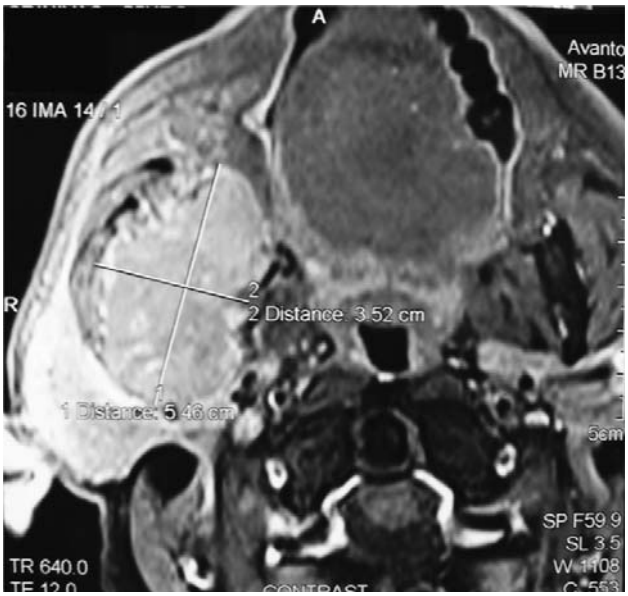

Figure IV: Axial MRI section showing expansile lytic lesion with heterogeneously enhancing solid soft tissue involving right ramus, condyle and coronoid process of mandible.

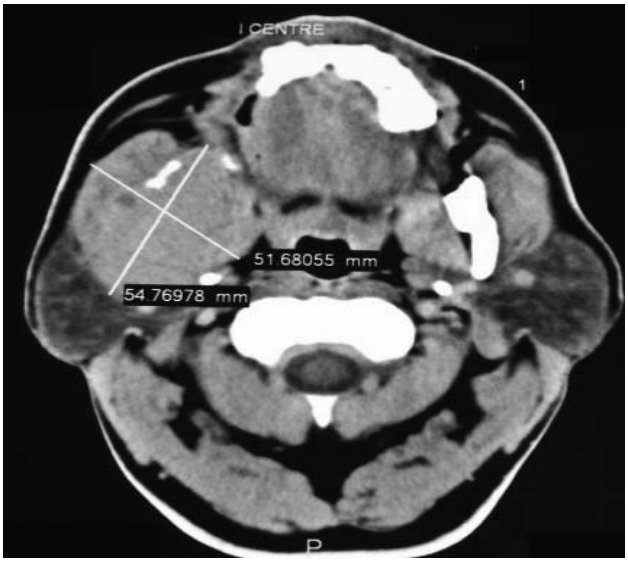

Figure III: Axial CT scan showing complete erosion of ramus of right mandible also notice mass infiltrating into the ipsilateral masseter muscle.

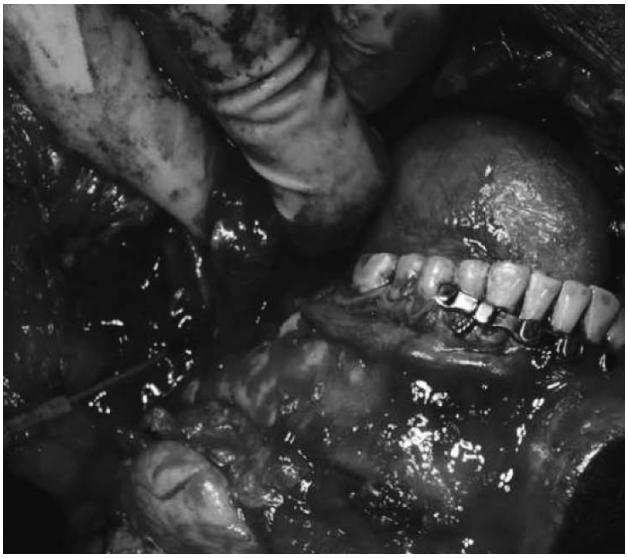

Figure V: Photograph showing surgical exploration of the site.

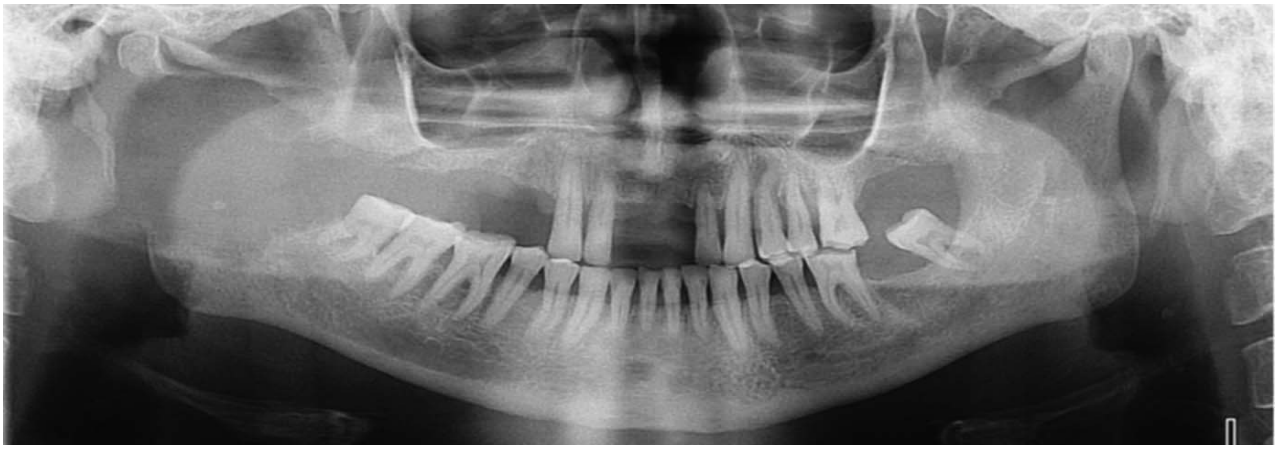

Figure II: Orthopantomogram showing complete destruction of right ramus and residual right condyle.

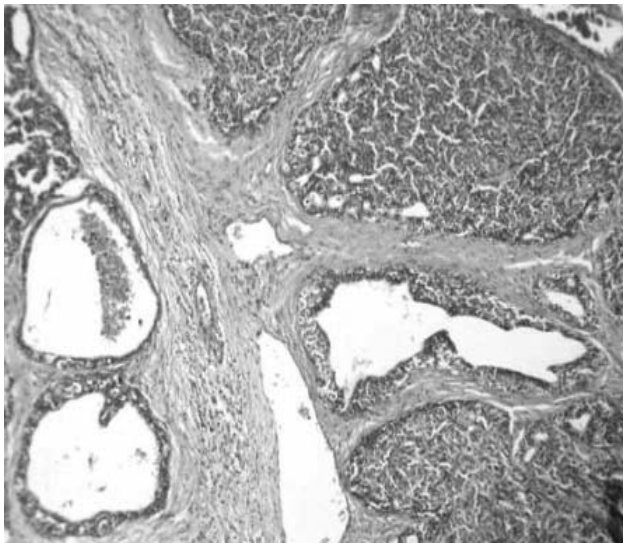

Figure VI (A): Photomicrograph (100X resolution, Haemotoxyline & Eosin) showing partially encapsulation, lesional cells in lobular pattern, occasional infiltration of connective tissue and cuboidal and columnar cells attempting to form duct-like structures.

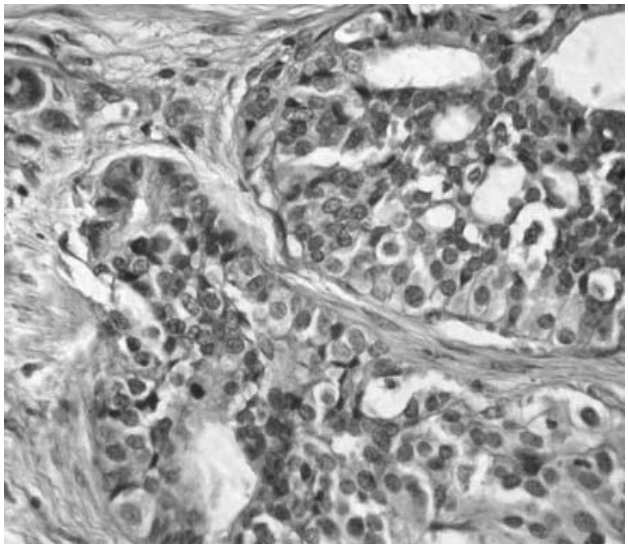

Figure VI (B): Photomicrograph (400X resolution, Haemotoxyline & Eosin) showing cells with angular nuclei (myoepithelial cells), microcyst formation, pleomorphism, hyperchromatism mitotic figures and areas of necrosis and hemorrhage.

but occasional cellular and nuclear pleomorphism and increased mitotic figures may be present. Although these tumors seem to be well circumscribed, nests of tumour cells frequently infiltrate adjacent parenchyma or tissues, including peripheral nerves. Necrosis may be observed in some tumors.[1, 7]

Rarely, some epithelial- myoepithelial carcinoma may be found in combination with other salivary gland tumors forming hybrid carcinoma or show dedifferentiation.[8, 9]

Although some studies have characteristically described the ductal cells as being positive for cytokeratin and the clear cells as immunoreactive for S-100 protein[10], this description is relative. Reactivity for S-100 protein is variable.[1]

Surgical excision with wide margins is indicated as the first choice for treatment because of the locally invasive behavior of these tumors.[11] A wide margin was given while excision was carried out for the present case.

Most studies indicate that EMC is a low-grade malignancy with local recurrence found in 23% to 50% of cases, lymph node metastases in 17% to 23%, and distant metastases to lung, kidney and brain in up to8- l0% and with fatal outcome in up to 8- l0%.[1, 12] the 5-year-survival rate is 67%. The prognosis is bad in tumors having a diameter of 4 cm or more and for those solid tumors predominantly composed of clear cells, forming nests and sheaths.[12] In presenting case, patient had no loco regional recurrence.

Conclusion

Although Epithelial-myoepithelial carcinoma is extremely rare in occurrence, have immense destructive capabilities. Hence a description of clinical presentation, radiographic extension and histopathologic presentation would be of great additive value to the sparse information of this rare tumor.

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