CASE REPORT

An unusual case of epithelial-myoepithelial carcinoma of the parotid gland radiologically simulating a benign lesion

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ABSTRACT

Epithelialmyoepithelial carcinoma (EMEC) is a rare low-grade malignant salivary gland neoplasm that most commonly occurs in the parotid gland but can also arise in minor salivary glands. Here, we present a case of EMEC in a 60-year-old male patient with a huge swelling in the left parotid gland region. Clinically and radiologically, it simulated a benign salivary gland neoplasm. However, fine-needle aspiration cytology and histologic examination revealed atypical myoepithelial cells in solid sheets or nests suggestive of epithelial-myoepithelial carcinoma. Diagnosis was further confirmed by positive immunohistochemical staining with calponin (CALP) and periodic acid-Schiff (PAS) for glycogen.

Key words: Biphasic pattern, epithelial-myoepithelial carcinoma, parotid gland

INTRODUCTION

Epithelial-myoepithelial carcinoma (EMEC) of the salivary glands is a rare tumor, accounting for less than 1% of all salivary gland neoplasms that arise most commonly in the parotid gland but has also been described in minor salivary glands. Patients typically present with a painless slowly growing mass; occasionally there is rapid growth, pain or facial weakness. Also known as *adenomyoepithelioma,* it is an uncommon low-grade epithelial neoplasm. It may commonly recur locally after resection with a tendency to metastasize. Here, we present an unusual case of EMEC that radiologically simulated a benign lesion but histopathologically turned out to be malignant.

CASE REPORT

A 60-year-old male patient reported with a huge swelling in the left parotid gland region, slowly enlarging in size since 5 years. On inspection and palpation, swelling was non-tender, pyramidal in shape, 12 × 10 cm in size, nodular and firm in consistency; temperature over the swelling was normal with no fixation of the tumor to the underlying structures. The ear lobule appeared to be raised with no facial paralysis or restriction in jaw movements [Figure 1]. On the affected side, the lymph nodes were not palpable. The case was clinically diagnosed as benign salivary gland neoplasm involving left parotid gland.

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Hematological investigations showed no significant alterations except increased eosinophilic count. Ultrasonography revealed hypoechoic lesion showing increased vascularity [Figure 2a]. Lateral view and posteroanterior view of skull was normal without any underlying bone resorption [Figure 2b and c]. Fine-needle aspirate showed atypical mononuclear cells suggestive of malignancy [Figure 2d]. Patient was advised a complete surgical excision. The tumor was excised from superficial lobe of parotid gland. No intra-operative complications were reported. Postoperatively, patient showed signs of paresthesia with transient signs of Bell's palsy, which were taken care of. Then the specimen was sent for histopathological examination. On grossing, it showed a well-encapsulated mass, measuring 10 × 12 cm in size and was firm in consistency. The cut section showed white fibrous mass inside the capsule [Figure 2e].

Histopathologically, it showed islands of tumor cells separated by dense bands of hyalinized fibrous connective tissue. Islands were composed of small ducts lined by inner luminal cuboidal cells and outer clear myoepithelial cells representing the characteristic *biphasic* pattern. The cuboidal epithelium was surrounded by clear cells, which interfaced with a thickened hyaline basement membrane. Nests of atypical myoepithelial cells, some in acinar pattern and some scattered in the connective tissue stroma were seen predominantly. These atypical cells had irregular ovoid shapes, prominent nucleoli and fine chromatin. Mitotic figures were not

common. Positive staining with periodic acid-Schiff (PAS) stain suggested the salivary gland origin of the neoplasm, whereas immunohistochemistry showed positivity for calponin (CALP) confirming the myoepithelial differentiation of the tumor.

DISCUSSION

EMEC is defined by the World Health Organization (WHO) classification in 1991 as a rare low-grade malignant neoplasm and accounts for less than 1% of salivary glands tumors. Though the name for this tumor was coined by Donath *et al*., (1972) EMEC was likely recognized as early as 1956 and reported under a variety of names such as *adenomyoepithelioma, clear cell adenoma, tubular solid adenoma* and *clear cell carcinoma*.[1] It arises most commonly in the parotid gland but has also been described in the submandibular gland and minor salivary glands.[2,3] There is a female predominance with the peak occurrence in the seventh decade.[4] However, in our case the patient was male and aged 60 years. EMEC tends to grow in a bulky lobulated fashion, with necrosis and hyalinization of large tumor nodules,[5] as like in our case [Figures 3a and 4b].

EMEC varies not only from carcinoma to carcinoma but also within the same neoplasm.[6] By 1987, a review of published findings had identified 35 cases and a 1989 study from Germany and Sweden added a further 21.[7]

The histogenesis of EMEC is uncertain. It has been suggested that there is bidirectional differentiation from a stem cell to form myoepithelial and intercalated ductal epithelial cells.[7] Our findings are consistent and explicate intimate association of these two cell types but do not further elucidate the histogenesis.

Although originally thought to be benign, it is now apparent that EMEC is a genuine low-grade malignancy.

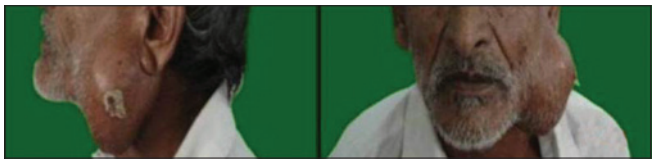

Figure 1: Extra oral swelling on the left side of face

It is histologically most significant for its biphasic cellular pattern, but in some carcinomas biphasic pattern is less apparent and appearance is dominated by solid groups of myoepithelial cells.[6] The tumor has a distinctive histopathological pattern with a proliferation of ductal structures [Figure 4a]. The ducts may be seen in cross section or longitudinally and they may be densely packed together or separated by abundant dense hyaline material. The inner cells of these ductules constitute the epithelial component of EMEC.

These mild to moderately pleomorphic cells have irregular ovoid shapes with prominent nucleoli and fine chromatin. Mitotic figures are not common. The outer cell layer that surrounds the ductules is the clear cell myoepithelial component of EMEC. The nuclei are smaller than those of the epithelial cells, with a definitely condensed and triangular appearance. The cells vary from being with "naked nuclei" to having abundant clear cytoplasm. The clear cells are glycogen positive and should stain for PAS and be sensitive to diastase [Figure 5a].[5]

In our case, biphasic pattern is less evident but ductal epithelial cell proliferation is seen and myoepithelial cells are arranged in solid sheets or nests without ducts [Figure 3b]. Some are present in acinar pattern and some are scattered in the connective tissue stroma [Figure 4c].

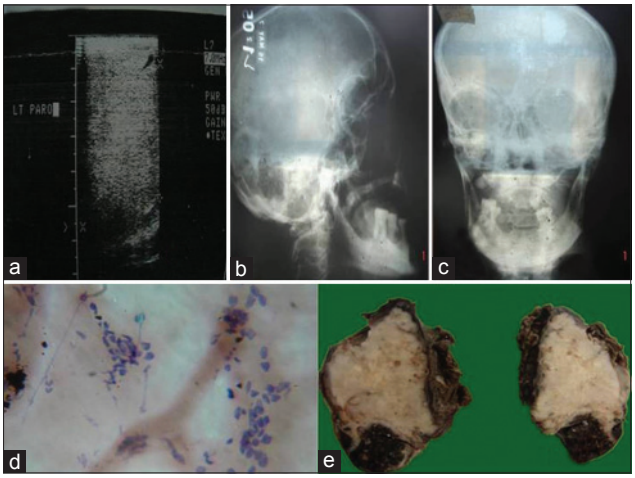

Figure 2: (a) Ultrasonography, (b and c) Lateral and posteroanterior view of skull. (d) Fine needle aspiration cytology, (e) Cut surface of the gross specimen

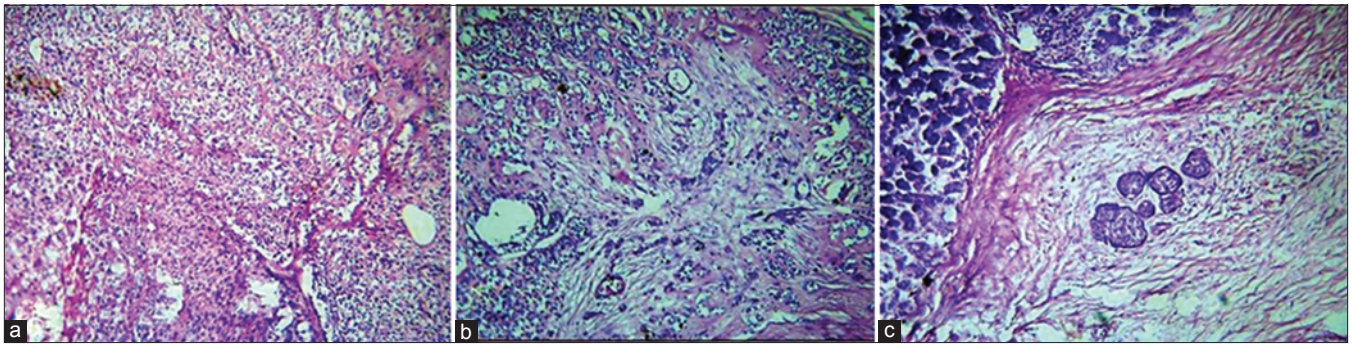

Figure 3: (a) Round or oval solid nests of myoepithelial cells (H&E, ×100), (b) Nests of tumor cells scattered in the stroma in an irregular pattern (H&E, ×100), (c) Infiltrated tumor islands in the capsule (H&E, ×200)

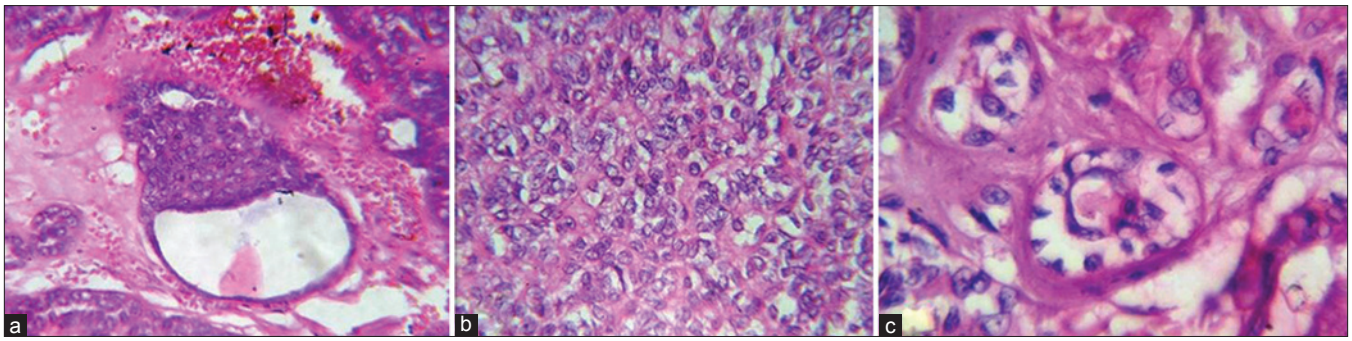

Figure 4: (a) Proliferation of ductal epithelial cells with increased vascularity (H&E, ×200), (b) Biphasic pattern with outer myoepithelial and inner cuboidal cells (H&E, ×400), (f) Nests of atypical myoepithelial cells, some in acinar pattern and some scattered in the connective tissue stroma (H&E, ×400)

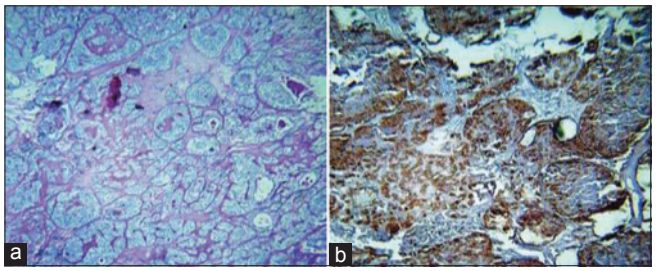

Figure 5: (a) Positive staining for PAS (PAS stain, ×400), (b) Positive staining for CALP (IHC stain, ×100)

Immunohistochemically, epithelial component is selectively well highlighted by all cytokeratins (CK) and epithelial membrane antigen (EMA). Myoepithelial component is demonstrated by S-100, smooth muscle actin (SMA), p63 and vimentin. The newer markers like CALP, caldesmon (CALD) and smooth muscle myosin heavy chain may be useful tools for identifying myoepithelial cells when myoepithelial cell differentiation is not easily identified on routine histologic sections. Immunohistochemical staining with CALP was positive in our case, which confirmed the diagnosis [Figure 5b].[8]

The intercalated duct and myoepithelial cells are capable of manifesting numerous patterns of differentiation, this leads to consideration of other salivary gland neoplasms in the differential diagnosis. Present case can be differentiated from pleomorphic adenoma, acinic cell adenocarcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, clear cell oncocytoma and metastatic renal cell carcinoma.

The number of cases reported till date of EMEC are 125, greater part of which is located in salivary gland with exception of some cases found in nasal cavity, paranasal sinuses, lacrimal glands or bronchus and lungs.[5]

The local recurrence rate of EMEC ranges from 17-60%; however, we did not observe recurrence during 3-years follow up. Distant metastases to lung, liver and vertebrae are known, but cervical lymph node involvement is relatively rare.[9]

CONCLUSION

Salivary gland neoplasms sometimes can be difficult to recognize because of their rarity. Not only should the pathologist always be alert in the diagnosis of these neoplasms but also clinicians should closely follow up on those patients diagnosed as EMEC because of uncertainty of our knowledge about its behavior.

REFERENCES

1. Simpson RH, Clarke TJ, Sarsfield PT, Gluckman PG. Epithelial-myoepithelial carcinoma of salivary glands. J Clin Pathol 1991;44:419-23.
2. Batsakis JG, el-Naggar AK, Luna MA. Epithelial-myoepithelial carcinoma of salivary glands. Ann Otol Rhinol Laryngol 1992; 101:540-2.
3. Morrow TA, Chun T, Mirani N. Epithelial myoepithelial carcinoma of the parotid gland.Ear Nose Throat J 1990;69:646-8.
4. Noel S, Brozna JP. Epithelial myoepithelial carcinoma of the salivary gland with metastasis to lung: Report of a case and review of literature. Head Neck 1992;14:401-6.
5. Silvers AR, Som PM, Brandwein M. Epithelial myoepithelial carcinoma of the parotid gland. AJNR Am J Neuroradiol 1996; 17:560-2.
6. Demet E, Ismail O, Murat E, Berna V. An epithelial myoepithelial carcinoma in parotid gland. Firat Tip Dergisi 2007;12:65-7.
7. Simpson RH, Clarke TJ, Sarsfield PT, Gluckman PG. Epithelial-myoepithelial carcinoma of salivary glands. J ClinPathol 1991;44:419-23.
8. Ramraje SN, Bharambe BM, Zode R. Bombay Hosp Journal 2010;52:141-3.
9. Puri T, Singh K, Sharma DN, Khurana N. Epithelial myoepithelial carcinoma of the base of the tongue: Pathology and management.Indian J Cancer 2004;41:138-40.

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