Epithelial Myoepithelial Carcinoma of Head and Neck Region

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Received: 21 February 2010 / Accepted: 24 November 2011 / Published online: 20 January 2012
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Abstract The objective of the study is to document the clinical, histopathological and immunological findings of epithelial myoepithelial carcinoma (EMEC) which is an uncommon neoplasm. The tumour represents less than 0.5–1% of all salivary gland neoplasms. As per literature, it involves the parotid gland most frequently and rarely, the minor salivary glands. We are reporting a series of four cases of EMEC over a period of one and half years, three from minor salivary glands at different sites and one from the deep lobe of parotid. Interestingly, the pre operative FNAC of all the cases revealed benign pathology. Recurrence occurred in three of them after tumour excision. One patient was subjected to post operative radiotherapy and one patient to chemotherapy. Histopathology and immunohistochemistry is very important in this tumour. There is no definitive guideline of treatment in post operative recurrence cases.

Keywords Salivary gland neoplasms · Fine needle aspiration cytology (FNAC) · Immunohistochemistry (IHC) · Epithelial myoepithelial carcinoma (EMEC)

Introduction

Neoplasms of salivary gland origin are histologically diverse. The overlapping microscopic features showed by many of the benign and malignant tumours pose a diagnostic challenge. EMEC was first introduced in World Health Organization (WHO) classification in 1991. Epithelial myoepithelial carcinoma was first described by Donath et al. in 1972 [1]. This is a low-grade malignancy, which is locally invasive. It has a high chance of recurrence, so complete surgical removal of tumour is very important. FNAC is often not conclusive for diagnosis. Diagnosis is usually based on histopathology and immunohistochemistry.

The aim of this study is to present four new cases of EMEC and to document their clinical, histopathological and immunological findings, and thus permit wider recognition of this distinctive tumour.

Case Reports

We have four cases of EMEC in last one and half year.

First patient, 30 year/female, presented to us in 2001 with painless swelling on the left side of face for 1 year. The examination revealed swelling in left buccal space. FNAC was suggestive of pleomorphic adenoma. The tumour was excised by sublabial approach. Differential diagnosis on postoperative histopathological examination was of EMEC and pleomorphic adenoma. The diagnosis was confirmed by IHC. IHC was positive for CK (epithelial component) and SMA, S-100 (myoepithelial component). Patient was asymptomatic till 2006 i.e. for 5 years.

Then patient again presented with diffuse fullness on left side of face extending from temporal fossa to maxillary region.

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On CECT showed large heterogonously enhancing mass in the left infratemporal region infiltrating the maxillary sinus, nasal cavity and left orbit. Tumour was excised by combined approach, total maxillectomy with removal of mass in left temporal region via trans zygomatic approach. Histopathological and IHC examination confirmed the diagnosis of EMEC and patient was subjected to chemotherapy.

Second patient, 28 year/male, presented with swelling in floor of mouth. The FNAC was pleomorphic adenoma. The tumour was excised and sent for histopathological examination. Histopathology revealed it to be EMEC. Patient was kept on regular follow up. Tumour recurred after 6 months. CT scan showed tumour approx. 3×3 cm infiltrating the floor of mouth with no involvement of mandible. Tumour was excised with wide margins with preservation of mandible. Floor of mouth was reconstructed with sternocleidomastoid flap. HPE revealed it to be EMEC and has some features of pleomorphic adenoma. The diagnosis was confirmed by IHC. IHC was positive for CK (epithelial component) and SMA, S-100 (myoepithelial component). Patient was subjected to post operative radiotherapy. There is no recurrence after 6 months of follow up.

Third patient, 17 year/male, presented with a firm swelling in retromandibular area. CECT scan showed mass involving deep lobe of parotid. FNAC revealed it be epithelial myoepithelioma. Total parotidectomy was done. Postoperative histopathological examination showed EMEC, confirmed by IHC which was positive for CK (epithelial component) and SMA, S-100 (myoepithelial component). The patient is on follow up for last 1 year with no recurrence (Figs. 1, 2, 3, 4, 5, and 6).

Fourth patient, 58 year/male, presented with firm swelling on the right side of hard palate. FNAC was suggestive of pleomorphic adenoma. Tumour was excised. HPE and IHC revealed it to be EMEC. IHC was positive for CK (epithelial component) and S-100 (myoepithelial component). Patient had positive deep margin, for which re-excision was done.

In all the four cases diagnosis was confirmed by immunohistochemistry as shown in the Table 1.

Discussion and Review of Literature

First case of EMEC was described by Donath et al. [1] in 1972 and recognized as a distinct pathologic entity in the 1991 WHO classification. It is a rare tumour, constituting 0.5% of salivary gland tumours [2]. We have one case of EMEC in floor of mouth, which is very rare. EMEC is usually seen in 60–80 year of age [1], but two of our cases were seen at less than 30 years. This tumour has a high

Fig. 1 Facial nerve branches after total parotidectomy for EMEC of deep lobe of parotid

Fig. 2 Sheets of cells with clear cytoplasm along with tubules Haematoxylin and Eosin ($\times 100$)

chance of recurrence and not infrequent distant metastasis [3]. Three out of four cases had recurrences in our study. It may be because we have treated these tumours as benign and had not taken adequate margins during tumour excision. But we have no case of distant metastasis.

FNAC is not conclusive. In all of our four cases, FNAC was suggestive of benign tumour. Three patients has features of pleomorphic adenoma while one patient has features of epithelial myoepithelioma. Histopathology showed two populations of cells i.e. clear cells and smooth muscle cells. It is difficult to differentiate EMEC from other clear cell tumours of salivary glands [4]. So the main differential

Fig. 3 Sheets of cells with clear cytoplasm along with tubules Haematoxylin and Eosin ($\times 400$)

Fig. 5 Immunohistochemistry slide (S-100 positive)

Fig. 4 Immunohistochemistry slide (cytokeratin positive)

Fig. 6 Recurrence of EMEC in floor of mouth

Table 1 Site, preoperative diagnosis, treatment, histopathology, IHC and recurrence of EMEC

Case	Site	FNAC	Treatment	HPE	IHC			Recurrence
					CK	SMA	S-100	
I	Left infratemporal region	Pleomorphic adenoma	Surgery + chemotherapy	EMEC	+	+	+	+
II	Floor of mouth	Pleomorphic adenoma	Surgery + radiotherapy	EMEC	+	+	+	+
III	Deep lobe of parotid	Epithelial myoepithelioma	Surgery	EMEC	+	–	+	–
IV	Hard palate	Pleomorphic adenoma	Surgery	EMEC	+	+	+	+

EMEC epithelial myoepithelial carcinoma, CK cytokeratin, SMA smooth muscle antigen

diagnosis is pleomorphic adenoma and clear cell tumours. So it is important to do immunohistochemistry in these tumours. We have done IHC with cytokeratin for epithelial component and S-100 and SMA (smooth muscle antigen) for myoepithelial component [5, 6].

Conclusion

EMEC is a rare tumour of salivary glands with high chances of recurrence; so complete removal of tumour is very important. There is no clear cut guideline to treat post

operative recurrence cases. Histopathology and immunohistochemistry is very important for diagnosis. This tumour can occur in younger age group.

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