



A Case of Epithelial–Myoepithelial Carcinoma in the Nasal Cavity Treated with Definitive Chemoradiotherapy

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SUMMARY

Epithelial–myoepithelial carcinoma (EMC) is an uncommon neoplasm that is predominantly seen in major salivary glands. Although it is considered as low grade, the high recurrence rate is quite common. EMC of the nasal cavity is extremely rare. A case of EMC detected in the nasal cavity is reported. A 44-year-old woman presented with a 1-year history of nasal congestion and bleeding. Magnetic resonance imaging (MRI) revealed a 4×4 cm necrotic expansile lesion involving the bilateral nasal cavity, extending to the nasopharynx, oral cavity, maxillary sinus, and ethmoid cells. The patient was found to be inoperable since complete surgical resection was not possible, and excisional biopsy was performed. Histopathological and immunohistochemical analyses led to a diagnosis of EMC. Definitive radiotherapy (RT) with concomitant cisplatin was applied. After 4 years, the patient is asymptomatic, and MRI showed a stable status with no progression. EMC is rarely seen in the nasal cavity. If possible, the first treatment modality should be surgery. The role of RT is controversial. To the best of our knowledge, this is the only case treated with definitive chemoradiotherapy for EMC in the nasal cavity.

Keywords: Chemotherapy; epithelial–myoepithelial carcinoma; nasal cavity; radiotherapy.

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Introduction

Epithelial–myoepithelial carcinoma (EMC) is an uncommon neoplasm that arises predominantly in the major salivary glands, especially in the parotid gland. [1] Rare cases have been reported in the nasal cavity and other head and neck regions. Very few cases have been reported outside the head and neck region, such as the vulva and breast. Although distant metastasis is rare, they are locally invasive. Histologically, EMC shows biphasic morphology. Heterogeneity of tumor that may contain four cell types, such as spindle, epithelioid, reticular, and clear cell, causes diagnostic pit-

fall.[2] Generally, tumor cells express calponin, S-100, cytokeratin, epithelial membrane antigen, smooth muscle actin, and P63.

These tumors have an excellent long-term survival. We report an extremely rare case of EMC of the nasal cavity and results of treatment. To our knowledge, this case is the third case treated with definitive radiotherapy (RT).

Case Report

A 44-year-old woman presented with nasal congestion and bleeding for 1 year. Interestingly, the patient's history revealed that she had also a small mass on her hard palate

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for 30 years. On endoscopic examination, the mass was seen occupying the bilateral nasal cavity and extending to the nasopharynx. On the other hand, there was a nodular mass with a diameter of 1 cm enclosed with normal mucosa on the hard palate. Magnetic resonance imaging (MRI) revealed a mass lesion, which was 4×5 cm in the transverse plane and 5 cm in the coronal plane (Fig. 1). The mass showed intense enhancement peripherally, and there was a necrotic area centrally. Hard palate and nasal cavity on both sides were involved. In addition, the mass was extending to the nasopharynx, oral cavity, left maxillary sinus, and ethmoid cells. There was also bony involvement on the inferomedial left bony orbit. With these imaging findings, the mass was considered to be inoperable, and excisional biopsy was performed.

Histopathological evaluation demonstrated that the tumor was composed of ovoid round cells with clear cytoplasm in a myxoid and hyaline stroma. The tumor was infiltrating the bone. In immunohistochemical analysis, tumor cells were found to express cytokeratin, calponin, P63, and GFAP diffusely and CK-7 and S-100 focally. The Ki-67 proliferation index of the tumor was 20%. There was no expression of synaptophysin, CD56, LCA, and CD20 detected. Finally, histopathological and immunohistochemical findings referred to EMC. The grade was not men-

tioned in the report. Definitive RT was applied due to inoperability. The patient received RT with concurrent cisplatin. Intensity-modulated RT was delivered to the primary tumor and bilateral level 1–3 lymph node regions with 6 mV X-rays using image-guided RT. Total radiation dose to the primary tumor and elective nodal regions were 70 and 50 Gy in 2 Gy fraction doses, respectively. After 4 years of follow-up, the patient was asymptomatic, and MRI showed stable disease with minimal regression (Fig. 2).

Discussion

EMC is a rare malignancy, and the majority of cases were reported in the major salivary glands. It has a slight female predominance, and the mean age of the patients at diagnosis is 60 years. It generally exhibits low-grade malignancy and is associated with favorable prognosis. However, it shows a locally infiltrative pattern and rarely metastasizes. We reported a case of EMC presenting with an invasive mass in the nasal cavity infiltrating the nasopharynx, maxillary sinus, ethmoid sinus, and hard palate with bone destruction. The patient's history of hard palate mass persisting for many years suggested that the disease has been growing very slowly.

The essential treatment modality is surgery, and surgical excision with negative margins is important.[3]

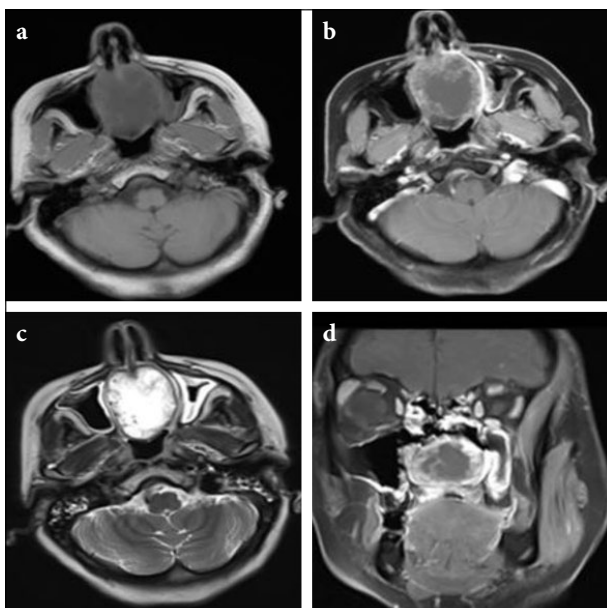


Fig. 1. MRI at the time of diagnosis. (a) Precontrast. (b) Postcontrast T1-weighted transverse sections show marked peripheral enhancement and central necrosis. (c) Fat saturated T2-weighted image. (d) Postcontrast T1-weighted fat saturated coronal image.

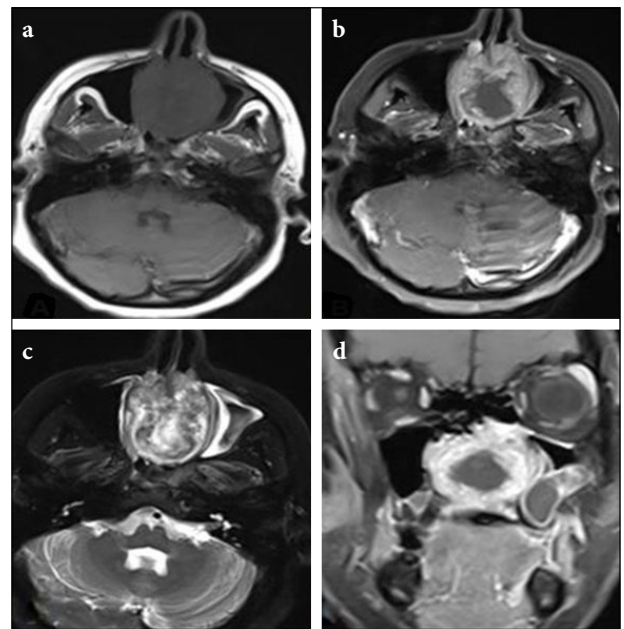


Fig. 2. (a) Precontrast T1-weighted image. (b) Postcontrast T1-weighted image. (c) Transverse T2-weighted fat saturated image. (d) Postcontrast T1-weighted fat saturated image.

The addition of postoperative treatment is controversial. Arora et al. reported that adjuvant RT can be applied in case of recurrence.[4] Moreover, adjuvant RT alone or concurrent with chemotherapy was reported to be effective for patients with myoepithelial carcinomas of the vulva with lymph node metastases.[5,6]

Lymph node involvement and distant metastases are rare.[1] However, the local recurrence rate is approximately 36%–42%.[3,7] Deer et al. reported that metastases to the periparotid and cervical lymph nodes occur in 10% of the cases. The most significant predictor factors for recurrence are surgical margin, lymphovascular invasion, necrosis, and myoepithelial anaplasia.[3]

Vazquez et al. analyzed the demographic, clinicopathologic, and survival features of 246 cases of EMC.[1] In this retrospective cohort study, which is the largest series of EMC, data were obtained from the Surveillance, Epidemiology, and End Results data (1973 and 2010). They reported that the survival results are excellent with a 10-year overall survival rate of 90.2%. They found that tumor size is a significant factor on survival. Patients with tumors <2 cm had a more survival advantage. In their study, 41.1% of patients received RT in addition to surgery, and survival benefit was not detected for patients who received adjuvant RT. Nevertheless, it was not a strong evidence since this was a registry-based study. The role of RT is not clear.

A definitive treatment protocol has not been determined since these tumors are seen rarely. Surgery is the main treatment. In the present case, the patient was treated with definitive RT due to inoperability and concurrent cisplatin, and the patient is free of any symptoms with stable disease on MRI after 4 years. In the literature, there are only two cases treated with definitive RT. One of the cases was published in 2004.[8] The patient was a 48-year-old man with stage T3N0M0 EMC at the base of the tongue. Surgery was rejected by the patient. First, two cycles of chemotherapy (cisplatin, doxorubicin, and 5-fluorouracil) were given. Then, using cobalt teletherapy unit, RT was applied at a dose of 66 Gy to the primary tumor and a dose of 50 Gy to the whole neck regions by conventional fractionation. The authors reported that a complete response was achieved.

The second case was published in 2015.[9] The patient was a 55-year-old man with myoepithelial carcinoma of the nasopharynx. The patient was applied chemoradiotherapy since the tumor was unresectable. However, there was still residual tumor after treatment.

Conclusion

In conclusion, to the best of our knowledge, our patient is the third case who was applied definitive RT in the

literature. Definitive RT appears to be an alternative for inoperable patients. Further cases will be necessary to define the optimal treatment protocol.

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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